Coats Disease : A Case Report

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ABSTRACT:

An 08 yr old male child presented to our O.P.D with U/L Leukocoria & Blindness in Left eye for last 15 days. There was a past history of injury to Left eye 1 ½ yrs back but without any consequent visual complaints.

On examination of Left eye the Ant. Segment was normal & on examination of Post.Segment there were vitreous cells with pigments, Bullous Total Retinal detachment with kissing bullae, Sub retinal exudates & a Supero temporal patch of Venous dilatation & hemorrhage. The VA of Left eye was PL-ve PR -ve .The Right eye examination was normal with VA of 6/6. A provisional diagnosis of Exudative Bullous Retinal detachment (LE) was made & a USG- B Scan was done in Left eye. The B Scan ruled out mass lesion & a final diagnosis of COATS DISEASE (LE) was made & the pt is currently under observation.

INTRODUCTION:

Coats disease is a rare idiopathic retinal condition, first described by George Coats in 1908, in which abnormal telangiectatic retinal vessels cause intra- and subretinal exudates and retinal detachment. It typically occurs unilaterally in children, and most commonly in young boys. Males account for approximately 80% of cases. Adults and adolescents may be affected as well. The mean age at diagnosis is 5 years. It may be seen as early as 1 month of age, and most cases are diagnosed before the age of 10 years. The severity of the condition is worse in younger patients (especially those younger than age 3 years), in whom the disease progresses more rapidly. The disease is unilateral in 95% of cases. There is no racial or laterality predilection. The incidence and prevalence are unknown. Coats disease has a sporadic occurrence, and it is not known to be associated with other organ abnormalities, ocular conditions, or systemic conditions. It is known to have a vascular etiology. There is no evidence of genetic predisposition; however, it has been associated with retinitis pigmentosa, muscular dystrophy, deafness, and Norrie disease.

CASE REPORT :

An 08 yr old boy from Phulbani district, presented to our OPD with the chief complaints of Unilateral Leukocoria & blindness in left eye for the last 15 days. The patient was apparently alright 15 days back when the parents of the child noticed a white reflex from the left eye of the boy in the dark & which mimicked the eye of a cat in the darkness. Then the pt was taken to a local hospital & on examination it was found that he is unable to see in the left eye & was referred to our OPD for further evaluation & treatment. His parents do not report any fever or concurrent illnesses. The patient has taken no recent medications (except for multivitamins). There is a past history of trauma to the left eye 1 ½ years back with the head of another child while playing & after which there was immediate peri-ocular swelling & which subsided on taking medications & without any consequent visual complaints. His parents deny any history of allergies, and his vaccination schedule is up to date. The patient was born full term and has one siblings who is a female child of 12yrs & without any similar episode or other ophthalmic disorder. There is no parental consanguinity.

On examination the pt is conscious, well oriented to time , place & person & of average built. The pulse is

80/min regular, B.P 110/70 mm Hg. His oral temperature was 98.6°F (37.0°C).There was no Icterus, Pallor, Edema Or Lymhadenopathy. The Examination of Cardiovascular, Respiratory, Gastrointenstinal , Musculosketal systems were normal.

On examination of the Left eye the VA was PLabsent & PR absent in all four quadrants. Mild exotropia (15 Degrees by Hirschber test) was noticed. There is absence of red reflex with a mid dilated pupil of around 6-7mm & with a relative afferent pupillary defect. Other Ant segment details were normal with a transparent Lens in normal position. On examination of the Posterior segment there was a total bullous retinal detachment with kissing bullae (three in number) & with prominent retinal vessels over the detached retina.The optic disc is not seen due to the kissing bullae.The surface of detached retina was smooth. There were cells in vitreous. Multiple subretinal exudates of greyish to yellowish in colour were present. There was no shifting fluid elicitated on change of posture .There was also a patch of venous dilatation along with hemorrhages seen in the Supero-temporal quadrant of the retina. The IOP of left eye was 20.1 mm Hg with Gonioscopy revealing open angle in all the quadrants,without any abnormal structures.

Visual Acuity in Right eye was 6/6. Ant & Post segment was normal in Right eye with a good red pupilary glow. The IOP of Right eye was 17.3mm Hg with Gonioscopy revealing open angle in all the quadrants.

