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Case Report

A Case of Unilateral Pseudoglioma of Eye in A Neonate

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Abstract: Pseudoglioma is a condition presenting as leukocoria other than retinoblastoma. Persistent hyperplastic primary vitreous is a type of pseudoglioma in an ocular developmental defect in which the hyaloid vasculature within the vitreous overgrows and fails to regress.In this case , 25 days old female child presented with the complaints of white pupillary reflex in the right eye since birth. Baby was born to second degree consanguineous parents. The child was born at full-term by normal delivery. The Antenatal period was uneventful. No history of oxygenation and birth trauma. Anterior segment examination of the right eye showed a shallow anterior chamber and leukocoria. Lens was cataractous and vascular. The left eye was normal. Examination under anesthesia revealed normal corneal diameter and Intraocular pressure in both the eyes. Fundus examination was normal in the left eye and cannot be performed in the right eye. Ultrasound B-scan in the right eye showed an echogenic band in the posterior surface of the lens and on colordoppler arterial blood flow was seen in the band. Hence, the patient was diagnosed to have Anterior persistent hyperplastic primary vitreous in the righteye.In this case ,child is advised to undergo Right lensectomy with cautery of the persistent anterior hyaloid stalk and anterior vitrectomy.

Keywords: Pseudoglioma, Retinoblastoma , Pupillary Strands, Persistent Hyperplastic Primary Vitreous, Leukocoria, Vitreous humor,Vitrectomy , lensectomy

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1. INTRODUCTION

Pseudoglioma usually presents as leukocoria other than retinoblastoma.[1] Persistent hyperplastic primary vitreous (PHPV) usually presents as sporadic, unilateral or rarely bilateral leukocoria. PHPV is a congenital developmental anomaly of the eye resulting from failure of the hyaloid vasculature and embryological primary vitreous to regress at birth. Persistent hyperplastic primary vitreous was first described by Reese in his Jackson Memorial Lecture in 1955. Goldberg in 1997 renamed PHPV as persistent fetal vasculature (PFV). Mutation in NDP gene has been identified in some cases and incidence was 1 in 15000 live births.[2] The spectrum of disease ranges from pupillary strands, Mittendorf's dot to dense retrolental membrane and Retinal Detachment. It is Associated with other congenital defects and neurological abnormalities. PFV may be associated with rare systemic syndromes like Walker-Warburg anencephaly, Patau syndrome, Trisomy 13, Oculopalatal-cerebral dwarfism, Fetal alcohol syndrome & Oculo-dento-osseous dwarfism. The majority of PFV cases occur sporadically.[3] A few cases of familial occurrences have been reported in dizygotic twins, in two brothers. Persistent fetal vasculature isdivided into Anterior, Posterior & Combined PFV. Autosomal dominant and autosomal recessive genetic forms of PFV have been described. Anterior PHPV occurs when the remnant vascular stalk is seen attached to the back of the lens but no longer extends back to the optic nerve.[4]The purely anterior PHPV is also known as persistent tunica vasculosalentis. Ocular findings in Anterior PFV are Leukocoria, Microphthalmia, Cataracts, Elongated ciliary processes, Shallow anterior chamber depth, Glaucoma,Retrolentalfibrovascularmembrane,Strabismus&Int ralenticular hemorrhage.[5]Most of Posterior PFV are sporadic and 83% unilateral. Common age of presentation is 6 years.Ocular findings in Posterior PFV are Leukocoria, Microphthalmia, usually clear lens, Retinal folds, Normal anterior chamber depth, Pigment maculopathy, Vitreous membrane and stalk, Strabismus, Macular hypopigmentation and hypoplastic optic nerve. 50% of patients can have micro cornea and immature angles. In posterior PHPV the remnant vascular stalk is seen arising from the optic nerve but not reaching the lens, thus usually not causing cataract. Posterior PFV can present as Bergmeister's papilla, as a vascular loop arising at the disc or as a glial mass overlying the inferior optic nerve head and extending into the vitreous causing peripapillary tractional retinal elevation in turn leads to phthisis bulbi.The purely posterior presentation of PHPV is termed falciform retinal septum and ablation falciformiscongenita. [6,7]The diagnosis of PFV is usually obvious clinically based on comprehensive eye examination. This can be confirmed by B mode ultrasonography with color doppler, computing tomography (CT) or magnetic resonance imaging (MRI).[8] A cone-shaped retrolental density is a characteristic finding of PFV on imaging studies. Imaging is useful in assessing the size, thickness, and vascularity of the retrolental fibrovascular mass. CT also enables the best assessment of the presence or absence of calcifications to help distinguish PHPV from retinoblastoma, which is frequently calcified. The goals in treatment are saving the eye from the complications of cataract, glaucoma and retinal detachment.Useful vision, and acceptable cosmetic outcome can be achieved by early intervention. Lensectomy with primary posterior capsulorhexis / with or without anterior or total vitrectomy and membrane excision may prevent further complications.Visual rehabilitation by aphakic contact lens and amblyopia therapy is possible with eyes that are fairly normal in structure after the lensectomy and membranectomy.[9]

