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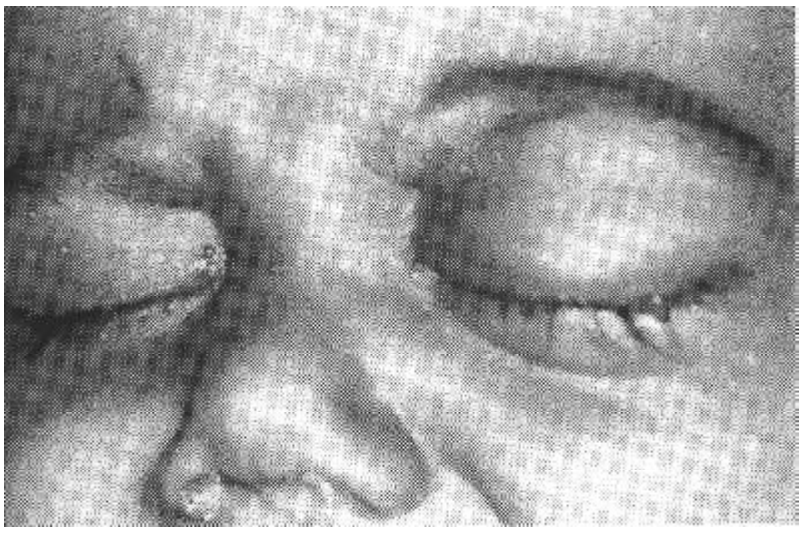


External clinical photograph depicting left orbital cellulitis

SANKARA NETHRALAYA

EDITORIAL

This issue of Insight comes to you as Sankara Nethralaya enters it's 25th year in service.

Ophthalmology is fast becoming a technology-based specialty. Phacoemulsifi-cation for cataract surgery, lasers for refractive surgery and of course, the technology driven vitreoretinal surgery exemplify this fact adequately. Adding to the list is the Wavefront technology, the current buzzword after LASIK in refractive surgery. Drs. Geetha Iyer and Srinivas K Rao explain the basics and effects on this technology on refractive surgery.

On the technology front again is the article on perimetry by R.Krishna Kumar which deals with the future of perimetry. This issue also has its complement of scientific articles including one on x-chrom contact lenses.

Dr Mahesh P Shanmugam
Dr Arun Narayanaswamy
Editors

INSIGHT & EYELIGHTS are now available on line at http://www. sankaranethralaya.org/publication.htm

Perspective:

Wavefront Technology

Iyer Geetha Krishnan and Srinivas K Rao

Refractive surgery for correcting the optical aberrations of the eye aims to make the eye optically perfect. In the past, the preoperative goal has been 20/20 uncorrected visual acuity with zero residual power. The new goal is 20/10 uncorrected visual acuity with aberration free postoperative vision.

Although recently introduced to ophthalmology, wavefront technology has been used for more than three decades to measure the quality of optical lenses, such as those used in astronomy. Data derived from wavefront technology allows the description and calculation of optical errors, and offers the promise of better understanding and treating human visual disorders.

WAVEFRONT

Wavefront describes the curve corresponding to multiple light rays emanating from the point source in the foveola after exiting the eye having passed through the different optical elements of the eye and are visualized through the pupil. Thus the wavefront describes the surface shape of the imperfections of the eye's optical system. In a perfect optical system, there are no distortions induced by the lens system. Such an ideal wavefront of an optical system with no aberrations is in one plane. A wavefront that is not free of distortions is said to have aberrations, which can be quantified mathematically using certain types of mathematical functions. Wavefront error is the difference between the actual and the ideal wavefront.

OPTICAL ABERRATIONS

When light rays pass from the tear film on to the cornea to the retina, they are bent and distorted by all the structures in the eye. The optical aberrations are classified as chromatic and monochromatic types. The degradation of image caused by the chromatic aberration is not analysed by the aberrometers. The monochromatic aberrations are measurable and are defined and quantified in terms of what are known as Zernicke's polynomials (ZP) consisting of Zernicke's terms (ZT).

The classic spherocylindrical correction of refractive errors that correct the optical aberration of defocus equals spherical correction (Zernicke term 4) and astigmatism (ZT 3 and 5) are familiar.

The so-called higher order aberrations are all the other aberrations except defocus (ZT4) and astigmatism (ZT3, 5) combined.

The most important and commonly used are the first 4 orders of ZP consisting of 14 terms of ZP, as follows:

0-order ZP (piston) - Term ZT0

1-order ZP (prism) - Term ZT1, 2 (tilt)

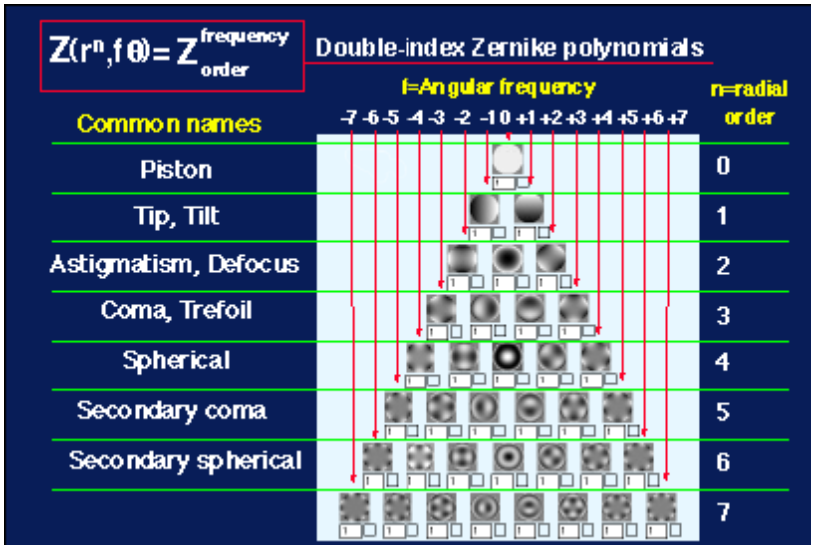
2-order ZP (spherocylinder) - Term ZT3, 5 (astigmatism), and ZT4 (defocus, sphere)

3-order ZP (coma) - Term 6, 9 (trefoil) and term ZT7, 8 (coma)

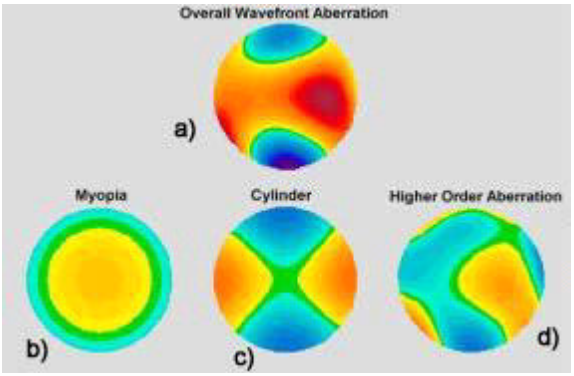
4-order ZP - Term ZT10, 14 (quadrafoil) and term ZT 11, 12, 13 (spherical aberration)

5-order ZP (secondary coma) - Terms ZT 15, 16, 17, 18, 19, 20 and so forth

In normal human eyes of young patients, third order ZP aberrations (ZPA) are responsible for approximately 40%, fourth order ZPA for 25%, and fifth to sixth order ZPA for 30% of higher order aberrations of the eye.



Visual catalog of the Zernicke modes, orders 0-7.



Measurement and evaluation of the different aberrations

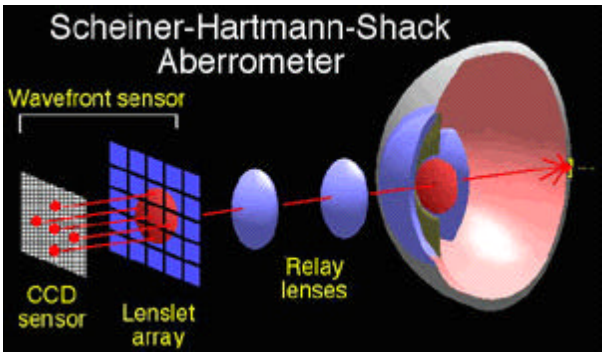
METHODS AND INSTRUMENTS MEASURING OPTICAL ABERRATIONS OF THE EYE

All methods of measuring the aberrations of the human eye evaluate how light that enters the eye is modified. With each of these approaches, light is imaged onto the retina, and either the image position on the retina or the wavefront as it emerges from the eye is measured.

