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Case Report

An unusual craniofacial cleft: Amniotic band syndrome as a possible cause☆,☆☆

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ABSTRACT

We report the case of a no. 4 Tessier cleft in association with an unknown cleft of the mandible extending to the external auditory meatus. This has not been previously published in the literature and its underlying pathology remains undetermined. The nature of the cleft, possible classifications, and potential embryologic origins will be discussed. Amniotic band syndrome is the most likely cause of the cleft.

1. Case report

The patient is a 6-year old female who was referred for craniofacial malformations present since birth. She had no other diagnosed medical conditions, physical exam revealed no concern for cardiac abnormalities or neurological impairment. Her parents reported no significant past medical history and her pregnancy was without complications.

The left sided cleft began at the lower eyelid lateral to the medial canthus and extended across the orbital rim, medial to the infraorbital foramen, and into the upper lip between the philtral column and commissure. There was distortion of the anterior maxillary sinus and the cleft extended into the alveolus between the lateral incisor and canine. There was right-sided deviation of the nose due to traction from anomalous insertion of the orbicularis oris. The findings were most consistent with a Tessier no. 4 cleft (Figs. 1–5).

The mandibular cleft was a soft tissue ridge within a depression 3 mm wide. It began at the lower lip vermillion border inline with the upper lip cleft, with a lateral extent between lower incisor and canine. This extended inferiorly and obliquely to the midline at the pogonion and then posteriorly to the angle of the mandible. The cleft paralleled the angle of the mandible until it terminated in the external auditory meatus. There was effacement of the concha, tragus, and anti-tragus as well as pre auricular skin tag. The tympanic membrane was intact and not involved. She had no hearing loss in the involved ear. Cranial nerves, including VII, VIII, and V, were intact bilaterally. The mandibular bone was not involved in the cleft (Figs. 1–5).

The soft tissues of the cleft were closed by incorporating a z-plasty infraorbitally. This allowed enough vertical length to approximate the lower eyelid and upper lid vermillion border without tension. The closure was successful in creating a functional lower lid and upper lip.

2. Discussion

Paul Tessier introduced the Tessier classification of clefts in 1976 and it has since become the standard for descriptions of craniofacial clefts [1]. The Tessier classifications describe clinical patterns of clefts formed by anomalies in the formation and organization of embryologic structures. Updated classification systems have been proposed that suggest an embryopathogenesis

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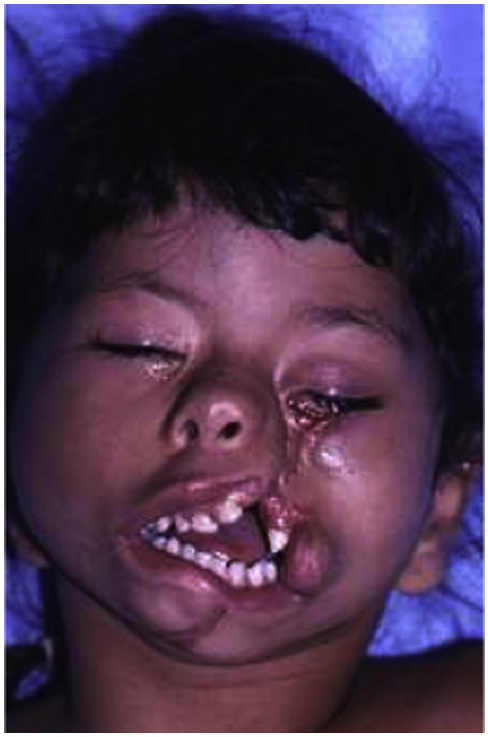

Fig. 1. Left sided cleft beginning at the lower eyelid, lateral to medial canthus, and extending to upper lip between the philtrum and commissure. This is consistent with a Tessier no. 4 cleft.

