

Proboscis Lateralis - A Case Report



Medical Science

KEYWORDS : Proboscis lateralis ; Congenital anomaly ; Tubular nose ; Nasolabial groove

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ABSTRACT

Proboscis lateralis is a rare cranio-facial malformation for which no embryological basis has been established however it is embryologically related to the median facial cleft , and may be associated with other anomalies of the eye and its adnexa and cleft lip and palate. A child with type 2 Proboscis lateralis was treated by single-stage simple excision of the defect with fashioning of the ipsilateral Lower lateral cartilage to appropriate contour for Naso-labial groove correction. This procedure helped in achieving satisfactory cosmetic result.

Introduction

Proboscis lateralis is a rare congenital anomaly which occurs in approximately 1 each 100,000 births [1]. There are just a few cases related in literature. It is typically described as a tubular, pendulum-like structure, with a consistency similar to the lateral nasal wall, which usually originates in the inner corner of the orbit, and, atypically, outside of it. It has a small longitudinal central channel, with a stratified epithelium, which discharges mucus, draining it to an orifice in the distal. The proximal end is closed, and it has no communication with adjacent structures. It is usually associated to nasal anomalies on the same side, with malformations in the nose area in most cases, as well as with heminasal aplasia or hypoplasia. Nasal cavity is partially developed, and the nose is rarely normal [2,3,4].

Such malformation is followed by other anomalies, and the ophthalmic ones being more frequent, present in 44% of the patients (anophthalmia, microphthalmia and colobomas), facial anomalies in 38% (maxillary hypoplasia or facial clefts) and craniocerebral anomalies in 18% encephalocele and holoprosencephaly [5,6].

Iris coloboma and cleft palate and labial clefts are more frequently associated. Histopathological examination shows the skin with sebaceous glands, conjunctive tissue with sebaceous glands and striated muscular fibers of the same type found in nasal muscles, and some cartilaginous element, corresponding to the normal sidewall nose cartilage [5].

The treatment adopted is surgical, with the proboscis as the ideal donor for the reconstruction of the nasal deformities associated, due to its texture, color and neighboring location characteristics. Surgery is recommended before 4 y.o., before school age, due to the psychological consequences both for the patient and the family [6,7,8,9].

Proboscis is apparently derived from the lateral nasal process, when several ipsilateral nasal abnormalities are observed. In the case of proboscis formation with a normal nose development, the possibility of an hypertrophy of a small part of the lateral nasal process is, remaining from the normal tissue, unleashed by an exogenous stimulation, during the 3rd or 4th week of fetal life [10,11].

Congenital malformations of the nose are rare and result from an aberrant embryological development. Differential diagnoses include duplications, polyrrhinia, supernumerary nostril, and proboscis lateralis.

Case Report :

A 6-year-old boy from lock-manufacturing vicinity of Aligarh presented with a 0.5 * 1.5 cm outward defect arising from his left nostril with no other associated mid face disorder. The left and

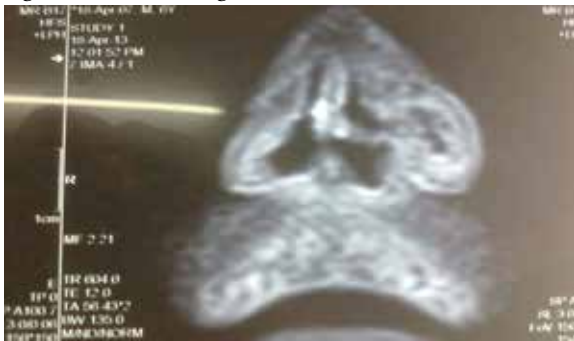
right nostrils of the child were functioning aptly. The defect was confined to left nasal cavity only with no associated defect of eye or its adnexa, lip and palate. The boy also had no history of difficulty in breathing and any discharge from the appendage. No other craniofacial or congenital abnormality was observed. A diagnosis of Proboscis Lateralis (Boo-Chai Type 2) was made and the condition explained to the guardian of the boy. Her mother wished to get the appendage removed. Proboscis excision was planned along with reconstruction of nose. Under General Anaesthesia using Lateral Rhinotomy approach the Proboscis, that ended blindly at the Left Middle Turbinate was excised completely. Mucosa and Skin were sutured with respective approximation. The patient was discharged 3 days later only to contract measles at the time of suture removal. He had to be transferred to Paediatrics but was then lost to follow-up, only to reappear after 5 months.



Fig 1 : patient profile



Fig 2 : patient profile

Fig 3 & 4 : MRI showing Nasal Disease**Fig 5 : 7 weeks post op patient profile****Discussion :**

Proboscis lateralis is a rare facial anomaly with occurrence of 1:100,000 births and result in incomplete formation of nose , eye and adnexa [12]. Its archival mention dates back to 1861 when Forster published in his monograph 'congenital anomaly of the human body' [13]. Later in 1884 Selenkoff described in detail the autopsy findings of a farmer with Proboscis[14]. Boo-Chai noted a 2:1 male female preponderance and after a review of literature classified proboscis lateralis into 4 groups[13].

- i.e. : type 1 - Proboscis with Normal Nose
 type 2 - Proboscis with Nasal defect only.
 type 3 - Proboscis with Nasal Defect + Eye & Adnexa defect
 type 4 - Proboscis with Nasal defect + Abnormality of Eye & Adnexa + Cleft Lip/Palate or both

Proboscis lateralis is primarily a malady of the mid-face however its embryological reason of development is still not clear. It is hypothesized that the embryological defect that results appears to involve the nasal placode which is a primary organizer of the nasal area of the mid-face[13]. The development failure or absence of medial and lateral nasal processes results in fusion of the maxillary process with the contralateral frontal process[15]. By and large there is heminasal hypoplasia/aplasia on the side of the proboscis although; in rare cases the nose is normal. It may or may not be associated with Cleft lip/palate disorder. Worth mentioning is the fact that majority of patients with Proboscis lateralis donot have grave CNS abnormalities.

Proboscis lateralis should be managed immediately in early childhood to evade psychosocial corollary related to this illness [16]. However another noteworthy fact is that the Proboscis should not be excised if future nasal reconstruction is anticipated because texturally & structurally the Proboscis make it an ideal substrate for nasal reconstruction.

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