1. Hartmann-Shack Aberrometry

Hartmann-Shack style devices are currently the most commonly used. These devices analyze an outgoing light that emerge or is reflected from the retina and passes through the optical system of the eye.

Light of specific characteristics is projected into the eye onto the macula. Rays that emerges from a single point in the foveola and pass through the optical system of the eye are analyzed. Multiple tiny lenslets in newer devices simulate apertures in the Hartmann disc located in front of the eye that isolate a narrow pencil of light emerging through different parts of the pupil. A charge-coupled device (CCD) camera registers the true position of each ray and compares it to the calculated reference position of such a ray for a perfect optical system of the eye without aberrations. This difference enables calculation of the aberrations of the true optical system of the eye.



2. Tscherning aberrometry

Tscherning aberrometry analyzes the ingoing light, which forms an image on the retina. A grid pattern formed by multiple spots is projected through the optical system of the eye and forms an image on the retina. This image is observed and evaluated by a method similar to indirect ophthalmoscopy and captured on CCD camera. The distortion of the grid pattern enables calculation of the aberrations of the optical system of the eye.

3. Ray tracing aberrometry

This device measures an ingoing light that passes through the optical system of the eye and forms an image on the retina. It measures one ray at a time in the entrance pupil rather than measuring all the rays at

the same time like previously mentioned devices. This decreases the chance of crossing the rays in highly aberrated eyes. The total time of scanning is 10-40 milliseconds.

WAVEFRONT vs. CORNEAL TOPOGRAPHY

Corneal topography recognizes complex patterns of the anterior corneal surface alone. Since the cornea provides nearly three quarters of the total dioptric power of the eye, corneal topography is an important diagnostic tool. But in order to measure the complete performance of vision of the eye, one needs to have also the information about optical distortions provided by the lens and the other optical media in the anterior and posterior chamber of the eye.

Wavefront measures the optical path in its entirety and is not limited to any given refractive surface. The cumulative sum of all the aberrations caused by all the eye structures is measured, though it is not possible to isolate the structure causing a particular aberration.

CUSTOM ABLATION

First the aberrometer is used to measure the wavefront distortions created by all the structures in the eye. Second, this information is used to guide excimer laser treatment to counteract the eyes' aberrations. Third the excimer laser uses a small spot-scanning beam to precisely place the custom ablation profile onto the cornea. This laser should have a fast eye tracker to null the saccadic eye movements that may degrade the effect of the precise ablation profile.

SUPERVISION

Using conventional laser ablation profile, patients with high corrections frequently report a decrease in functional vision when target contrast or the light intensity is reduced. Supervision is vision that is significantly improved over that provided by more traditional forms of correction. If optical aberrations in the eye could be limited; the theoretical limit of foveal acuity would be 20/8 to 20/10 (depending upon the biological variation in the foveolar receptor diameters in a particular eye). Beyond this, the retinal image will be under sampled causing the visual percept to be distorted. The benefits of aberration correction are much greater in the contrast domain than in the spatial domain (improving visual acuity).

Uncorrected image correction (e.g. by glasses)

Image after sphero-cylindrical correction

Image after wavefront aberration correction

CLINICAL APPLICATIONS

1. To objectively measure the refractive sphere and cylinder.
2. To compare with other measurement units such as corneal topography and verify the source of astigmatism.
3. To quantify and understand irregular astigmatism.
4. To carry out wavefront guided ablations.

CLINICAL LIMITATIONS

1. Analyzing the wavefront requires a clear optical system.
2. Lack of understanding of the effect of postoperative flap dynamics and wound healing on ocular aberrations.

3. The optimal wavefront characteristics for human vision are yet to be determined.

4. The dynamic fluctuations in the higher order aberrations of the eye are not well understood.

The field of refractive surgery is undergoing a major evolution and the wavefront places us at the threshold of a new frontier in ophthalmology.

Suggested Reading:

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Rapid Resolution of Posterior Scleritis following Intravenous Methylprednisolone Therapy

Rupak Kanti Biswas and Jyotirmay Biswas

Posterior scleritis remains one of the under diagnosed treatable conditions in ophthalmology. Awareness of the entity, typical clinical features, ancillary tests like fundus fluorescein angiography and in particular ultrasonography can establish the diagnosis in majority of cases. The various treatment modalities described are retrobulbar injection of corticosteroid, oral steroid, non-steroidal anti-inflammatory agents (NSAIDs) and systemic immunosuppressive agents[1]. Intra venous methylprednisolone (IVMP) has been described in severe uveitis, but its role in posterior scleritis has been reported rarely in literature. Mc clauskey et.al reported successful resolution of 13 cases of severe anterior scleritis and 1 case of posterior scleritis[2]. We report a case of posterior scleritis with profound, painless visual impairment, which resolved successfully following a course of 3 days of IVMP.

CASE SUMMARY:

A 49year old female presented with sudden onset of decreased vision in right eye of 1 day duration, which was not associated with pain or redness. She was also complaining of seeing black area in central region of the right eye. There was no complaint in left eye. Her past history did not reveal any systemic diseases like diabetes, hypertension. There was no history of joint pain, skin rashes, erythema or lymph node enlargement.

On examination her best-corrected visual acuity was counting finger at ½ meter, <N36 in right eye and 6/6, N6 in left eye. Pupillary reaction was brisk and well sustained in both eyes with no relative afferent pupillary defect. Slit lamp examination revealed unremarkable findings except early posterior subcapsular cataract in both eyes. There were no active

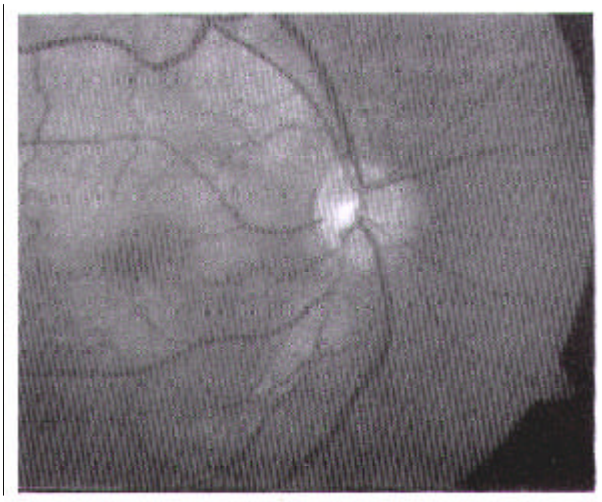

Fig.1a - Fundus photograph showing large subretinal mass with secondary retinal detachment involving posterior pole of right eye.

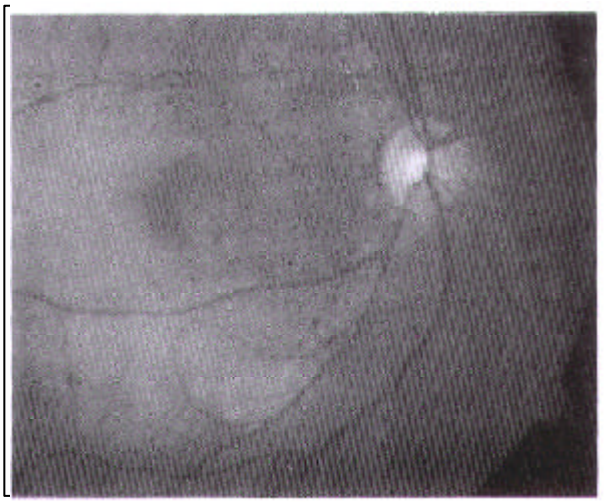

Fig.1b -- Fundus photograph following 3days of IVMP therapy showing marked resolution of fundus lesion.