2. CASE REPORT

A 25 days old female baby weighing 2.5 kg at birth delivered by elective caesarean section was brought to the Paediatric outpatient department with a whitish pupil in the right eye since birth as noticed by the parents (Figure 1). Baby is a precious child born to second degree consanguineous parents after 7 years of marital life. Mother had one abortion at 3rd gestational month previously. TORCH test was positive and she was treated for that before this conception. She conceived within 8 months of vaccination. Pregnancy was confirmed by UPT after 45 days of amenorrhoea. First and second trimester was uneventful. She was diagnosed to be oligohydramnios by USG at third trimester and planned for elective caesarean section**.** Baby cried immediately after birth. Breastfeeding started immediately after birth. baby sucking and swallowing well . No neonatal intensive care unit admission. Written consent was obtained from the parents for the publication of the study and consent for the publication from the institution was also taken.

3. MEDICAL HISTORY

No history of any birth trauma and usage of hyperbaric oxygen.no significant medical history.

4. FAMILY HISTORY

No significant family history.

5. OBSERVATION

Baby sucking and feeding well ,passing urine and stools normally.On General examination Child is alert, active , afebrile, hydration is good,All peripheral pulses felt, CRT <3sec .No pallor, no icterus HEAD TO FOOT EXAMINATION- Eyes-whitish pupil in the right eye, No neuro cutaneous markers .On systemic examination Cardiovascular system first heart sound and second heart sound present, no murmur.Respiratory system -Bilateral air entry present, no added sound.per Abdomen soft ,non tender ,bowel sound present,no significant organomegaly .central nervous system- Anterior fontanel at level ,Tone normal.

6. INVESTIGATIONS

Thorough anterior segment examination was done. Ophthalmic Examination revealed normal corneal diameter and intraocular pressure in both the eyes.

Table 1: ANTERIOR SEGMENT EXAMINATION		
STRUCTURES	**RIGHT EYE**	**LEFT EYE**
Lids, Conjunctiva, Cornea	Normal	Normal
Anterior chamber	Shallow	Depth & Content Normal
Iris	Brown, Radial pattern	Brown, Radial pattern
Pupil	2.5 mm, Round, Reacting to light,LEUKOCORIA +	2.5 mm, Round, Reacting to light
lens	**Cataractous and Vascular** (Figure 2)	Clear
Fundus	No view	Normal

Table 1- Illustrate that the Examination of anterior segment of Right eye-Lids ,conjunctiva ,cornea is normal. Anterior chamber of the right eye is shallow . Right eye pupil shows leukocoria but 2.5mm in size , roundin shape ,Reacting to light , Right side lens shows cataractous and vascular .There is no view in fundus when compare to left eye .Left eye is normal –lens is clear ,pupil is round in shape , 2.5mm in size ,reacting to light .Iris is brown in colour , typical radial pattern .Anterior segment of left eye is depth and content normal, lids ,conjunctiva ,cornea is normal ,Fundus of the left eye is normal

Fig 1 (Patient image)

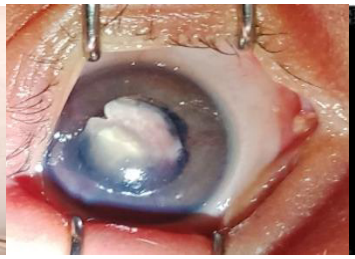

Fig 2 (Cataractous and vascular lens)

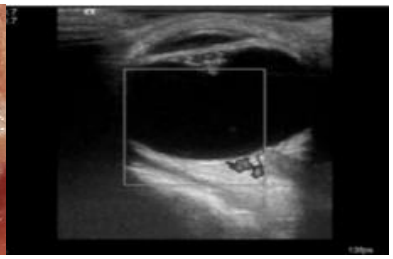

Fig 3 (Band in posterior surface of lens in B scan)

7. SPECIAL TEST

B SCAN Showed a band in the posterior surface of the lens (Figure 3) in the Right eye & Normal study in the left eye
CT IMAGING showed Cone shaped retrolental density & absence of calcification exclude RETINOBLASTOMA

8. DIAGNOSIS

Baby was diagnosed as a case of Anterior persistentfetal vasculature (PFV) of Right eye based on the above investigations.

