A rare case of cleft number nine associated with atypical cleft number two

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The incidence of the craniofacial cleft is rare ranging between 1.43 and 4.85/100,000 births. Tessier number nine cleft being the rarest, there are a few reports of detailed ophthalmologic examinations performed in them. In this study, 1-day-old female neonate delivered by normal vaginal delivery at term, weighing 1480 g presented with right eye dystopia, cleft extending through the lateral third of the upper eyelid, brow ending at the temporal region, conjunctival congestion, clear cornea 10 mm in diameter, normal anterior chamber, pupil 2 mm reactive to light, clear lens, and normal fundus. Cleft extended downward from the right medial canthus involving the nasal ala and left forearm had an oblique-crease with camptodactyly. We thus report a case of anterior segment abnormality with an oblique craniofacial cleft. The cause of which is unclear, amniotic band syndrome being a possible cause.

Key words: Amniotic band syndrome, craniofacial cleft, ocular manifestations

Craniofacial clefts are disfiguring facial anomalies. Incidence is rare, ranging between 1.43 and 4.85/100,000 births.[1] In 1976, Tessier proposed an anatomic classification of the craniofacial clefts using the orbit as the primary structure of reference.[2] The number nine cleft is the rarest, described as lateral orbital cleft with soft tissue deformity involving the lateral third of the upper eyelid and underlying bony disruption extending into the temporal fossa.[3,4] To date, there have been a few reports of detailed ophthalmologic examination in cases of the craniofacial cleft. Here, we report a case of anterior segment abnormality with Tessier nine cleft, nasoschisis, and a left forearm crease.

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Case Report

A 20-year-old primi with regular prenatal care, no adverse obstetric or genetic history, normal prenatal laboratory test results and normal antenatal scans presented at term with premature rupture of membranes (PROM) in labor. A live 1480 g baby girl at 38 weeks, was delivered. The Apgar scores were 7 and 8 at one and 5 min, respectively. On subsequent examination, the neonates left eye and adnexa were normal. The right eye showed a cleft extending from the lateral third of the upper eyelid, eyebrow to involve the temporal fossa, with the absence of middle third of the upper eyelid with ectropion of the medial two-third and lateral third. Lateral third of the right eyebrow was absent with severe lower eyelid retraction with lateral canthal displacement exposing the right globe [Fig. 1]. A deficiency of the superior and lateral orbital rims was felt resulting in a lateral displacement of the globe. There was severe injection and chemosis of the bulbar conjunctiva, cornea measured 10 mm in horizontal diameter, clear. The anterior chamber was well formed, iris normal in color and pattern, pupil 2 mm round, regular and reacting to both direct and indirect light and lens appeared clear. The fundoscopic examination found normal optic nerves and maculae. A deep furrow was noted downward from the medial canthus medial to the lower punctum not involving the horizontal canaliculus, not involving the floor of the orbit. The lacrimal passage being patent with dysplasia of the right nasal ala. No cleft palate or lip was noted. The left forearm showed splitting of the superficial skin with crease extending from medial aspect of the elbow to base of left-hand index finger it thus being flexed with evidence of camptodactyly [Fig. 2]. Due to the extreme risk of exposure keratopathy, a moisture chamber was placed over the right eye, and ciprofloxacin eye ointment, preservative free moxifloxacin eye drops were applied four times daily. At 72 h after birth, the patient developed right eye exposure keratopathy despite topical therapy [Fig. 2]. X-ray skull anteroposterior and lateral view showed evidence of bony defect of the superior orbital rim with distortion of the greater wing of the sphenoid and squamous portion of the temporal bone [Fig. 3]. Subsequently, under general anesthesia the child underwent complete temporary tarsorrhaphy [Figs. 4 and 5] to prevent further corneal thinning, perforation, and left forearm contracture release. Thus addressing the cause of primary concern.