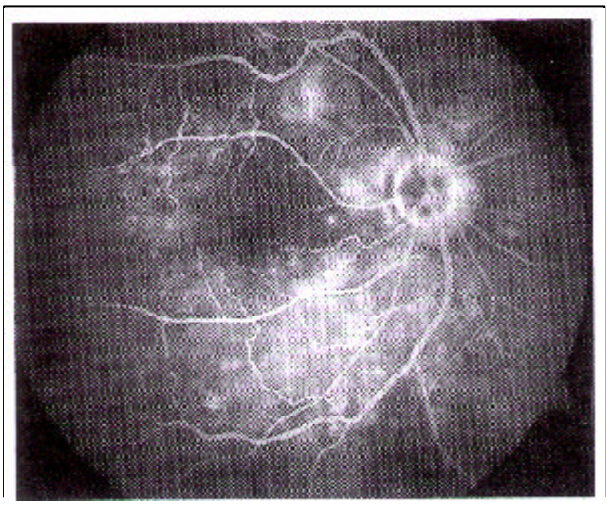

Fig.2 -- Late arteriovenous phase of fundus fluorescent angiogram showing mottled hyperflourescence in posterior pole and pinpoint hyperfluorescent dots.

signs of inflammations in the anterior segment in both eyes. Fundus evaluation of right eye with indirect ophthalmoscope revealed elevated lesion of approximately 8mm x 3mm involving the macular area with white subretinal precipitate and retinal striae (Fig.1a). Fundus evaluation of left eye with indirect ophthalmoscopy showed few areas of pigment epithelium alteration nasal to the disc, rest was within normal limit. Ultrasonography of right eye showed localised retinal elevation at macula with choroidal thickening of 2.8mm (Fig.3a). Optic disc was normal. Fundus fluorescein angiography of right eye showed pinpoint hyperflourescence and pooling of dye in late stages (Fig.2). Patient was clinically diagnosed to have posterior scleritis. She was given 3 doses of 1gm intravenous methylprednisolone (IVMP) daily after physician clearance. After the 3rd dose of IVMP her best-corrected visual acuity was improved to 6/6, N6 in right eye. Fundus evaluation of right eye revealed marked resolution and flattening of the lesion (Fig.1b). Post IVMP ultrasonography of right eye showed marked decrease in retinal elevation and choroidal thickness (1.7mm) (Fig.3b). She was advised to take oral Prednisolone 60mg per day, which was tapered gradually at 10mg per day per week.

On follow up after 2 weeks, her right eye vision was maintained as 6/6, N6. Fundus evaluation with indirect ophthalmoscope revealed regressed posterior scleritis in right eye. Ultrasonography of right eye showed further decrease in choroidal thickness (1.4 mm).

DISCUSSION:

Posterior scleritis usually mimics other conditions like choroidal mass or central serous retinopathy. Most common presenting symptoms are visual impairment and pain of varying severity, although diplopia, flashes and pain in ocular movement may also be present. The visual impairment may be mild when it is simply due to transient hyperopia

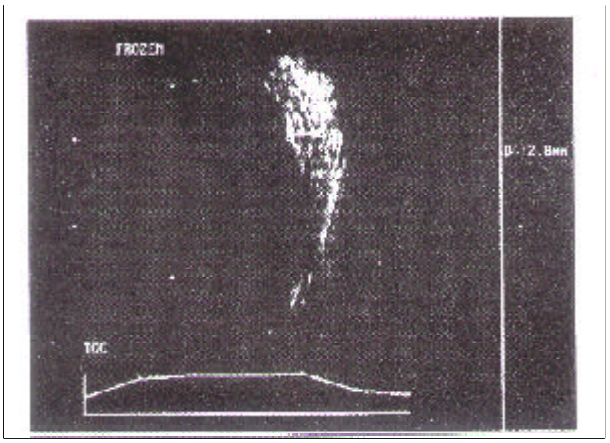

Fig.3a - Ultrasonography of right eye showing marked choroidal thickening (2.8 mm) with retinal elevation.

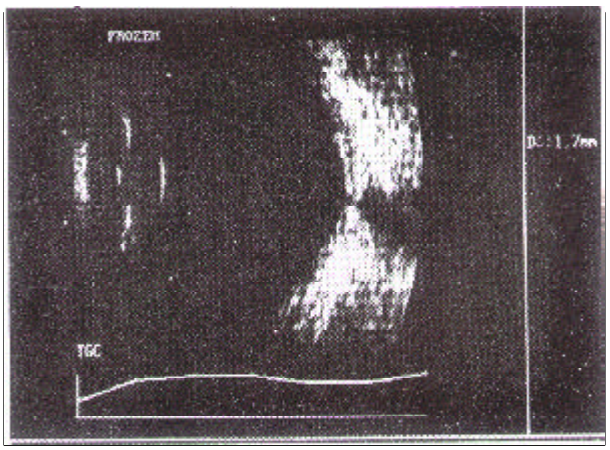

Fig.3b - Ultrasonography of right eye showing decreased choroidal thickening (1.7 mm) following 3 doses of IVMP therapy.

resulting from posterior scleral thickening[3]. But when the inflammation involves the macular area or there is cystoid macular edema - there may be severe visual impairment. Retinal detachment or optic neuritis are the complication of posterior scleritis which can also cause moderate to severe visual loss.

Pain, similarly, is also a consistent feature of posterior scleritis, and was surprisingly absent in our case, producing diagnostic dilemma. Pain occurs due to the stretching of the Tenon's capsule by edema, stretching of scleral sensory nerve endings by edema or due to optic nerve sheath swelling[3]. When the muscle sheath is involved, there may be pain with ocular movements.

The common fundus findings in a case of posterior scleritis include choroidal folds, sub retinal mass, disc edema, macular edema. Our case presented with elevated retina in macular region with sub retinal precipitates, mimicking sub retinal mass. There were horizontal retinal striae surrounding the macular area (Fig 1a).

In absence of typical signs and symptoms, diagnosis is confirmed by ultrasonography, which shows the characteristic flattening and thickening of posterior coats and T-sign. The characteristic T-sign occurs due to collection of fluid in subtenon space and around optic nerve, which produce a lack of echoes in the edematous areas[3]. In this case though there was gross choroidal thickening but T-sign was absent. Fundus fluorescein angiography was done to rule out central serous retinopathy. Angiography did not show any focal leak, but showed pooling of dye in the late phases, with multiple pinpoint areas of hyper fluorescence. The choriocapillaries showed mottled fluorescent pattern, suggestive of choroidal involvement.

On systemic examination, she was not having any evidence of vasculitis, erythema, skin rash. There was neither any evidence of joint abnormality or deformity, nor any enlarged lymph node. Mc Clauskey and coworkers reported that 29% of posterior scleritis had associated systemic diseases, the risk of which increases after age of 50 years[4].

Management modalities also vary according to the severity of the disease and the area of involvement[1]. The various modalities include NSAIDs, high doses of systemic steroids, systemic immuno-suppressive therapy, retrobulbar steroid injection and rarely IVMP in very severe cases. Systemic steroid and NSAIDs are the mainstay of treatment in mild to moderate cases of posterior scleritis not involving the macula. Treatment started with 1mg/ kg/ day of oral steroid and adjusted according to the response to treatment. Non-steroidal anti-inflammatory agents are also important in controlling the inflammation. However in refractory cases systemic immunosuppressive agents like cyclophosphamide in the dose of 2mg/ kg/ day in two divided doses is recommended[5]. Mc Clauskey and coworkers showed one case series of 14 cases in which they have mentioned 1case of posterior scleritis resolved successfully following IVMP therapy[2]. They have suggested IVMP as the treatment in severe form of ocular inflammatory diseases[6]. In our case as the lesion was involving the macula, we treated her with 3 doses of 1gm/ day of IVMP. It showed a dramatic and almost complete resolution of lesion as well as visual improvement. Wakefield and coworkers reported the side effects of IVMP, which included psychological disturbances, hypertension, elevated blood glucose levels, but cessation of treatment was not necessary in any patient[6]. Transient irregularity of heart rate with ectopics is also mentioned. Our patient had experienced only transient rise of blood sugar level for which cessation of treatment was not needed and it had come down to normal level after completion of treatment.

Our report indicates that an acute posterior scleritis with profound visual impairment can be managed by IVMP therapy. Such treatment can cause rapid resolution with good visual outcome. However one should keep in mind, the side effects of and risk

involved in intravenous methylprednisolone therapy.

