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An Unusual Case of Persistent Primary Hyperplastic Vitreous

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I. Introduction

Persistent hyperplastic primary vitreous (PHPV) is a pathologic entity resulting from abnormal persiste nce of the fetal fibro vascular primitive stroma (hyaloid system) of the eye, [1,2] which should disappear by the ti me of birth. Exact incidence of PHPV is not known but is a rare entity. In our case it is associated with cataracto us lens, hypo plastic iris and increase axial length of the eye ball. Usually PHPV is associated with decreased axi al length.Such an atypical clinical presentation of Persistent Hyperplastic Primary Vitreous has been found unus ual in the available text.

II. Case History

A male child of 12 years of age and having non-consanguineous parents, presented to the out patient de partment of Assam Medical College with white pupillary reflex and diminution of vision of the left eye since bi rth. There was neither any history of local or systemic disease nor any history of operative procedure or trauma to the left eye.The antenatal and postnatal birth history was normal.Immunisation was up to date according to In dian Academy of Paediatrician guidelines.There was no family history of any congenital disease.

Systemic examination was within normal limits. On local examination, his facial features, ocular adene xa including bony orbital rims and soft tissues were found normal. Visual acuity was perception of light (PL) ne gative. IOP was 32.7 mmHg in left eye. Left eye was exo deviated and the cornea had mild stromal haze with no rmal corneal diameter. Anterior chamber was deep with hypo plastic iris all around.The lens was cataractous an d calcified with some fibrous adhesions extending from the calcified lens till root of the hypoplatic iris(Figure 1, 2, 3). The posterior segment couldnot be visualsed except some large vitroeus cysts seen with fibrous adhsio ns(Figure 3). Angle details were not correctly evaluated because of the corneal haze and fibrous bands. Right ey e was normal with visual acuity of 20/20.

On Investigation, Ultrasonography with color doppler showed presence of persistent hyperplastic prima ry vitreous with flow of blood in both artery and vein.Large vitreous cysts were found along with the adhsions.T he cysts were homogenous. Interestingly the axial length of that eye was increased. CT scan and MRI confirme d radiological diagnosis of a thin, triangular band of enhancing soft tissue extending from the lens through the v itreous body up to optic nerve.Left eye axial length was 27.7mm (Figure 5, 6, 7.). The lens was abnormally thic kened and globular. Chest X-ray was normal. Routine blood examination including ESR was normal. Ultrasoun d abdomen showed normal study. Serological tests of toxoplasmosis were found to be negative.

III. Discussion

Leukocoria in children requires imaging to establish the diagnosis and for further treatment. Retinoblast oma is the most common intraocular malignancy in children presenting with the symptom. In leukocoria,causes like Coat's disease, persistent hyperplastic primary vitreous,congenital cataract etc must be included in the diffe rential diagnosis before embarking on aggressive therapy for ocular tumors. [2, 3]

In the fetus, the primitive lens and vitreous receive their blood supply mainly via the hyaloid system.Th e anterior portion of this arterial system involutes at 8 months of life and is usually present in premature infants. The posterior portion of the arterial system normally regresses by 7 months of life, but is also occasionally prese nt in premature infants. In the absence of hypertrophy, the primitive blood vessels of the hyaloid system regress completely. The anterior and posterior hyaloid vascular systems may persist independently or together. Most pat ients with PHPV have a combination of both anterior and posterior types.

In posterior PHPV, findings include vitreous membranes, a stalk extending from the optic nerve to the posterior lens (a remnant of Cloquet's canal that carries the hyaloid artery), dysplasia of the optic disk, an indisti nct macula that may be hypo pigmented, retinal folds, and a clear lens. In anterior PHPV, findings include a shal low anterior chamber, elongated ciliary processes, enlarged iris vessels, infantile or juvenile-onset glaucoma, cat aract, intralenticular haemorrhage, and a retrolental fibro vascular membrane. A persistent hyaloid artery may al so be present in some cases of anterior PHPV. A and B scan ultrasonography may assist in precise measurement and accurate diagnosis of PHPV. [5]

In patients with antero-posterior PHPV in which vision is unsalvageable, a lensectomy may be done. In posterior PHPV, when rehabilitation of the eye is deemed possible, a vitrectomy may be performed. In cases of purely anterior PHPV, a lensectomy-membranectomy and anterior vitrectomy are done. Prognosis depends on th e type of PHPV. Treatment of posterior and combined antero-posterior PHPV has a less favourable outcome, wi th most patients attaining perception only of motion or light.The surgical line of action in this case is highly deb atable.