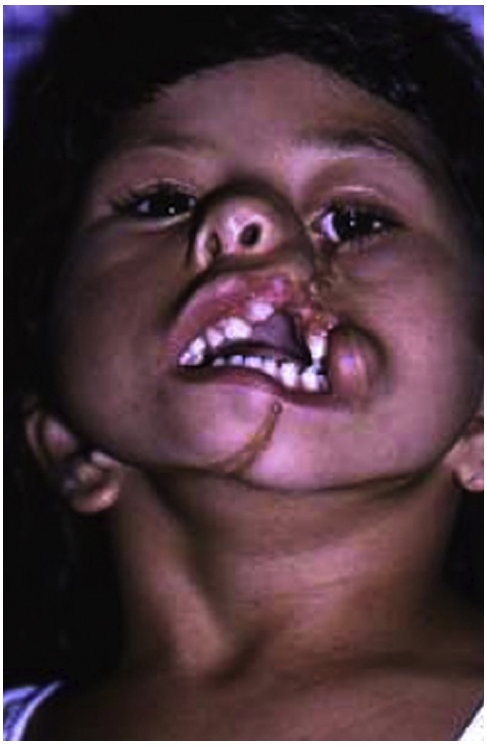

Fig. 3. Collinear cleft extending inferiorly, obliquely, and laterally from the lower lip, across the pogonion, to the right angle of the mandible.

for cleft development and also allow for a greater description of the defects [2]. Certain combinations of clefts frequently occur in the same patient leading to named syndromes such as Goldenhar's, a 7 and 8 cleft, and Treacher Collins, a 6, 7, and 8 cleft [3]. Non-syndromic combinations of clefts are rare, as is the no. 4 cleft [4]. Rare facial clefts as a group occur in 1.43–4.85 per 100,000 births [3].

Presented here is a rare facial cleft not described by traditional Tessier classification. It begins with a left sided no. 4 cleft. An additional unclassified cleft then begins on the paramedian

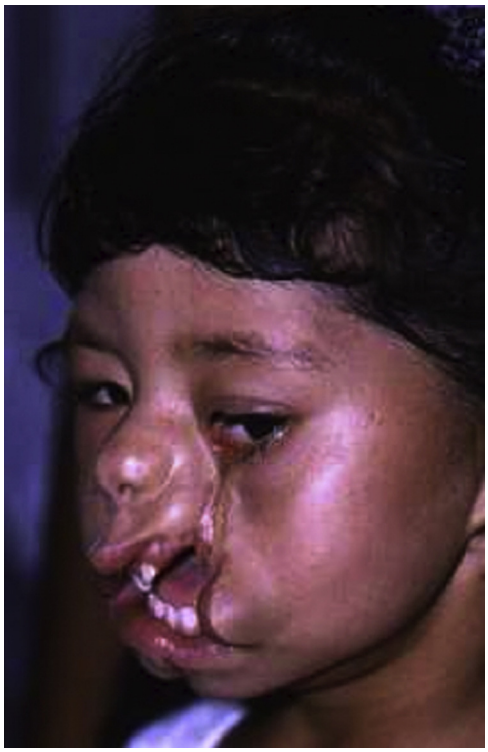

Fig. 2. The cleft extends across stoma and is collinear with lower lip and the obliquely oriented mandibular cleft.

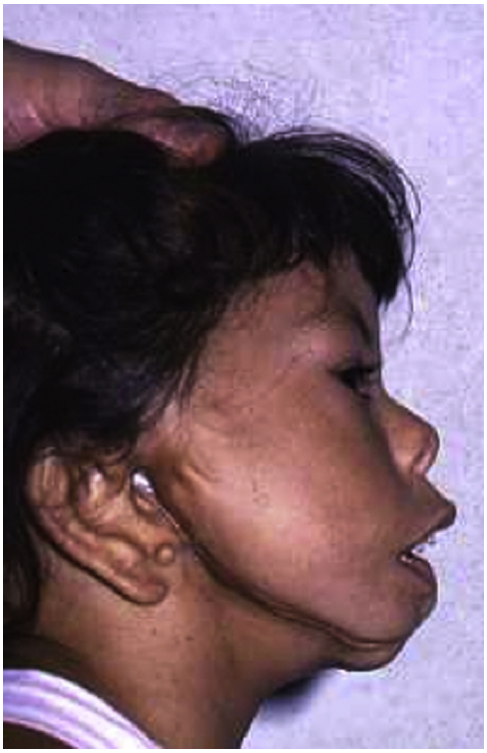

Fig. 4. The cleft paralleled the mandible and terminated at the external auditory meatus. A soft tissue ridge is apparent.