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Bilateral Retinal Vascular Occlusion Secondary to Septic Emboli arising from a Scrotal Ulcer in A Child – A Case Report

Amit Nagpal, Jyotirmay Biswas and Kannan M Narayanan

ABSTRACT

A 4-year old child with features of bacterial septicemia and a scrotal ulcer of 20 days duration presented with acute onset profound bilateral loss of vision. Examination revealed central retinal vein occlusion in one eye and branch retinal artery occlusion in the other eye. The child received intravenous methyl prednisolone, antibiotics and ACTH. At 3 months follow up there was partial visual recovery in one eye.

This retrospective interventional case report illustrates that pediatric patients presenting with retinal vascular occlusion should be thoroughly examined for the septic foci. Aggressive therapy with intravenous steroids and antibiotics may be useful in restoring functional vision.

INTRODUCTION

There are a few isolated case reports of combined retinal vascular occlusions in the literature.[1-5] In most of these instances the occurrence was unilateral. One third of these cases are reported to be due to retrobulbar injection[6]. Other causes reported include vasculitis, neoplastic infiltration of optic nerve and septic emboli from sources like infective endocarditis. We further report a case of bilateral vascular occlusion in a 4 year old child, in whom there were concomitant features of bacterial septicemia secondary to a scrotal ulcer. There was partial visual recovery in one eye at 3 month follow up.

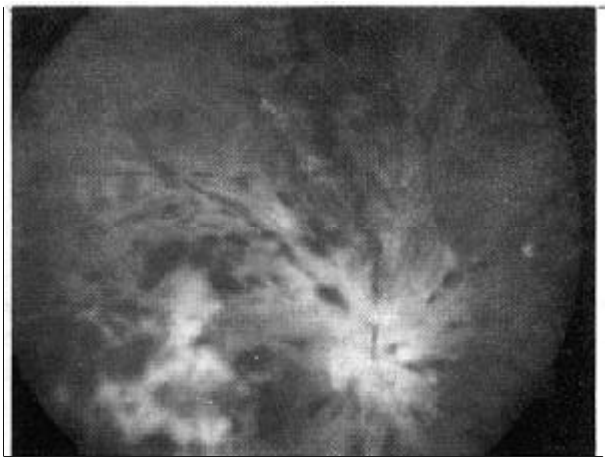

FIG 1: Fundus photograph of right eye at the first presentation, suggestive of central retinal vein occlusion.

CASE REPORT

A 4-year old boy was referred to us with the complaint of sudden painless profound loss of vision in both eyes. He had fever, erythematous maculopapular rash, anasarca, multiple generalized lymphadenopathy, hepatosplenomegaly and a scrotal ulcer for the last 20 days. Echocardiography was done to rule out infective endocarditis. A diagnosis of gram-negative septicemia was made and intravenous antibiotics; ceftazidime, ofloxacin

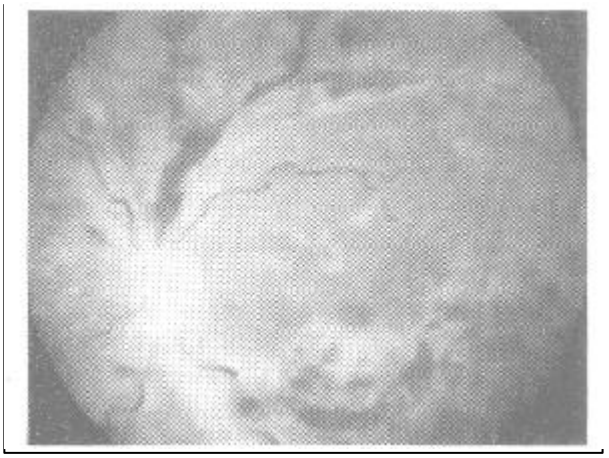

FIG 2: Fundus photograph of the left eye at the first presentation, suggestive of inferotemporal branch retinal artery occlusion secondary to arteritis.

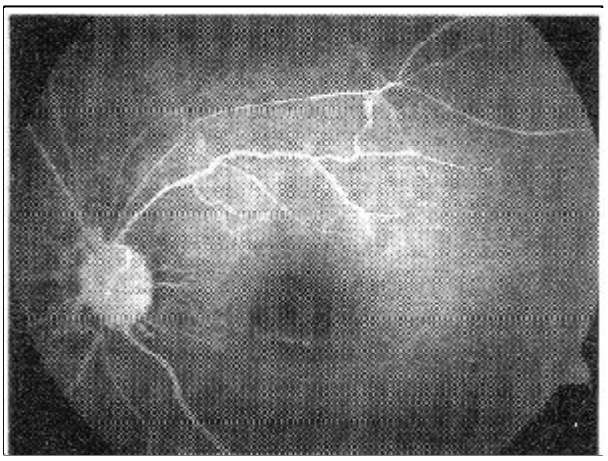

FIG 3: Fundus Fluorescein angiogram of the left eye showing collaterals.

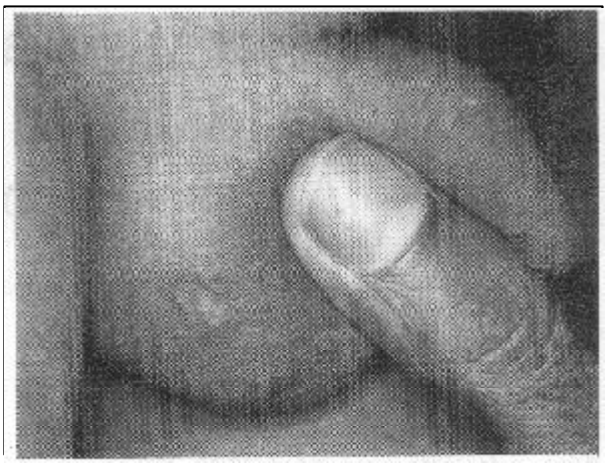

Fig 4: External photograph of the healed scrotal ulcer.

and metronidazole were administered, following which he developed sudden loss of vision in both the eyes for which he was given intravenous methyl prednisolone and ACTH. A computerized tomography scan of brain was done which was with in normal limits.

On ophthalmic examination the child denied perception of light in both the eyes. Examination of the anterior segment revealed no abnormality except ill sustained pupillary reactions in both the eyes. The anterior vitreous face showed 2 + cells in both eyes. Fundus examination of the right eye revealed edematous and pale optic disc with blurred margins, splinter hemorrhages, sclerosed and tortuous retinal blood vessels, sub retinal exudation and multiple dot and blot hemorrhages scattered in all the quadrants (Fig 1). Fundus examination of the left eye showed pale optic disc, sclerosed blood vessels, with segmented blood columns, multiple hemorrhages along the arcades, pale-edematous retina with superficial retinal opacification and a partial cherry red spot formation (Fig 2). Fluorescein angiography of the left eye showed severe retinal capillary non-perfusion and abrupt termination of the mid sized blood vessels along with collateral vessels on the disc and near superotemporal arcade (Fig 3). The culture of the scrotal ulcer revealed streptococcus viridians. A diagnosis of central retinal vein occlusion in the right eye and branch retinal artery occlusion in the left eye secondary to arteritis was made as a cause vascular obstruction due to septic emboli from the scrotal ulcer (Fig 4). Guarded visual prognosis was explained. The patient was advised to continue the antibiotics and oral steroids. On follow up after 3 months, the best-corrected visual acuity was no perception of light in the right eye and 6/60 in the left eye. Fundus examination of the right eye showed extensive fibro vascular proliferation over the disc (Fig 5). Left eye showed a pale optic disc with sclerosed blood vessels, regressed retinal hemorrhages, pigmentary changes and relatively avascular retina (Fig 6).

The child was seen after 4 months, there was no perception of light in the right eye and vision in the left eye remained 6/60. The fundus examination revealed the similar picture as before.