Persistent hyperplastic primary vitreous is usually associated with other anomalies like micro-ophthalm os, small lens diameter, polar cataract, glaucoma and corneal scarring. In our case, persistent hyperplastic primar y vitreous was associated with increased axial length of the eye ball, cataractous lens and a hypo plastic iris for 3 60 degrees around,vitreous cysts and with normal corneal diameters.In this case increased axial length is an unu sual findings as PHPV is usually associated with microphthalmos.

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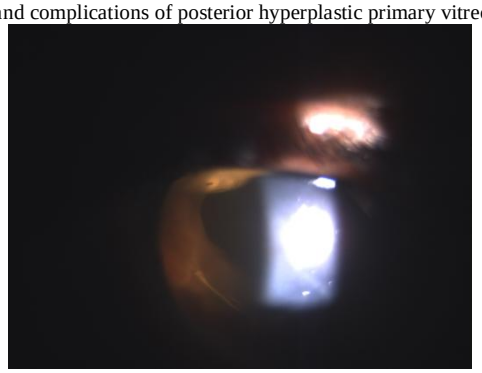

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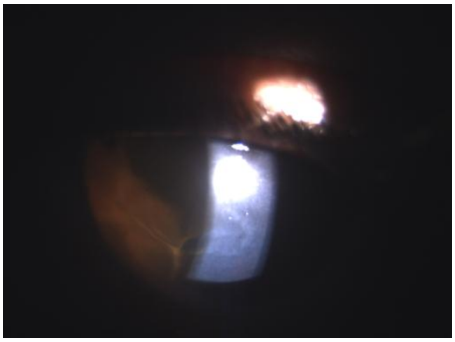

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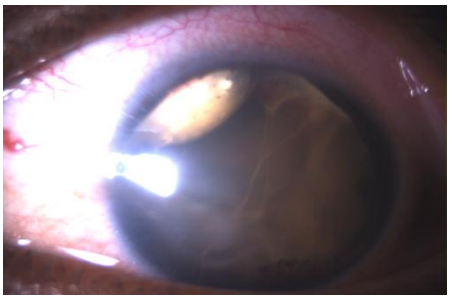

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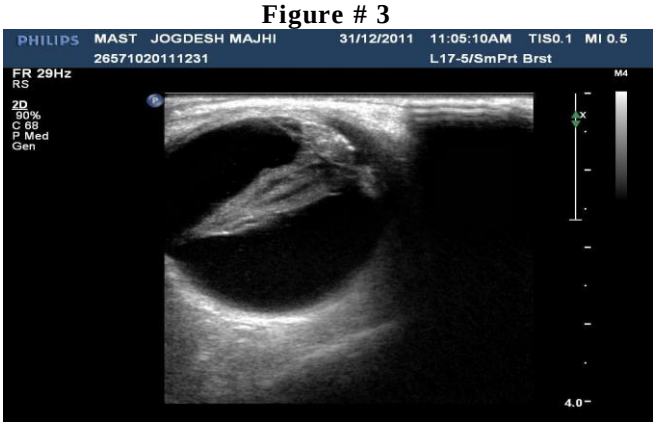

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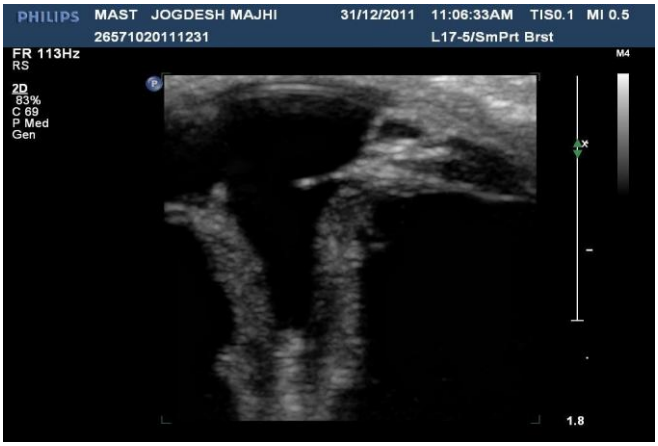

