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CASE REPORT

Proboscis Lateralis: A Rare Case

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ABSTRACT

Proboscis lateralis (PL) is a rare congenital anomaly with a characteristic appearance. We present such a case in 5-year-old child which was managed by excision. Complete surgical excision at the base of the proboscis is desirable as a primary procedure if there is adequate ipsilateral nasal development or as a delayed excision if the proboscis is to be used in nasal reconstruction. Definitive cosmetic reconstruction with bone or cartilage should be planned at a later age keeping in mind the possibility of further growth and development of the nasal complex, however no such reconstruction was done in the present case as there was no gross asymmetry.

Keywords: Congenital, Anomaly, Nose, Proboscis, Lateralis.

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INTRODUCTION

Proboscis lateralis (PL) is a rudimentary nasal structure or appendage that is located off-center from the vertical midline of the face. It is a rare craniofacial malformation frequently associated with abnormalities of the eyes and or ocular adnexa. Its reported incidence is less than 1 in 100,000.[1] The various localization points of PL may be at the embryonic fusion line between the anterior maxillary process and the frontonasal process, level 1, supramedial canthal area; level 2, medial canthal area; level 3, inframedial canthal area; level 4, supranostril area.[2]

There is no consensus regarding the pathogenesis of PL, however it appears that it is a developmental defect involving the nasal placode, which is the primary organizer of the nasal area of the midface.[3,4] PL is included in a spectrum of malformations described as developmental field defects. This terminology refers to those conditions which represent an embryological area where an error causes a major anomaly which, in turn, disturbs contiguous developing areas. It occurs more often in the midline.[5] We report a case of PL in 5 years old child.

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CASE REPORT

A 5-year-old male child presented with the malformation of the nose. The child had mucocutaneous out growth from the inframedial canthal area on the left side of the nose (Fig. 1). There was associated hypoplasia of the left nasal cavity. He had a normal 10-year-old elder brother and 8-year-old elder sister. Both the 30-year-old mother and the 35-year-old father were normal. His parents had the habit of neither drinking nor smoking. There was no report of this deformity in either parent's family. The pregnancy was unremarkable, without maternal diabetes infections, and resulted in a normal spontaneous vaginal delivery at home. There was no history of any consanguinity among the parents. There was no history of any drug intake during pregnancy or any exanthematous fever or exposure to the X-rays in the antenatal period. The eyes of the child were normal including the normal fundus and there was no other associated malformation of the face or any other part of the body.

The patient had a tube-like structure attached to the left side of the dorsum nasi. It was about 2 to 3 cm in length and 1 cm in diameter, with a small discharging tract. There was mild hypoplasia of the nasal cavity on the side of the proboscis. There was no other craniofacial anomaly. There was no cleft palate, cleft lip or choanal atresia. There were no associated central nervous system anomalies which are commonly associated with midline proboscis. The tip of the structure was hollow, it accepted a 6 FG infant feeding tube and occasionally released mucous discharge. The infant feeding tube could go upto 1 cm through the proboscis, but could not be negotiated in the nasal cavity. CT was obtained to evaluate the full extent of the congenital anomaly and to rule out communication between the intracranial compartment and the proboscis prior to surgical repair. The right side was normal and the right nasal airway was patent (Fig. 2).

Surgical excision of the proboscis was performed at the age of 5 years. Since the nasal alae were relatively well developed, reconstruction was unnecessary, and the soft tissue appendage was simply amputated from its origin at the left inframedial canthal area. The raw area was sutured in layers (Figs 3 and 4). Sutures were removed on the 10th postoperative day (Fig. 5).