DISCUSSION

Combined occlusion of central retinal artery and vein in pediatric patients has been reported in the literature, the reported risk factors were; infective endocarditis [1,2] inflammation, coagulopathies, neoplasia [3-5], blunt trauma [7], and systemic lupus erythematosus.[8] Our case is unique because one eye had central retinal vein occlusion and the other had a branch retinal artery occlusion secondary to septic emboli from a scrotal ulcer. The pathogenesis was assumed to be a disruption of the arterial blood flow in to the retina by infectious emboli that caused

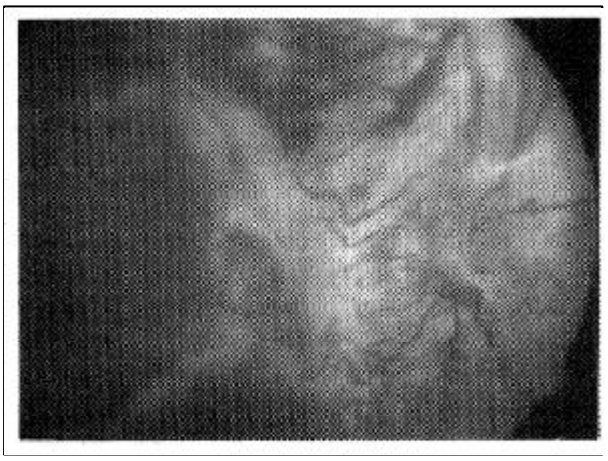

Fig 5: Fundus photograph of the right eye at 3 month follow up showing extensive fibrovascular proliferation over the optic disc as a sequelae of CRVO.

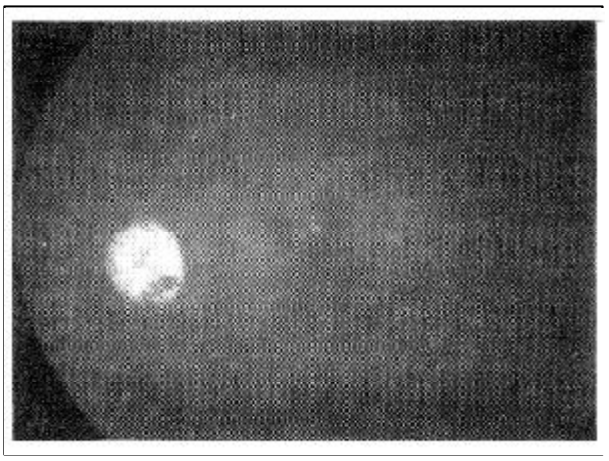

Fig 6: Fundus photograph of the left eye at 3 month follow up showing pale disc and severely sclerosed vessels as a sequelae of Branch retinal artery occlusion.

stagnation of capillary blood flow and venous thrombosis. None of the administered antibiotics have been reported so far in the literature as a cause of retinal vascular occlusion. To our knowledge this is the first case in a pediatric patient of bilateral vascular occlusion secondary to infective emboli other than infective endocarditis. Ophthalmologists should be aware of the fact that a thorough systemic examination and investigations of all the patients with retinal vascular disease is mandatory to rule out an underlying septic emboli.

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X-Chrom Contact Lens

S Premnandhini

Introduction:

Colour vision plays a crucial role in our day today living. Though a concept taken for granted it has enormorous clinical applications. Colour vision appears to be well developed soon after birth, by the age of 2 or 3 months, the infant has good colour discrimination ability as that of adults[1].

Most clinicians are aware that colour vision is anomalous in a relatively large percentage of the male population and a much smaller percentage of women. About 1 in 12 males and 1 in 200 females have anomalous colour vision of the Red-green type, but only 1 in every 50,000 males & females have inherited anomalous Blue-Yellow vision[2]. This defect is inherited constant throughout life and cannot be cured in the usual clinical sense.

As colour vision is being used more in the work place, poor performance in a job may be related to defective colour vision. Mostly the hereditary linked colour vision deficiency goes undetected, as the person will be 'asymptomatic'. Mostly, its only during the time of vision screening that many get to know about their colour deficiency, sometimes it might be during a crucial time of a job selection.

The current increased relevance of colour vision in the workstation demands a parallel increase in interest in assisting persons with colour defects. One of the popular ways of assisting is the use of x-chrom contact lens.

Background knowledge:

Dichoptic colour experiments were performed with colour normals and with protanopes and deutranopes. Experiments indicate that the cortical perception achieved by summing input of each eye when one eye has a filter in front of it may aid colour discrimination for dichromats[3].

The red filter when used gives brightening of reds relative to other colours. These changes in relative brightness can of course be seen by any person whether his colour vision is norma or defective. With the red filters, the green becomes darker and red remains bright. For colour-normal person, this tells him little more than he had already seen, but for the dichromat this is new information and he can make the Red-Green distinctions, which he couldn't make before[4].

It should be noted that, **the coloured filters enables to differentiate between certain colours and are definitely not "cures" for colour defects.** There is no known filter that allows a colour defective to have a normal colour vision.

X - Chrom contact lens:

The best-known filter that is specifically promoted as an aid for colour defective patients is the X-Chrom lens, which is available as a hard and soft contact lens. Indeed, the X-Chrom contact lens is a red lens that allows some blue light transmission. In 1971 Zeltzer introduced the hard X-chrom contact lens in polymethyl meth acrylate for wear on one eye[5]. The lens is not only intended to produce differences in intensity between the two eyes[6,7]. The lens helps colour defective patients whose occupation calls for differentiating browns, greens, oranges, reds, purples and blues. Its usefulness probably results from the brightness difference cues for different colours in each eye, the stereolustre effect that occurs when objects on one eye are very different from the other and small chromaticity (colour) shifts that an be produced by this filter.

The X-Chrom contact lens is worn on the non-dominant eye of colour deficient individuals (dichromats & anomalous trichromats). The fitting procedure is the same as that of any conventional contact lens.

Clinicians are cautioned that the clear improvement of performance on Ishihara, Drorine and AO pseudoisochromatic plates with this lens, or infact most red filters, does not reflect comparable improvement in colour discrimination in the real world. The improved performance in the test plates can be attributed to the fact that a red filter essentially destroys the carefully balanced design of these test plates[2].

X-Chrom lens can be extremely helpful in distinguishing colours in an array and simply doesn't give an overall improvement in colour discrimination.

Points To Remember:

- Care should be taken not to disrupt the binocular vision when prescribing X-Chrom lens in one eye
- Patient should be cautioned about wearing the lenses at night
- The patient may experience a false distance preception of any and all objects in motion; this phenomenon is called pulfrich phenomenon8.
- Due to the intensity difference between the eyes, patient may report a lustre or glittering appearance of red objects. The patient can learn this and other chromatic and brightness differentials.

For totally colour-blind patients, filters that cut out short wavelengths (blue) can be extremely effective in improving outdoor vision[2].

Case 1:

A 30 year old man came to our outpatient clinic with difficulty in colour perception. He also reported that he was denied a government job due to this deficiency. There was no history of any systemic illness. His best corrected visual acuity was 6/6+; N6 in each eye. Anterior and posterior segment did not reveal any abnormality.

His colour vision recorded with the pseudo-isochromatic chart was noted as follows:

OD: 1/14 (correct response)

OS: 2/14 (correct response).

An X-chrom contact lens was fitted in his non-dominant eye (OS) and the response recorded was OU: 8/14 (correct response)

The lens maintenance and handling was explained and the patient was instructed to use the X-chrom contact lens only during the time of need.

Case 2:

A 27 year old male reported to our clinic with complaint of difficulty in colour perception. As the patient had an occupational need, he was interested to go in for X-chrom contact lens.

His initial colour vision was 0/14 (either eye), tested with Ishihara's test plates. With red filter in either eye (OD / OS) the correct response was noted as 1/14 (measured binocularly).

With red filter in both eyes, colour vision was noted as 14/14 correct response.

Usually a X-chrom contact lens will be fitted only in one eye (non-dominant eye). Case 1 obeys this rule whereas in Case 2, interestingly, the patient improved only with X-chrom contact lenses in both eyes.

The speciality of colour vision is vast and the use of X-chrom contact lens is only at its incipient stage in our country. The second case presented in this paper makes us wonder about the complexities involved in colour vision and more research is needed in this area.