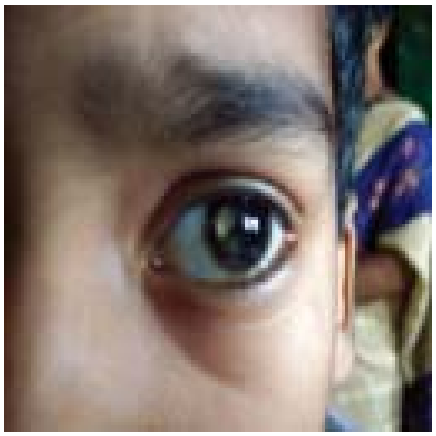
Due to the above findings a Provisional diagnosis of " Total Exudative Bullous retinal detachment" was made. B-scan ultrasonography of left eye revealed no mass lesion with calclflcation which ruled out Retinoblastoma. So a final diagnosis of "Coats disease with Total Bullous retinal detachment" was made.

DISCUSSION:

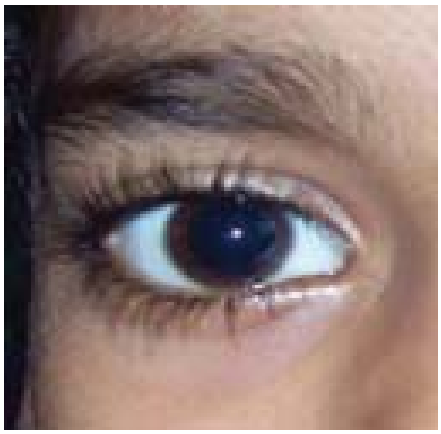
Clinically, Coats disease presents with decreased vision, strabismus, leukocoria (white pupillary reflex or 'cat's eye reflex'), and redness. It may, however, be detected incidentally. It is not generally associated with pain, except for in advanced cases with secondary associated neovascular glaucoma. It has not been associated with infectious or inflammatory conditions, and the great majority of patients have no associated systemic medical problems. The clinical presentation shows great variability. Typically, the ophthalmoscopic examination reveals a localized area of subretinal exudates associated with vascular abnormalities. Vascular changes may include peripheral retinal telangiectasia, capillary and small vessel dilatation and tortuosity, sheathing, capillary nonperfusion, and small aneurysms located at the equator of the eye and the ora serrata (most commonly, inferotemporal). The posterior pole is less frequently affected. Exudation is a common feature that presents in most cases as flat intraretinal and subretinal exudates. These exudates initially appear in areas of telangiectasia and progress to become more widespread. Macular protein accumulation can occur directly from macular telangiectasia or indirectly from peripheral disease. A dense exudate or white nodule in the macula can progress to a disciform lesion that indicates a poor visual prognosis. Retinal hemorrhages may be seen. The vitreous remains clear until the advanced stages. Retinal cysts may be seen and are common in chronic retinal detachments of different etiologies.

The diagnosis may be suspected clinically by indirect ophthalmoscopic evaluation of both eyes, but ancillary testing is extremely helpful to differentiate Coats disease from retinoblastoma, which is a more feared disease. A- and B-scan ultrasonography will show low internal reflectivity consistent with an exudative retinal detachment, and it will rule out the presence of a solid tumor. Intravenous fluorescein angiography helps to visualize the telangiectatic vessels as irregular dilated tortuous vessels filling in the late arterial and early venous phases. Microaneurysms are seen as "light bulb aneurysms". The angiogram will also show progressive leakage from abnormal vessels, adjacent areas of capillary dropout, and late staining of intraretinal exudates. Associated

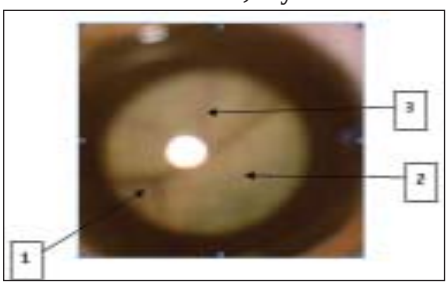
Srimanta Naik, 08yrs HM



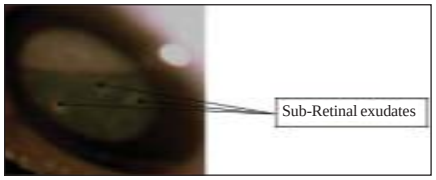
MILD EXOTROPIA (15 Degrees) LE



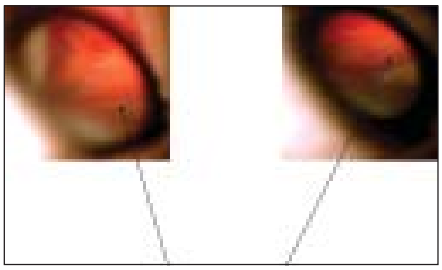
RIGHT EYE (DILATED)



