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CLINICAL

A CASE REPORT OF CONGENITAL CORNEAL OPACITY

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ABSTRACT

INTRODUCTION: Congenital corneal opacity refers to the opacification of cornea present at birth. It can occur as an isolated anomaly or with systemic association. It is a rare condition and is an important cause of corneal blindness. Prevalence of congenital corneal opacities is estimated to be around 3 in 100,000 newborns. Sensory visual deprivation due to opacification of cornea in the phase of visual development leads to sensory deprivation amblyopia. Early diagnosis and intervention is essential. **CASE DETAILS:** A 10 days old male child is brought by his parents to the ophthalmology OPD with complaints of whitish discolouration of cornea in right eye noted since birth. Baby was full term with birth weight 2.4 kg. There was h/o maternal infection during pregnancy. On ocular examination, corneal clouding of right eye was present. B scan USG of both eyes was normal. On systemic examination, hepatosplenomegaly and pan systolic murmur suggestive of VSD were present. Hearing evaluation detected sensory neural hearing loss. TORCH titre came out to be positive for rubella and CMV. This is a rare case of unilateral congenital corneal opacity secondary to congenital rubella and CMV infection. **CONCLUSION:** Congenital Corneal opacities, though rare, pose a significant morbidity to a child's visual function. Various genetic, developmental, infectious and metabolic conditions cause congenital corneal opacities. Early diagnosis of underlying aetiology and surgical intervention is essential to prevent sensory deprivation amblyopia. A detailed evaluation and timely intervention decrease the morbidity and mortality.

KEYWORDS

congenital corneal opacity, congenital rubella infection, congenital cytomegalovirus infection

A CASE REPORT OF CONGENITAL CORNEAL OPACITY

INTRODUCTION: Congenital corneal opacity refers to the opacification of cornea present at birth. It can occur as an isolated anomaly or with systemic association. It is a rare condition and is an important cause of corneal blindness. Prevalence of congenital corneal opacities is estimated to be around 3 in 100,000 newborns. Sensory visual deprivation due to opacification of cornea in the phase of visual development leads to sensory

deprivation amblyopia. Management of congenial corneal opacity consists of early diagnosis and treatment. Penetrating keratoplasty is indicated for the corneal opacity.

CASE DETAILS: A 10 days old male child is brought by his parents to the ophthalmology OPD with complaints of whitish discolouration of cornea in right eye noted since birth.

Birth History: Second born male child, Full term normal

vaginal delivery, delivery was uneventful, baby cried immediately after birth, no h/o NICU admission. Birth weight was 2.4 kg, h/o maternal infection during pregnancy.

Family history: H/o death of first born child due to cardiac anomaly at the age of 1 year. H/o polyhydramnios during 1st pregnancy.

Ocular Examination:



Clinical photo of right eye congenital corneal opacity

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RS: AEBE

PA: Hepato-splenomegaly

- Paediatric referral for further evaluation

INVESTIGATIONS:

-TORCH TITRE

Rubella IgG titre: positive (85.2 IU/mL); Rubella IgM: negative (10 AU/ml)

CMV IgG titre: positive (156 U/mL); CMV IgM: negative (5 U/ml)

-CBC: WBC count: 10,000/cumm; RBC count: 4.69 milli/cumm;

platelets: 3 lakhs/cumm; Hb: 16.8 g/dl

-LFT: Direct bilirubin: 0.36 mg/dl; Indirect bilirubin: 7.54 mg/dl;

-USG Cranium: Normal

-USG Abdomen: hepatosplenomegaly

-Hearing evaluation: SNHL

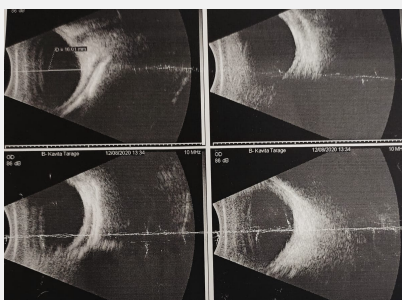
-2D-ECHO: VSD

DIAGNOSIS

Right Eye Congenital Corneal Opacity secondary to Congenital Rubella and Cytomegalovirus infection

PLAN OF MANAGEMENT:

Penetrating Keratoplasty



B scan of both eyes

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SYSTEMIC EXAMINATION

CNS: NAD

CVS: Pan-systolic murmur

DISCUSSION:

Congenital corneal opacity refers to the opacification of cornea present at birth. It can occur as an isolated anomaly or with systemic association. It is a rare condition and is an important cause of corneal blindness. Prevalence of congenital corneal opacities is estimated to be around 3 in 100,000 newborns. (1) Unilateral corneal opacities are generally isolated, while bilateral opacities are often associated with systemic conditions and a complete genetic workup is essential. (2) Sensory visual deprivation due to opacification of cornea in the phase of visual development leads to sensory deprivation amblyopia.

Differential diagnosis of congenital corneal opacities :(3)

-Corneal dystrophies: - Congenital hereditary endothelial dystrophy

- Congenital hereditary stromal dystrophy

- Posterior polymorphous corneal dystrophy

-Peter's anomaly

-Infections: - congenital rubella, herpes simplex, cytomegalovirus and bacterial

- Anterior segment dysgenesis: sclerocornea, anterior staphyloma

-Dermoid

-Trauma: forceps delivery trauma

-Metabolic disorders: mucopolysaccharidoses, cystinosis

-Congenital glaucoma

-Developmental anomaly of iridotrabeular system: Axenfeld-Reiger syndrome

Management of congenial corneal opacity consists of

-Early diagnosis and treatment. Penetrating keratoplasty is indicated for the corneal opacity.

-Treatment of concurrent ophthalmic and systemic conditions.

-Prevention of sensory deprivation amblyopia and visual impairment.

Treatment of congenital corneal opacity is often challenging and complicated because of reduced scleral rigidity and increased elasticity. This increases the risk of anterior displacement of lens-iris diaphragm and wound retraction with associated lens expulsion.

Early diagnosis of underlying aetiology and surgical intervention is essential to prevent sensory deprivation amblyopia. (4, 5)

CONCLUSION: Congenital Corneal opacities, though rare, pose a significant morbidity to a child's visual function. Various genetic, developmental, infectious and metabolic conditions cause congenital corneal opacities. Early diagnosis of underlying aetiology and surgical intervention is essential to prevent sensory deprivation amblyopia. A detailed evaluation and timely intervention decrease the morbidity and mortality.

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