Rare Case of Primary Congenital Glaucoma with Hypoplasia Corpus Callosum

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CASE REPORT

RARE CASE OF PRIMARY CONGENITAL GLAUCOMA WITH HYPOPLASIA CORPUS CALLOSUM

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Primary congenital glaucoma is a rare disease that causes elevated intraocular pressure within the first three years of life. Few studies have explored the association of primary congenital glaucoma with malformation of corpus callosum. We report on a six-month-old female presenting with unilateral primary congenital glaucoma associated with hypoplasia of corpus callosum in Indonesian infant. The patient had already undergone trabeculectomy surgery. However, there no obvious improvement following the procedure given the severity of the condition. **Conclusion**: The failure rate of surgery in severe primary congenital glaucoma conditions is still very high, and therapy can usually preserve vision if early identification of mild or moderate form is made.

Keywords: Primary congenital glaucoma; Hypoplasia of corpus callosum.

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INTRODUCTION

Congenital glaucoma is defined as maldevelopment of some portions of the eye, which may or may not be associated with a systemic anomaly, and which causes elevated intraocular pressure (IOP) within the first three years of life.[1] However, when glaucoma develops in the absence of any secondary cause or related systemic problem, the term primary congenital glaucoma (PCG) is applied.[2,3] The incidence of PCG is relatively rare, with overall prevalence at 1:10,000 births. Most of the PCG is expressed bilaterally, and asymmetric expression should be suspected in clinically apparent unilateral cases.[2]

Hypoplasia of the corpus callosum (HCC) exists when all the components of the callosum are present, but the structure is thinner than normal.[4] Some recent studies have reported some syndrome-associated congenital glaucoma with malformation of corpus callosum.[5] The objective of this paper is to report a rare case of congenital glaucoma in an Indonesian infant with hypoplasia corpus callosum.

CASE REPORT

A six-month-old female was referred to the Paediatrics and Ophthalmology Department Soetomo Hospital, Surabaya, Indonesia due to protuding left eyeball. There was a bulge in the centre of the left eye since a week before admission (Figure 1–4). Patient was irritable and presenting photophobia as well as blepharospasm. Tears were coming out of her left eye. There was a previous history of trabeculectomy procedure in the left eye at the age of 40 days. The baby was born spontaneously with birth weight 1,000 grams at 28–29 weeks of gestational age. The Apgar score was 7–8, meconeal amniotic fluid was present, but no history of birth trauma. The mother was in healthy condition with good antenatal care. The USG anatomy of vitreoretina was in the normal limit (Figure-5). Examination under anaesthesia is presented in table-1.

The head MRI showed hypoplasia of the corpus callosum (Figure-6, 7). The patient was diagnosed as PCG with descemetocele on left eye and seclutio of the pupil, with anterior synechia on the right eye. Enucleation or evisceration therapy on the left eye and pupiloplasty for the right eye were considered in order to let the light through the pupil into the retina, and allow the patient to learn to see with her right eye. Unfortunately, the patient's parent refused the procedure.

The patient was six months of age; otherwise, it was three months of corrected age. Evaluation of growth and development using the Denver II chart showed delay in the aspects of fine motor adaptive and social personal.

Table-1: Examination under anaesthesia

Right eye		Left eye
Light perception +	Visual acuity	Decreased of light perception
Normal	IOP	36 mmHg
Hyperaemia -	Conjuctiva	Hyperaemia -
Clear with Φ 10 mm	Cornea	Cloudy with Φ 16 mm, descemetocele +
Deep	Anterior Chamber	Flat
Anterior synechiae	Iris	Anterior synechiae
Seclutio	Pupil	Seclutio
Difficult to evaluate	Lens	Difficult to evaluate
Fundus reflex + Normal cup and disc ratio	Fundoscopy	Fundus reflex + Difficult to evaluate cup and disc ratio