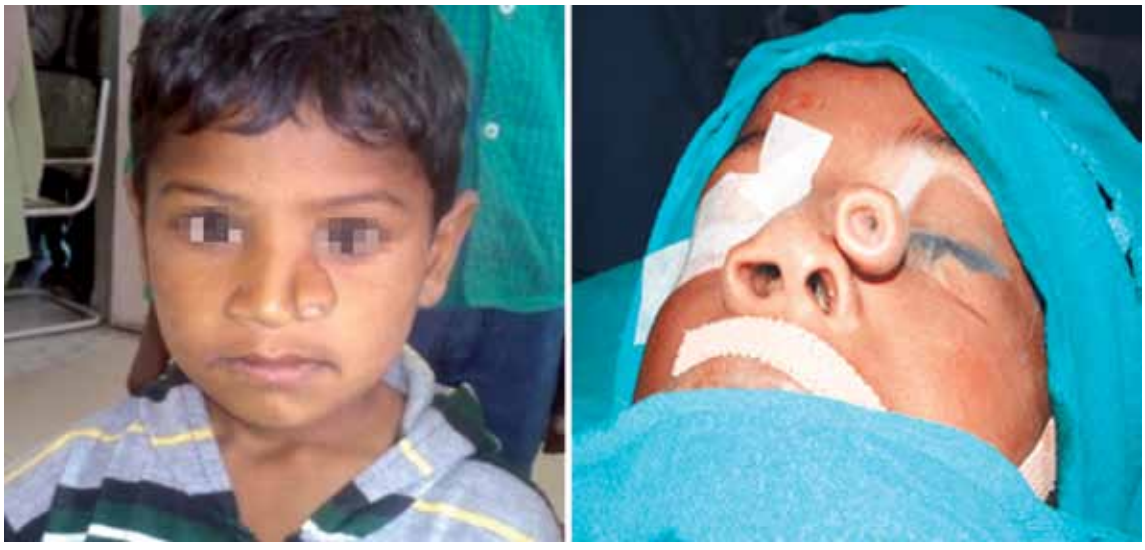


Fig. 1: Proboscis lateralis

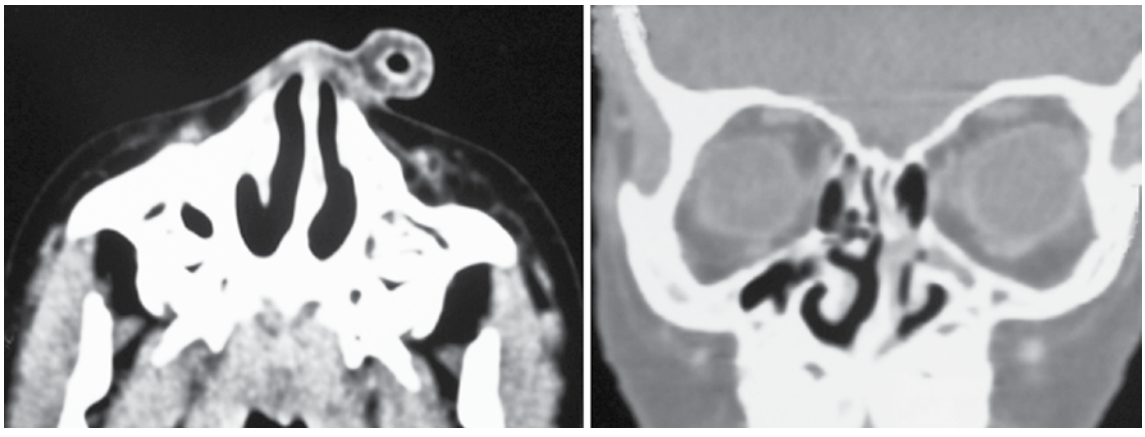


Fig. 2: Axial and coronal view of CECT of the patient with proboscis lateralis

DISCUSSION

Proboscis lateralis typically manifests as a soft, 2 to 3 cm long and 1 cm diameter trunk like process originating from the region of the medial canthus. It is characterized by a central tract lined with squamous and ciliated respiratory epithelium that typically expresses clear mucus from the blind dimple at its distal end as was the case in our patient. The precise embryologic mechanism responsible for the development of it has not been defined. Popular theories include imperfect fusion of the lateral nasal and maxillary processes and aberrant fusion of the maxillary process of the affected side to the medial nasal process (globular process).[6] Rontal and Duritz correctly pointed out that these theories do not adequately explain the associated anomalies and suggested a primary insult to the nasal placode as the likely mechanism for PL development.[7]