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Retinoblastoma mimicking Orbital Cellulitis

Manisha Agarwal, Jyotirmay Biswas, Krishna Kumar and Mahesh P Shanmugam

Introduction

Retinoblastoma commonly presents as leukocoria and strabismus; rarely as uveitis, spontaneous hyphaema, preseptal cellulitis, pseudohypopyon, secondary glaucoma and phthisis bulbi.[1] Orbital cellulitis like manifestation, though rare has been previously reported by Shields et al[2] and Mullaney et al,[3] the prevalence being 1.3% in 450 cases and 4.8% in 292 cases respectively. Many previously described cases of retinoblastoma presenting as orbital cellulitis, have had marked associated anterior segment involvement, including corneal opacification and enlargement, glaucoma, uveitis, and rubeosis iridis.[2,3]

We report the clinical and histopathological features of four retinoblastoma patients presenting with orbital cellulitis. The orbital cellulitis like picture may be misdiagnosed to be of infective etiology leading to improper medical treatment, delaying institution of appropriate treatment such as enucleation. This clinical implication and the rarity of such a presentation make it important to recognize orbital cellulitis like presentation of retinoblastoma.

Case reports

Case -1

In 1991, a 21-month-old male child presented to us with diminution of vision, lid edema and ecchymosis, steamy cornea, shallow anterior chamber, minimal hyphaema and low intraocular pressure (IOP) in the right eye, of 3 weeks duration. His left eye was buphthalmic with corneal vascularization, multiple macular opacities, anterior chamber calcific deposits and normal IOP. Fundus examination could not be done in either eye due to corneal opacity. Ultrasound and CT-Scan examination of orbit showed intraocular mass lesions with calcification suggestive of retinoblastoma. The child underwent enucleation of his left eye. Histopathological examination of the enucleated eye showed increased corneal diameter with iris adherent to posterior corneal surface, ciliary body atrophy and disorganization, presence of large undifferentiated basophilic tumor cells interspersed with extensive areas of necrosis, calcification, haemorrhage and fibrous tissue. Tumor cells extended till the posterior corneal surface involving the angle, which was closed. Focal choroidal and pre-laminar optic nerve invasion was present.

The child was lost to follow up and examined again after a month, with right eyelid edema, chemosis, mucopurulent discharge, steamy cornea and shallow anterior chamber. Histopathological examination of the enucleated right eye showed poorly differentiated and markedly necrotic tumor filling the vitreous cavity with ciliary body necrosis, choroidal and extrascleral invasion. The child was referred to the local cancer hospital for chemotherapy and radiotherapy.

Case-2

In 1995, a 1-year-old male was brought to us after the parents noticed a white reflex in the left eye, following a fall. The left eye showed signs of orbital cellulitis such as periorbital edema, proptosis, conjunctival chemosis, buphthalmos, corneal edema, extensive rubeosis with dilated and nonreacting pupil, leukocoria and no view of the fundus. IOP could not be assessed due to periorbital edema. Right eye fundus showed Reese Ellsworth stage 5B retinoblastoma. Ultrasound and CT- Scan of left eye showed evidence of retinoblastoma with suspicion of extraocular extension.

Left eye was enucleated which on histopathological examination showed closed angle, poorly differentiated and extensively necrotic tumor filling the vitreous cavity, iris necrosis, iris and ciliary body haemorrhage, focal choroidal invasion, post-laminar optic nerve invasion and no extrascleral extension (Figure-1). The patient underwent external beam radiation for the right eye, following

which the tumor regressed. The child was subsequently lost for follow up.

Case-3

In 1998, a 3- month- old male child presented to us after the parents noticed a white reflex in the left eye, ten days after birth. Left eye was buphthalmic and showed lid swelling, circumcorneal congestion, chemosis, corneal haze, leukocoria with no view of the fundus and restricted ocular movements. IOP was raised. (Figure-2)

Ultrasound and CT-Scan orbit showed preseptal swelling and an intraocular mass lesion with no evidence of extraocular extension on the left side, the probable diagnosis being retinoblastoma.

The patient was treated with oral systemic steroids (Prednisolone acetate - 2.5 mg/day) for 2 days following which the lid edema and orbital reaction decreased minimally. Left eye was enucleated and on histopathlogical examination it showed closed angle , ectropion uveae, tumor cell invasion of ciliary body and anterior chamber, rubeosis iridis, a poorly differentiated and necrotic tumor in the vitreous cavity with choroidal and pre-laminar invasion of the optic nerve. Right eye with stage 3A Retinoblastoma, confirmed on ultrasound (Figure-3) was initially treated conservatively. The tumor did not respond to conservative treatment and the child was lost to follow up after partial treatment. Subsequently the child presented with advanced retinoblastoma with extra-ocular disease for which exenteration was advised. Parents refused further treatment.

Case-4

In 2001, a 1 ½ year old boy presented with squint of 6 months duration and right eyelid edema, chemosis forming a prolapsing mass and hazy cornea, of 1 week duration. Right eye fundus showed a large retinoblastoma tumor filling the entire vitreous cavity. Left eye showed leukocoria and stage 5 retinoblastoma. CT-Scan showed bilateral mass lesions suggestive of retinoblastoma with no extraocular extension. The child was treated with oral systemic steroids (Prednisolone acetate- 5 mg/day) for three

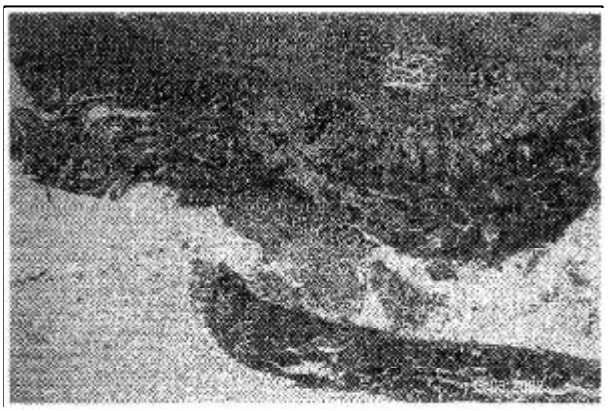

FIG-1 Photomicrograph of retinoblastoma (Hematoxylin and eosin)showing extensive necrosis, iris and ciliary body necrosis.

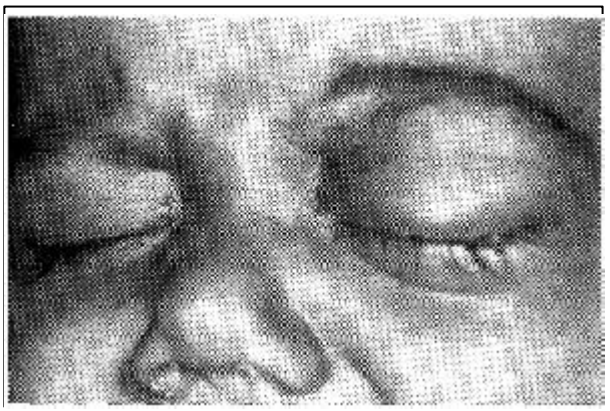

FIG-2 External clinical photograph of case-3.

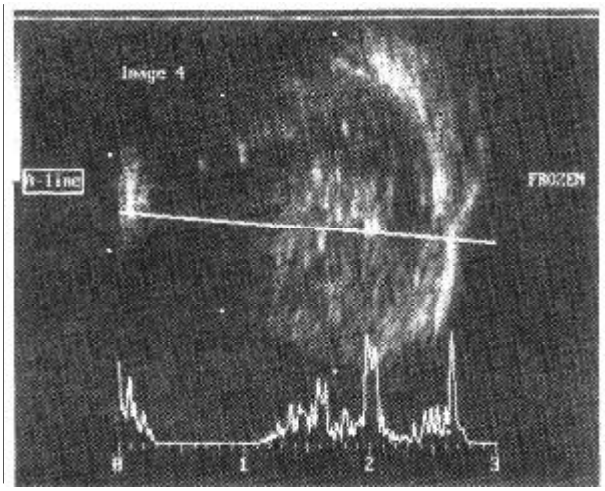

FIG-3 Ultrasound showing intraocular mass with no extraocular extension.

days prior to right eye enucleation. Histopathlogical examination of right eye showed peripheral anterior synechiae, iris necrosis and haemorrhage, a well

differentiated, extensively necrotic tumor with invasion of anterior chamber and pre-laminar optic nerve without extraocular extension. The child was advised chemotherapy for left eye. Two months later the left eye tumor remained active with secondary retinal detachment and vitreous haemorrhage. He was advised enucleation, which the parents refused and was then lost for follow up.