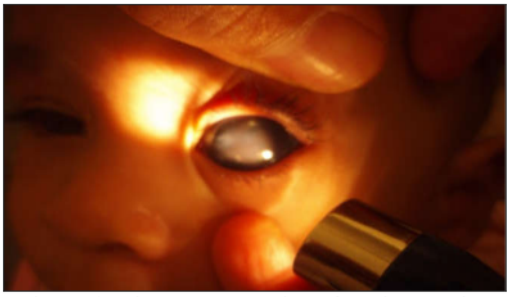

Figure-1: Right eye: seclusion of pupil, anterior synechiae, and normal cornea

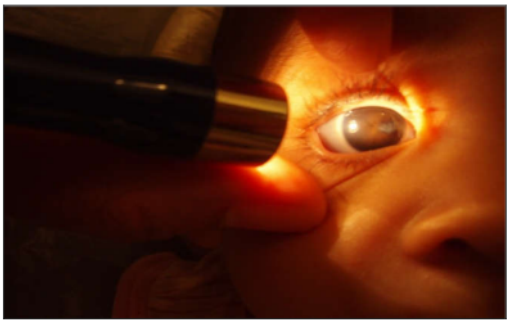

Figure-2: Left eye: seclusion of pupil, anterior synechiae, enlargement of a cloudy cornea and descemetocele

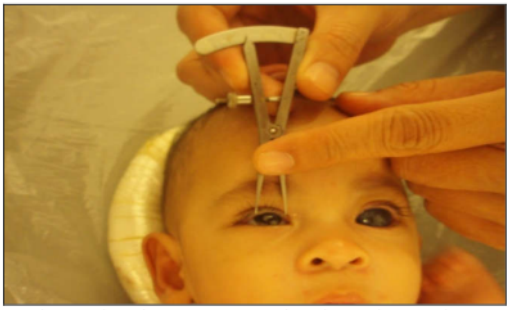

Figure-3: Rigth eye: examination with calliper revealed corneal diameter was 9 mm

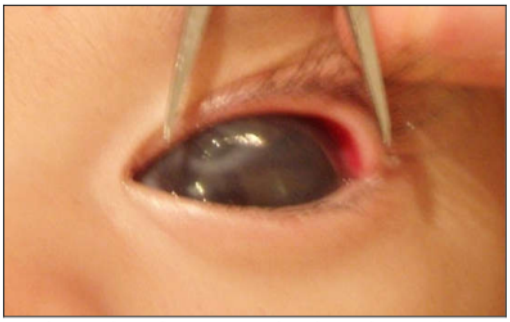

Figure-4: Left eye: examination with calliper revealed corneal diameter was 24 mm

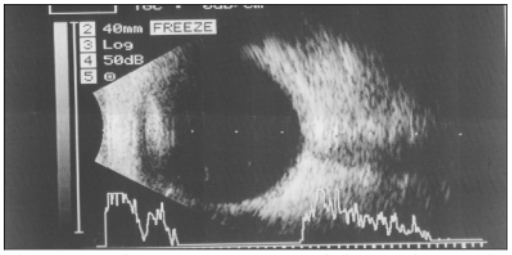

Figure-5: Left eye: The anatomy of vitreoretina was in the normal limit and axial length was 23 mm