TOTAL BULLOUS RETINAL DETACHMENT WITH 3 KISSING BULLAE



SUB RETINAL EXUDATES WITH PROMINENT RETINAL VESSELS



PATCH OF TELANGIECTACTIC VESSELS WITH HEMORRHAGES IN SUPERO-TEMPORAL QUADRANT

complications of Coats disease include neovascular glaucoma (approximately 10% of patients), angle closure glaucoma, anterior chamber cholesterolosis (3%), retinal/disc neovascularization, vitreous hemorrhage, secondary retinal vasoproliferative tumor, and intraretinal cysts.

Based on the clinical appearance and progression, Coats disease may be classified in 5 grades, as follows

- Grade I: Isolated focal exudates
- Grade II: Massive elevated exudation
- Grade III: Partial retinal detachment
- Grade IV: Total retinal detachment
- Grade V: Secondary complications

A more recent classification system has emerged, which is based on the prognosis. It grades the disease in the following manner

- Grade I: Telangiectasias only
- Grade II: Telangiectasias and exudation
- Grade III: Exudative retinal detachment
- Grade IV: Total retinal detachment with secondary glaucoma
- Grade V: End-stage disease

If untreated, Coats disease shows progressive worsening in most cases, with a variable speed; therefore, early treatment is recommended. The disease does not respond to steroid or antibiotic treatment. Recommended treatments depend mainly on the stage of the disease, and they include observation, laser photocoagulation, cryotherapy, retinal detachment repair with pars plana vitrectomy and/or scleral buckle, and enucleation. Because treatment involves general anesthesia and frequent follow-up, discussions with a patient's parents (if the patient is a minor) regarding diagnosis, prognosis, and treatment goals are of extreme importance. Laser treatment has been used with a high success rate in cases with less exudation. Treatment is directed to areas of vascular leakage and nonperfusion, which decreases or eliminates further exudation and leads to resolution of the exudates and serous detachment. Fluorescein angiography is very useful in guiding laser treatment.

Peripheral lesions are better treated with cryotherapy or a combination of laser and cryotherapy. Treatment may have to be repeated, as recurrences may follow. In more advanced cases, drainage of the subretinal fluid may be necessary, with or without the use of a scleral buckle. Pars plana vitrectomy has been recently used to treat exudative and tractional retinal detachments. Despite the resolution of exudates, subretinal fibrosis and scarring may limit the visual prognosis. Because the disease is unilateral in most cases, patients can live a normal life with the use of polycarbonate glasses for protection of the fellow eye, especially during sports activities.

There are a few new treatments in various stages of clinical trials, but are years away from becoming available. Some doctors are experimenting with a drug called Avastin, Avastin is an FDA approved therapy designed to inhibit angiogenesis, the process by which new blood vessels develop. In cancer patients, this anti-angiogenic process starves the tumor of new blood cells. In Coats' disease, it is believed that Avastin would greatly inhibit the development of new blood vessels. This could potentially eliminate the need for repeated sessions of laser therapy and the scar tissue it causes.

CONCLUSION :

A young child within the age of 10yrs presenting with a unilateral white pupillary reflex should induce the suspicion of Retinoblastoma, or ROP or Coats disease. But in our case the B-scan ruled out any mass with calcification thus ruling out Retinoblastoma & as the child had an uneventful birth history & the retinal detachment was of an exudative type instead of tractional, thus ruling out R.O.P. The age & mode of presentation was very typical for Coats diseae , but the telangiectactic vessels were found in the Supero-Temporal quadrant instead of the usual inferotemporal quadrant. In this patient, enucleation was not preferred treatment because secondary complications like neovascular glaucoma, elevated intraocular pressure, closed anterior chamber angle were absent & there was absence of pain. It was important to rule out the possibility of retinoblastoma by a USG B-Scan, because if left untreated, it would have been fatal . The pt is now currently under observation . Because the disease is unilateral in most cases, patients can live a normal life with the use of polycarbonate glasses for protection of the fellow eye, especially during sports activities.

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