Discussion

Orbital cellulitis is an uncommon presentation of retinoblastoma.2The pathogenesis of this response is unknown. Shields et al have suggested that the inflammatory signs could be secondary to a necrotic tumor that outgrows its blood supply or the tumor in one eye induces an immune response which may lead to necrosis of the tumor in the fellow eye.[2]

Large necrotic tumors have a tendency to present with orbital cellulitis like picture.3 Anterior segment involvement by the tumor has commonly been found in patients with Retinoblastoma presenting as orbital cellulitis.[3] Meir et al and Haik et al have suggested anterior segment involvement with necrosis of iris and ciliary body as a trigger to inflammation in adjacent orbital soft tissues.[4,5] Tumor necrosis with leaching of necrotic products has been suggested as a mechanism by Mullaney et al.[3]

In all our patients, anterior segment involvement by the tumor was noted. In addition, ciliary body and iris necrosis and invasion were also noted in all the patients. Three of the five enucleated eyes showed angle closure and one showed peripheral anterior synechiae. Two had rubeosis iridis as well. All had large tumors with extensive necrosis. Three out of five eyes had increased corneal diameter. (Table-1)

The pathogenesis of orbital cellulitis like reaction may be multifactorial. Large tumors invading the anterior segment may outgrow the blood supply leading to necrosis, inflammation and neovascular reaction. In addition, necrosis of the ciliary body and iris may also induce the associated soft tissue inflammation. Angle closure was seen in all our patients, secondary to either neovascularization or the mechanical effect of

Table-1 Histopathological characteristics

S. No.	*Eye*	Type of tumor	*Rubeosis*	Necrosis	*C.B Invasion*	Optic nerve invasion			Choroid invasion	Extraocular extension	Anterior segment involve.
						LAMINAR		Surg. End Invo.			
						Pre	Post				
1.	OD	Undifferentiated	-	+	+	+	+	-	+	+	C.B necrosis & closed angle
	OS	Undifferentiated	-	+	-	+	-	-	+ Focal	-	C.B atrophy, Angle closed & invasion+
2.	OS	Undifferentiated	+	+	-	+	+	-	+ Focal	-	C.B +Iris necrosis & haem, closed angle
3.	OS	Undifferentiated	+	+	+	+	-	-	+	-	Angle closed &invasion+
4.	OD	Well differentiated	-	+	-	+	-	-	-	-	PAS + Iris necrosis + haem.

the tumor pressing the iris-lens diaphragm forwards. This may lead to increased IOP causing further ischaemia and necrosis of the tumor, thereby setting up a vicious cycle. Thus the combination of raised IOP and extensive inflammation due to extensive tumor necrosis, ciliary body and iris necrosis may give rise to orbital cellulitis like reaction. Despite most of the previously reported cases of orbital cellulitis not having extraocular invasion,one eye in our series and one reported by Mullaney et al showed extraocular disease. Hence a careful pre-operative orbital imaging with CT-Scan or MRI is very essential to rule out extraocular disease in these patients.

Most of the children with orbital cellulitis like reaction present with a hazy media obscuring fundus details. The presence of pseudohypopyon, anterior segment inflammatory signs may also mimic an inflammatory etiology and not retinoblastoma. The differentiating features are absence of systemic symptoms such as high fever, history of frontoethmoidal sinusitis, normal pupils and fundi on examination, leukocytosis, positive blood culture and cloudy sinuses on investigation.[2] However in all children with orbital cellulitis like presenting picture where the fundus could not be seen, an imaging study is essential to rule out intraocular retinoblastoma. The difference on CT-Scan being that retinoblastoma appears as an intraocular mass lesion often with calcification and in cases of true infectious cellulitis the soft-tissue swelling is confined to the anterior portion of the orbit adjacent to a cloudy sinus.[2] Delay in the diagnosis of retinoblastoma can lead to possible extraocular spread.

The orbital cellulitis like reaction is usually managed with 3 days of oral corticosteroids resulting in reduction of the perineural soft-tissue edema which is often responsible for false-positive interpretation of the tumor invasion into the optic nerve on CT-Scan.[6] Intravenous prednisolone in the dose of 1mg/kg which can be increased to 1.5mg/kg if the resolution is slow, has also been advocated.[3] In most of the children the inflammation decreases after 5 days of treatment to enable enucleation. It is essential to decrease the associated orbital inflammation preoperatively to facilitate enucleation and obtaining a long optic nerve stump.

In conclusion, advanced necrotic retinoblastoma often involves the anterior segment and can give rise to orbital cellulitis like picture. It is essential to differentiate this from infective orbital cellulitis to enable proper management.We recommend that all children presenting with signs of orbital or ocular inflammation should have a complete ophthalmic examination including a fundus examination. If the fundus is not visible or in case of doubt, ultrasonography, CT-Scan or MRI should be performed to exclude the diagnosis of retinoblastoma.

However one should not presume the absence or presence extraocular extension of the retinoblastoma based on only clinical findings.

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(contd. on page 24)

geniculate body. Most ganglion cells in the magnocellular lamina of the lateral geniculate body give a more transient and phasic response and are sensitive to motion. The retinal ganglion cells that project to the magnocellular lamina (M cell) usually have larger cells. M cells constitute approximately 10% of the total number of retinal ganglion cells and have less redundancy than the P cell. When a low spatial-frequency sinusoidal grating undergoes high temporal-frequency counter phase flicker, the grating appears to have twice as many light/dark bars and its spatial frequency appears to be doubled. This phenomenon is known as the frequency doubling illusion. The illusion is believed to reflect the function of a subset of M cells [3]. Recent studies of glaucomatous pathology have demonstrated that the retinal ganglion cells with larger cell bodies are selectively damaged earlier in the disease process of glaucoma[4]. Frequency doubling perimetry (FDP) operates based on the frequency doubling illusion. A prototype device (Welch Allyn, USA) is available for the FDP measurements. The FDP device randomly presents a sinusoidal grating in 1 of the 17 test areas located within the central 20 degrees radius of the visual field. The sinusoidal grating has a spatial frequency of 0.25 cycles per degree and undergoes a 25 - Hz counter phase flicker. An advantage of frequency doubling technology perimetry is its shorter time, primarily the results are less affected by blur, pupil size differences and it has lower test-retest variability than white on white perimetry and SWAP.

4. High -Pass Resolution perimetry:

Midget ganglion cells which project to the parvocellular layers of the lateral geniculate nucleus are the most numerous, comprising 70 % of the total of ganglion cells. These cells handle acuity and resolution tasks and prefer stimuli with low temporal frequencies and high spatial frequencies, along with color. High-Pass Resolution perimetry resolution task developed by Lars Frisen, using the Ophthimus High-Pass Resolution perimeter (High Tech Vision, Malmo, Sweden) is a test that is designed to measure the response of parvocellular mechanisms[2]. The test presents spatially filtered rings across 50 test locations in a 30-degree visual field. Fourteen different ring step sizes are used, with threshold designated as the smallest ring size that can be resolved by the patient. This method is comparable with standard fields for detecting vision loss and is superior for identifying change over time [5]. It is user-friendly test, takes approximately 5 minutes and gives feedback for correct responses. Hereby we can see a sample High -Pass resolution perimetry report depicting field defect (Fig).

6. Flicker perimetry:

The ability to detect a rapidly flickering stimulus is believed to be mediated by magnocellular mechanisms. It is considered to be a strategy to detect early glaucomatous damage. Studies by a number of investigators have shown that flicker is able to detect early damage that is not demonstrated by conventional automated perimetry [2]. Flicker perimetry has been implemented in a commercial perimeter, the Medmont M600 (Medmont Pty Ltd., Camberwell, Victoria, Australia). The flicker stimulus has a fixed contrast level, whereas its luminance is varied to obtain threshold. The major advantage is that flicker perimetry is more resistant to optical degradation (Blur, Cataract, etc) than conventional perimetry [6].