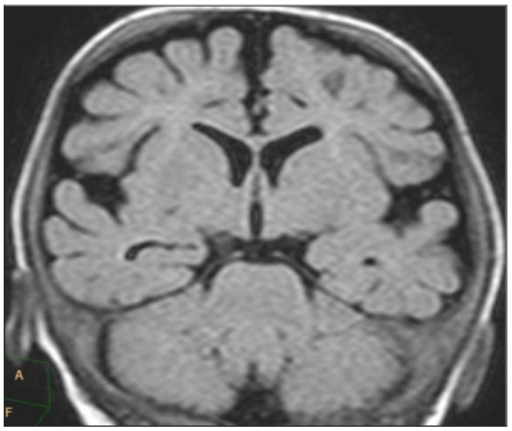

Figure-6: The Head MRI coronal view showing hypoplasia of corpus callosum

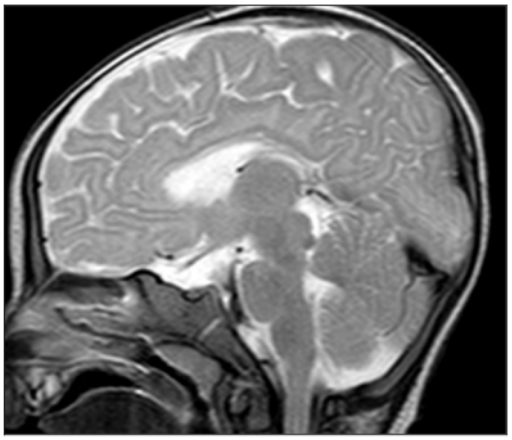

Figure-7: The Head MRI sagittal view showing fypoplasia of corpus callosum

DISCUSSION

The manifestations of glaucoma in infants are different from those associated with open angle glaucoma in adults. These differences provide the key to early diagnosis. Photophobia, blepharospasm, and epiphora frequently result from the corneal changes that occur secondary to elevated intraocular pressure (IOP) in the

infant eye.[6] When the condition reaches this stage, the eye is painful and tearing is present. In our case, the baby became sensitive to light and attempted to avoid it by covering the eye or burying the head in a blanket.

The size of the cornea is important in the diagnosis of PCG. Al-Umran KU found that the corneal diameter on preterm infant ranged from 7–7.5 mm to a maximum of 10 mm. The walls of neonatal eyes are distended by an elevated IOP because the corneal and scleral collagen has not yet sufficiently hardened. Before the age of six months, the rigidity of the cornea is less than the sclera; consequently, the cornea enlarges more rapidly under the influence of an increased IOP. The final diagnosis of congenital glaucoma can usually be made only when infant is under general anaesthesia (EUA) by evaluating (1) the IOP with Schiotz, Perkins or Tono-Pen tonometer, (2) length of the horizontal cornea with calliper, (3) the angle of anterior chamber with gonioscopy, and (4) the fundi (1,2).[1,2] In this patient, the enlargement of cornea was accompanied by the symptoms of tearing and blepharospasm. The EUA revealed left cornea was larger than normal value. The left cornea was so cloudy that it caused some difficulties in evaluating the Hab's striae and fundi. The cloudy cornea with corneal enlargement could be seen in the first week of life due to increasing of IOP, which reached 36 mmHg (normal IOP was less than 25 mmHg under anaesthesia). The left AXL was longer than normal value because immature collagen remained distensible due to elevated IOP, leading to eye enlargement. Paediatric glaucoma is classified into PCG when caused by congenital anomalies that affect aqueous outflow, and secondary glaucoma when caused by post-birth factors such as ocular trauma, surgery, and inflammation.[3] Diagnosis of PCG in this case is made by exclusion of other causes of glaucoma. PCG is better treated with surgery than medication because of the pathophysiology of the disease.[2,7] Surgical treatment includes goniotomy, trabeculotomy, or trabeculectomy, or a combination of these procedures.[7] The surgery planned is usually trabeculo-trabeculectomy, due to the severity of the glaucoma, but in practice only trabeculectomy is performed, due to technical constraints in finding Schlemm's canal. This is consistent with research by Dietlein *et al.* who performed trabeculectomy in four patients who initially planned trabeculo-trabeculectomy because Schlemm's canal could not be localized.[8] The trabeculectomy that had already been performed in this patient resulted no significant improvement. There is more failure with trabeculectomy than goniotomy or trabeculotomy because of complications including scarring over the bleb.[1,8,9] Corpus callosum is the largest connective structure in the brain. HCC exists when all the components of the callosum are present, but the structure is thinner than normal.[10] HCC is a strong predictor of developmental delay and significantly associated with poor outcome in all of the developmental domains. Anderson *et al.* found that among very premature infants, corpus callosum grows at lower rate postnatal than in utero. Some recent studies have reported some syndrome-associated congenital glaucoma with malformation of corpus callosum.[5] This patient suffered from HCC with developmental delay, but the other congenital anomaly associated with a syndrome has not been proven. In conclusion, PCG associated with HCC is a rare disease and presents challenging management. Recognizing the early presenting signs and symptoms of congenital glaucoma for medical practitioners is important. The failure rate of surgery in severe PCG conditions is still very high, and therapy can usually preserve vision if early identification of mild or moderate form is made.

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