9. DISCUSSION

Vitreous humour is an inert, transparent, colourless, jelly like, hydrophilic gel that serves the optical functions and also acts as important supporting structures for the eyeball. The vitreous cavity is bounded anteriorly by the lens and ciliary body and posteriorly by the retina. It weighs nearly 4g. Vitreous has strong attachment at 4 regions, which are oraserrata, optic disc, macula & back of the lens.Vitreous is divided into three parts , which are hyaloid layer or membrane, cortical vitreous and medullary vitreous. Primary or primitive vitreous is mesenchymal in origin and is a vascular structure having the hyaloid system of vessels. Secondary or vitreous proper is secreted by neuroectoderm of optic cup and it is an avascular structure. When the vitreous fills the cavity, primitive vitreous with hyaloid vessels is pushed anteriorly and ultimately disappears. Tertiary vitreous is developed from neuroectoderm in the ciliary region. [10]Our case is a type of Anterior PFV, which is described as when the remnant vascular stalk of hyaloid vasculature is seen attached to the back of the lens but does not extend back to the optic nerve. Anterior PHPV is also known as persistent tunica vasculosa lentis.[11]Anterior PFV consists of funnel shaped retrolental fibrous tissue and the Stem of the funnel contains remnants of hyaloid artery. This retrolental fibrous tissue attaches to elongated ciliary processes and draws them centrally causing cataract formation, shallow Anterior chamber and angle closure Glaucoma. Insufficiency of vitreous endostatin plays an important role in pathogenesis. Globe can be normal or microphthalmic.[12,13] In the late stage, it can even be larger due to pupillary block Glaucoma. Anterior PFV is typically associated with cataract, glaucoma and retrolenticular membrane. Iris vessel engorgement and recurrent intra ocular haemorrhage can result in tractional retinal detachment leading to phthisis bulbi. Congenital anomalies of optic disc and macula are often seen.[14]Differential diagnosis includes retinoblastoma, congenital cataract, Retinopathy of prematurity, Coat's disease, vitreoretinal dysplasia, ocular toxocariasis. lensectomy is performed in Anterior PFV presenting with cataract. Endodiathermy of the fetal vasculature can be performed to prevent the hyphema occurring intraoperatively. In case of thick plaques, it can be cut using Vitrectomy or micr-scissors. Glaucoma is the major cause of blindness in Anterior PFV which is treated with Antiglaucoma drugs, Trabeculectomy and glaucoma drainage devices. Anterior PFV with Glaucoma has a better prognosis when intervened at earliest.[15,16]The important factor for good outcome is whether the posterior pole is involved or not. There is a wide range of treatment options and potential outcomes exists due to its wide spectrum of presentation. [17,18]Treatment plans including early surgery may result in relatively good visual outcome in selected patients. The optimal age for surgery is 4-6 weeks of age.[19]Patient was advised to undergo right lensectomy with cautery of the persistent anterior hyaloid stalk and anterior vitrectomy via pars plana approach. [20,21]Regular follow up is essential in these patients as the disease is often progressive. Visual rehabilitation by replacement of the lens function typically requires use of an aphakic lens postoperatively and patching therapy to minimise amblyopia is the norm.[22,23]The treatment priority is mainly saving the eye from complications of cataract, glaucoma and retinal

detachment, thereby the retainment of vision and acceptable cosmetic outcome can be achieved.[24,25]

10. PROGNOSIS AND FOLLOW UP

Prognosis depends on the type of the disease, whether the posterior segment is involved or not , time of intervention, response and compliance to the treatment. Patients with Anterior PFV have a good prognosis when intervened at the earliest. Regular followup is very essential & crucial in these patients to preserve the visual potential as the disease is often progressive.[25,26]

11. CONCLUSION

Selecting the patient appropriately for the surgical management plays an important role . In patients with severe optic nerve involvement / foveal hypoplasia, surgery should be avoided. Refractive correction & amblyopia treatment is the most important factor in visual rehabilitation. Unfortunately compliance with the treatment in babies is extremely difficult. Persistent fetal vasculature (PFV) requires meticulous care, early treatment and regular follow ups . It needs a multidisciplinary approach involving pediatricians, pediatricophthalmologists , vitreoretinal surgeons and contact lens specialists .

12. AUTHOR'S CONTRIBUTION STATEMENT

Dr.Hamsa v s conceptualized and gathered the data with regard to this work. Dr.Jagadeeshwari and Dr.Naveenchandher analyzed these data and necessary inputs were given towards the designing of the manuscript. All authors discussed the methodology and results and contributed to the final manuscript.

13. CONFLICT OF INTEREST

Authors declare no conflict of interest.

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