7. Motion perimetry:

The ability of detecting motion has been a visual function of interest for detection of glaucoma because of the reports that M-cells and large-diameter fibers may be preferentially damaged early in glaucoma. Evidence from many investigators indicates that this procedure is effective in detecting glaucomatous visual loss and that these deficits precede those found with conventional perimetry[7]. There are several methods of performing motion perimetry. The procedure employed by Fitzke, Johnson and others involves detection of the direction of motion

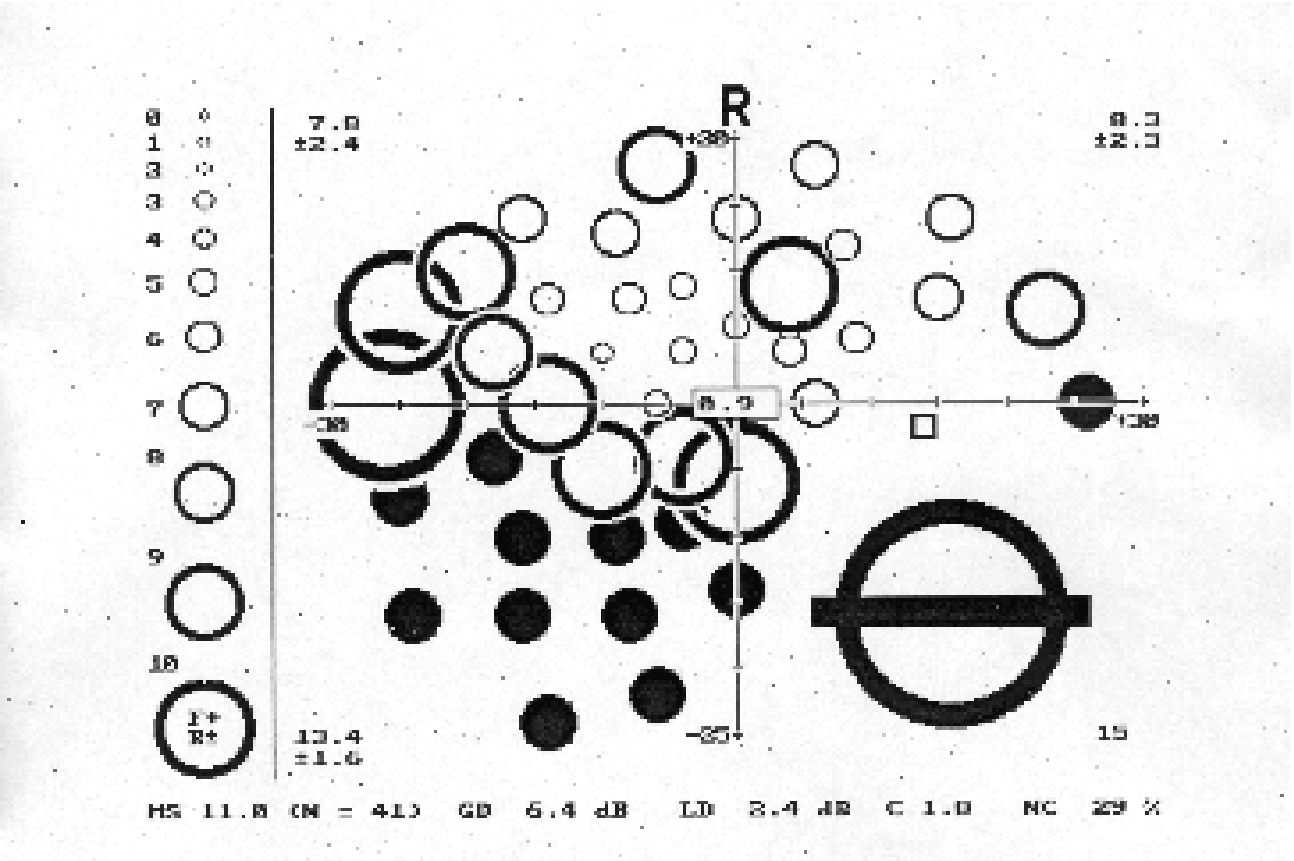

Figure: High-Pass resolution perimetry report showing inferior arcuate defect

of a single dot or line stimulus. This procedure determines the minimum displacement of the stimulus necessary to detect movement. Motion perimetry has several advantages. First, motion is a very salient stimulus for peripheral vision, thereby making this a test that is relatively easy for patients to perform. Second, like flicker perimetry, motion perimetry is highly resistant to optical degradation produced by blur or scattered light from cataract or corneal opacities. Third, large changes in pupil size do not appear to have much effect on motion perimetry thresholds. Finally, motion perimetry is less affected by background luminance and contrast than other visual functions7. However at present time commercial version of this method is not available.

8. Multifocal visual evoked potential:

Multifocal visual evoked potential is an objective measurement of the visual field. There are two Multifocal techniques that are presently being used, the Multifocal electroretinogram, which measures the local electrical responses of the retina throughout the central visual field and the Multifocal visual evoked potential, which measures the localized electrical responses from the primary visual portion of the brain for the central visual field. The usefulness of the multifocal electroretinogram is yet to be studied. Recent investigations of the Multifocal visual evoked potential have indicated that it can provide an objective determination of glaucomatous visual field loss that shows very good correspondence with standard automated perimetry. Multifocal perimetry seems to be a promising new technique for objectively assessing the central visual field and will probably be most useful for patients who are difficult to test with conventional perimetric procedures.

At the present time, these new techniques are able to supplement conventional automated perimetry by providing some additional capabilities. In future, as these techniques are refined, they may possibly become the standard for visual field testing.

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Perimetry Update

R Krishna Kumar

Perimetry is useful clinical tool for the detection of ocular and neurological pathology, differential diagnosis of eye disease and follow-up of the disease. The introduction of automated static perimetry, which occurred some 15 years ago, has changed the way of the management of ocular disease, especially glaucoma. The present perimetric methodology, however, has some drawbacks: like long examination time and inefficiency in detecting early glaucomatous changes. In this article, we discuss few of the developments that address these issues.

1. White-on White automated perimetry:

To reduce the test time, the recent Humphrey field analyzer (HFA-II), has 2 new threshold strategies: SITA standard (Swedish interactive thresholding algorithm) and SITA fast. The SITA standard strategy matches the precision of the full threshold strategy while reducing test time by half[1]. Both SITA and SITA fast use four methods for reducing time: reducing time between presentations, starting the examination of each location with a better estimate of the expected threshold, reducing the testing performed at each test point, and reducing the time spent in catch trials. In addition to blind spot monitoring in HFA-I, HFA-II has gaze-tracking capability that will monitor fixation continuously during the examination.

Tendency Oriented Perimetry (TOP) is an efficient test strategy that has been developed for the Octopus 101 and 300 Series perimeters (InterZeag Ac, Schlieren, Switzerland). It uses a staircase procedure, but does so by sequentially evaluating neighboring locations so that each location is tested only once. The visual field is divided into squares of four neighboring test locations and the first location in each square is tested with the initial stimulus intensity .The stimulus intensity for the second presentation is adjusted according to whether or not the patient responded to the first stimulus. The second location in the square is tested with the new stimulus intensity, a similar procedure is employed for the third and fourth members of the square and a threshold estimate is thus obtained after 4 stimulus presentations. TOP is four time faster than conventional staircase threshold procedures.

2. Blue-on-yellow (B/Y) perimetry or short wavelength automated perimetry (SWAP)

Blue-on-yellow perimetry or short wavelength automated perimetry is used to detect glaucomatous visual field changes much earlier than the conventional white-on-white perimetry does. The blue-on-yellow perimetry is incorporated with the Humphrey Field Analyzer. It uses blue stimuli (440 nm wavelength), narrow band, 1.8-degree target of 200 milliseconds projected on a bright yellow background (530 nm-cutoff-filtered, 315 apostilb) to test selectively the short - wavelength sensitive pathway. The small bistratified ganglion cells mediate the patient response to this test. These cells are few, comprising 6% to 10 % of retinal ganglion cells. The major drawback to this testing method is the test time of 15 minutes. It is also influenced by significant cataract[2].

3. Frequency Doubling Perimetry

Visual information derived from the retinal ganglion cells project to the lateral

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