Discussion

Craniofacial clefts are extremely rare and disfiguring congenital anomalies of the face. Tessier in 1990 proposed an anatomic

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classification of craniofacial clefts. Of these, Tessier number nine facial cleft is found to be the rarest. The exact incidence of which is unknown, due to the rarity of its occurrence.[3] Tessier number nine cleft in literature is described as an upper lateral orbital cleft involving the lateral third of the upper eyelid and underlying bony disruption extending into the temporal fossa.[3,4] It is most commonly seen as a cranial extension of number five facial cleft.[3,4]

In studies by Van Der Meulen, this anomaly has been described as a frontosphenoid dysplasia. There are various presumed mechanisms that induce facial cleft, such as dysraphisms of facial processes, invagination of the amniotic band, and insufficient blood flow *in utero*. In our case, signs like PROM, multiple anomalies point toward amniotic band syndrome being the likely cause.[5,6]

Figure 1: Right eye examination-cleft extending from the lateral third of the upper eyelid, eye brow to involve the temporal fossa, with ectropion of the upper eyelid, lower eyelid retraction, and lateral canthal displacement

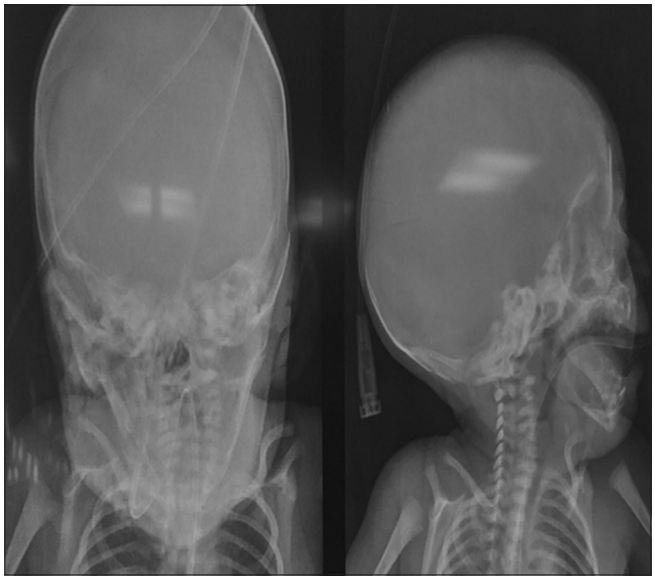

Figure 3: X-ray skull anteroposterior and lateral view showed evidence of bony defect of the superior orbital rim with distortion of the greater wing of the sphenoid and squamous portion of the temporal bone

Amniotic band syndrome is sporadic in occurrence with an incidence of 1:15,000 births. It includes a collection of fetal malformations caused by the entanglement of amniotic remnants after rupture of the amniotic sac.[7] Although it primarily affects external structures, associated malformations of internal structures also have been reported.[8,9]

The type and extent of ophthalmic defects depend on the band location and timing. Defects may include a combination of bony orbital clefts or hypertelorism; lid abnormalities such as colobomas, ptosis, and ectropion; lacrimal outflow obstruction; and exposure keratopathy.[7] Other congenital anomalies associated with it include clubfoot, facial cleft, cleft lip and palate, and supernumerary teeth.

Given the variety of craniofacial lesions encountered, the surgical approach needs to be tailored to the craniofacial defect.

Figure 2: Right eye exposure keratopathy. Left forearm shows splitting of the superficial skin with crease extending from medial aspect of elbow to base of left index finger with evidence of camptodactyly

Figure 4: Right eye postcomplete temporary tarsorrhaphy

Figure 5: The child 7 days postright eye complete temporary tarsorrhaphy

Oculoplastic surgery to address functional needs is of primary concern. Later, cosmetic issues can be addressed.

In the case described, the neonate presented at birth with a craniofacial cleft number nine with soft tissue and underlying bony involvement, nasoschisis suggestive of an atypical cleft number two with no evidence of cleft number five[11] and a left forearm crease which could be a result of amniotic band sequence. Even with the high risk of amblyopia the child underwent a complete temporary tarsorrhaphy to prevent further corneal thinning and perforation.

The child was then advised to undergo regular follow-ups at 4 weeks followed by at 3 monthly intervals. Release of the tarsorrhaphy sutures of a few millimeters distance centrally would be done to assess the status of the cornea, perform a retinoscopy to determine the presence of an underlying refractive error in view of the risk for amblyopia and to determine the timing for release of the tarsorrhaphy with further reconstruction of the upper eyelid and lateral canthal tendon.

The various oculoplastic alternatives include lateral canthotomy and cantholysis with Tenzel procedure, rotational flaps for medium-sized defects.[10,11]

The present study thus describes the ophthalmic management of Tessier number nine cleft and thus, may be helpful in counseling and estimating the impact of this syndrome on the eye when in utero.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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