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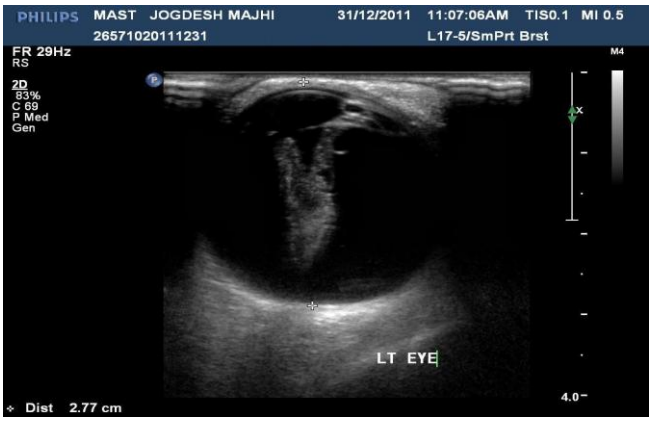

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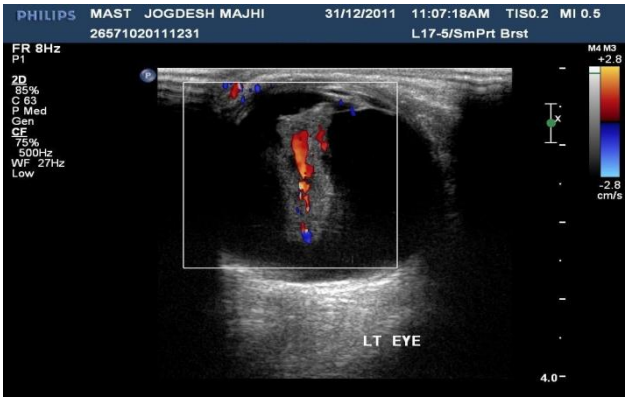

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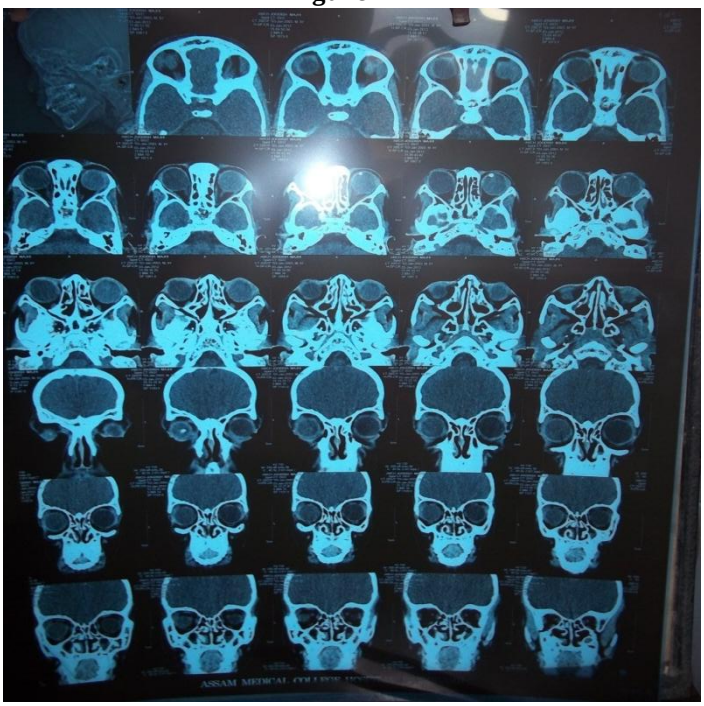

Figure # 8

IV. Conclusion

Persistent hyperplastic primary vitreous is an important cause of leukocoria. Many a times it associated with multiple disorders of eye. In our case it was associated with cataractous lens, hypo plastic iris,vitreous cysts and increase axial length of the eye ball (27.7mm). Also in our case the hyaloid arterial system was still persiste nt with clear blood flow in colour Doppler imaging. We are reporting this case because of such an atypical clinic al presentation of Persistent Hyperplastic Primary Vitreous.