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CASE REPORT

Congenital Lateral Nasal Proboscis with Orbital Mass

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ABSTRACT

Proboscis lateralis is a rare craniofacial malformation frequently associated with congenital ocular abnormalities. These abnormalities included anophthalmia, microphthalmia, microcornea, lenticular opacities, cyclopean eye, and colobomas of the choroid, retina, iris and eyelids. We report a case of unilateral proboscis lateralis in a newborn infant with an associated solid orbital mass.

Keywords: Proboscis lateralis, Orbital mass, Congenital.

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INTRODUCTION

The congenital malformation of heminasal aplasia with lateral nasal proboscis is a rare sporadic disorder that was first described by Forster[1] in his article on congenital malformations. Proboscis lateralis is a rare facial anomaly (1:100,000 birth).[2] This malformation is almost always associated with anomalies of the eye and its adnexa, and may also be associated with a cleft lip or cleft palate. Proboscis is typically described as a pendular mass arising from the region of the inner canthus. It is like a tubular structure and is lined by normal skin. This trunk like appendage is generally 2 to 3 cm in length and 1 cm in diameter and has a central tract lined with respiratory epithelium. Khoo Boo Chai collected 34 cases from literature, classified them into four groups.[3] Group I has a lateral proboscis with a normal nose; group II, lateral proboscis with an ipsilateral deformity of the nose; group III, ipsilateral deformity of the nose and the eye and/or ocular adnexa; and group IV, cleft lip and/or palate in addition to the nasal and eye and ocular adnexa. The present report describes a patient with lateral nasal proboscis with an orbital mass.

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CASE REPORT

A 3-day-old male child presented with abnormal growth above and medial to the right upper eyelid since birth. This was associated with incomplete formation of the upper eyelid which was causing incomplete closure of the palpebral fissure. The right nostril was not formed. The systemic history was unremarkable. On examination a pedunculated mass lesion with normal appearing skin was present which was located on superomedial aspect of the right upper eyelid (Fig. 1). The patient had an associated firm orbital mass in superomedial quadrant that was free from overlying skin and underlying bone. The lesion was nontender and not associated with any other sign of inflammation. Pediatric consultation was requested which ruled out any other systemic abnormality. Examination of the oral cavity and palate revealed no abnormality. CT scan of the orbits revealed an underlying orbital mass (Fig. 2) that was heterogeneous and related to superior orbital wall. The mass was not communicating with the cranium or nose. The CT scan also revealed right heminasal aplasia. The various management options were discussed with the parents of the child and the patient was planned for lateral nasal proboscis and orbital mass excision with upper eyelid reconstruction under general anesthesia. The pedunculated mass along with orbital mass was excised and sent for histopathology. The features of the pedunculated mass on histopathology were consistent with lateral nasal proboscis (Fig. 3) without any immature tissue or malignancy. The orbital mass contained fibrous tissue with interspersed vessels and mesenchymal tissue with presence of epithelium lining similar to the lateral nasal proboscis.

DISCUSSION

Proboscis lateralis describes a rudimentary nasal structure or appendage that is located off-center from the vertical midline of the face. Although, it was initially reported that no sex predilection existed, Boo-Chai[3]

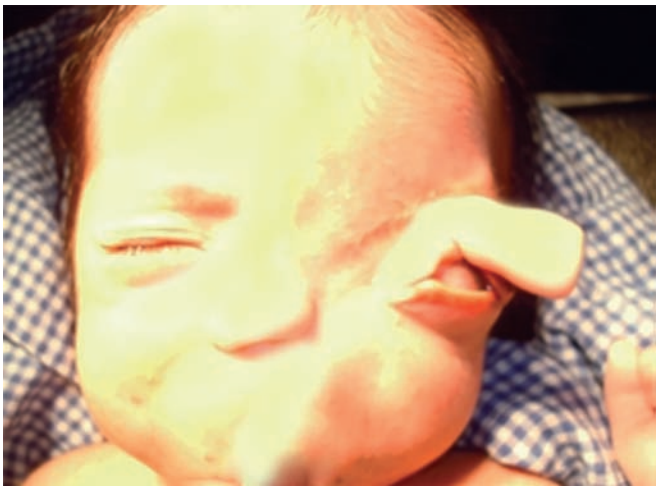

Fig. 1: Clinical photograph of the patient showing lateral nasal proboscis and heminasal aplasia