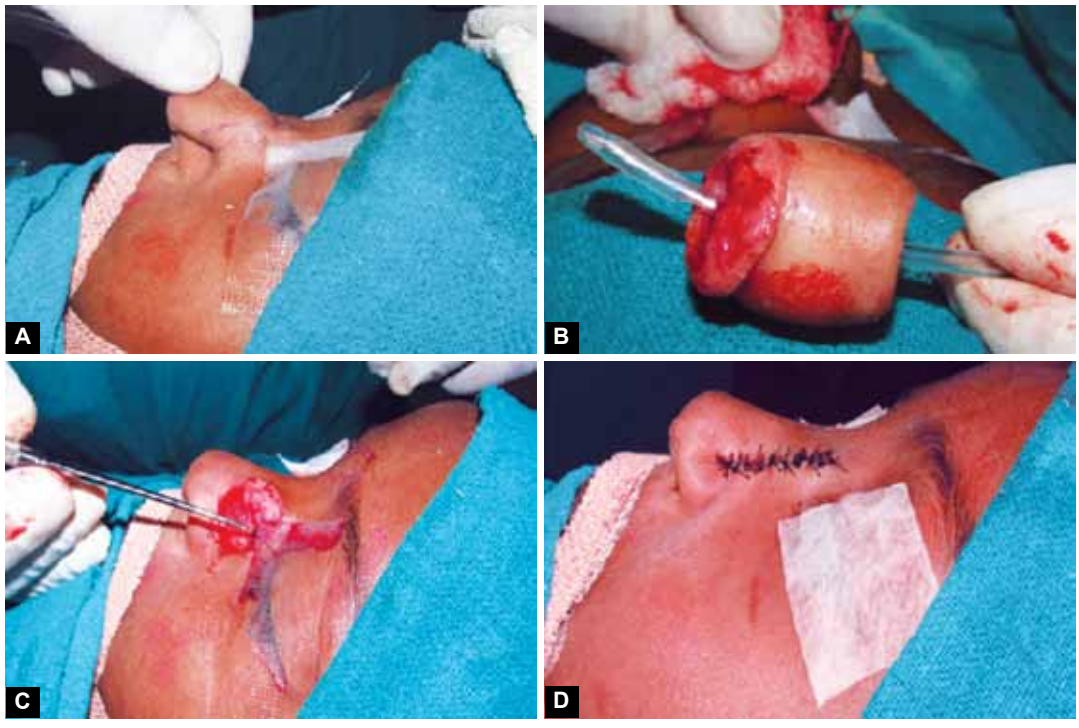
The presence or absence of ocular abnormalities was used by Boo-Chai to help categorize patients with proboscis lateralis into four groups:

- *Group I*: Lateral proboscis with normal nose (least common).
- *Group II*: Lateral proboscis with an ipsilateral deformity of the nose (second in frequency).
- *Group III*: Lateral proboscis with ipsilateral deformity of the nose, eye and or ocular adnexa (the most common type).
- *Group IV*: Lateral proboscis with ipsilateral deformity of the nose, eye and or ocular adnexa, plus cleft lip and/or palate.[6]

PL with or without heminasal aplasia appears to be the mild end of the spectrum of atypical clefting syndrome.[6]

For the complete evaluation of this anomaly CT scan is important which allows assessment of growth of facial and skull bones as well as CNS implications. Management should start early in childhood to avoid psychological consequences related to this deformity. For the heminose reconstruction, use of proboscis itself is the best option. Later secondary procedures are required to correct skeleton deformity and groove of the inset of proboscis with normal heminose.

Initial reports regarding the treatment of proboscis lateralis recommended simple surgical excision of the proboscis. More recently, surgical management of it has been approached with reconstruction in mind. Many authors advocate use of the proboscis to reconstruct the nose because



Figs 3A to D: (A) Marking of the incision site, (B) amputated proboscis, (C) raw area postamputation and (D) lateral view of nose after suturing of the raw end

Fig. 4: Asymmetry of the nasal cavities

it provides excellent color and texture match and is readily available.[8] Given the high degree of variability of associated anomalies, an individualized approach is suggested for surgical management. In general, surgical repair should be accomplished as early as possible without adversely affecting the cosmetic outcome.[9]

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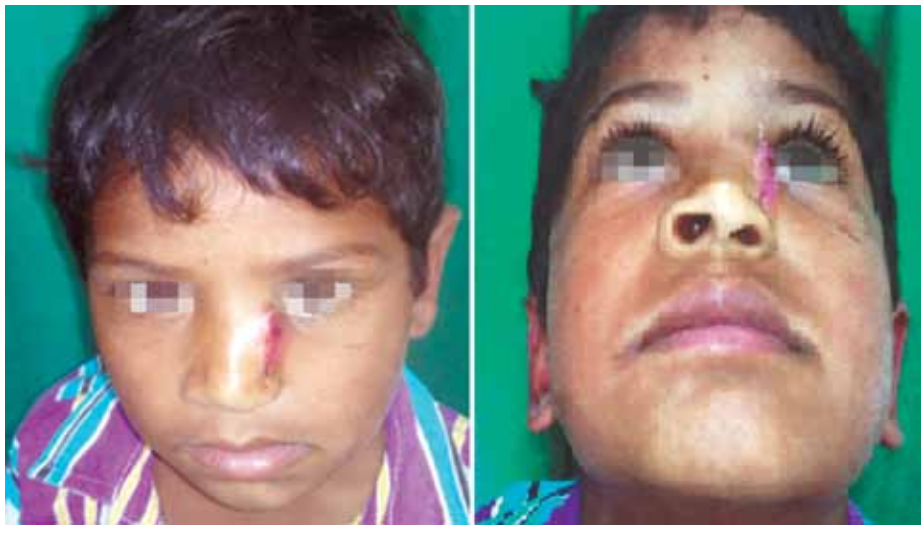

Fig. 5: Postoperative picture after suture removal

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