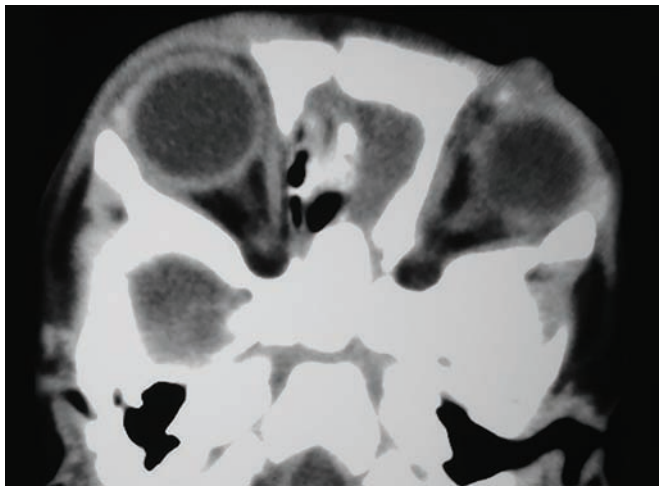

Fig. 2: CT scan showing hyperdense heterogeneous orbital mass

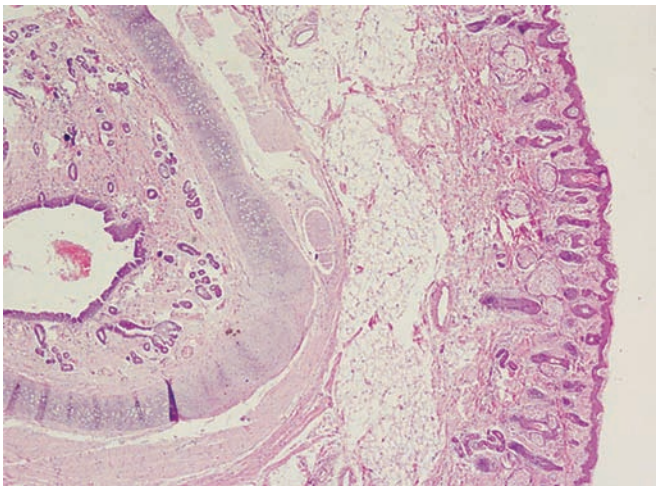

Fig. 3: Photomicrograph showing central lumen and epithelial lined mucosa with cartilage in the stroma (viewed under 10× using hematoxylin eosin stain)

noted a 2:1 male female preponderance. Review of 9 cases subsequent to Boo-Chai[3]s report supports the notion of a male preponderance with a 3:1 male female ratio.[4-10] There does not seem to be any racial predilection in proboscis lateralis. Lateral nasal proboscis is a rare anomaly resulting in incomplete formation of one side of the nose and other variable abnormalities in the adjoining regions of the face, without associated brain malformations. Heminasal hypoplasia or aplasia is the most common abnormality in proboscis lateralis although, in rare cases, the nose is normal. Of interest to the ophthalmologist is the frequent association of abnormalities of the eye and ocular adnexa with proboscis lateralis. Although, Wang et al[11] reported that ocular defects are rare in patients with proboscis lateralis, a subsequent review of the literature by Boo-Chai[3] noted that 24 of 34 patients with proboscis lateralis had associated anomalies of the ipsilateral eye and/or ocular adnexa. These abnormalities included anophthalmia, microphthalmia, microcornea, lenticular opacities, cyclopean eye, and colobomas of the choroid, retina, iris and eyelids. In our case, there was upper eyelid coloboma. Association of orbital mass with lateral nasal proboscis is very rare. Although association of cystic orbital masses such as encephalocele[12] and cystic teratoma[13] are documented in the literature, benign solid orbital mass sharing histopathological features of lateral nasal proboscis is not known. To the best of our knowledge, the present case represents first such pathology. Because there is some variability in facial anomalies and the degree of nasal hypoplasia seen with proboscis lateralis, management must be individualized. When marked hypoplasia or aplasia of the nasal ala is present, reconstruction is indicated. The structure and texture of the proboscis make it an ideal substrate for nasal reconstruction, and for this reason, the proboscis should not be excised if future nasal reconstruction is anticipated. Depending on the size and location of the proboscis and the degree of nasal hypoplasia, a variety of techniques may be used to reconstruct an esthetically acceptable nare, including use of the proboscis as a pedicle flap.[3,4] Since, the parents of the child did not want to wait any longer for surgical reconstruction, so surgical excision was done and the rudimentary tissue was sacrificed followed by eyelid reconstruction.

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