

Proboscis lateralis: Review of literature and a case report

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Abstract

Proboscis Lateralis is a rare congenital anomaly, which 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 Group three proboscis lateralis was treated by using the flaps of the proboscis lateralis, one for the correction of maxillary hypoplasia and the other for correction of the grooving in the nasolabial region.

This technique helped in achieving a good cosmetic outcome.

Various methods have been suggested for the treatment of proboscis lateralis, ranging from the extirpation of the proboscis, to the tunneling method. We have suggested the use of the flaps from the proboscis, which gave us the advantage of a one-stage correction of the proboscis along with the maxillary hypoplasia, and to minimize the grooving in the nasolabial region.

INTRODUCTION

Proboscis lateralis is a rare congenital anomaly, which is embryologically related to the median facial cleft^{1,2,3,4}. Various craniofacial anomalies are associated with it, apart from hemi nasal aplasia or hypoplasia. The reported incidence of proboscis lateralis is less than one per 100,000 live births, without any sexual preference². The proboscis may be unilateral as well as bilateral, the former being the more common type^{1,3,6,7,8,10}. There is a tube like rudimentary nasal structure which is attached along the embryonic fusion lines between the anterior maxillary process and the frontonasal process^{1,5}. The length of the proboscis may vary from 1 to 4 cms in length^{1,3}. It has a stratified columnar lining and ends as a blind tract, without any connection with the nasal cavities.

According to Boo- Chai⁶ and Guerrero et al.,⁷ it was Forster who first mentioned this anomaly in his monograph entitled "Congenital anomalies of the human body" in 1861. However, according to Binnis,⁸ Selenkoff is believed to be the first, since his detailed report of the autopsy findings of a farmer patient with proboscis was published in 1884^{2,3}. The patient had lacked the right nasal bone, right frontal and maxillary sinus, vomer, premaxilla, right upper second incisor, and the right lateral nasal wall.

Antenatal diagnosis is now possible with development in radio diagnostic techniques⁴. MRI and CT studies have shown that there are rudimentary nasal bones, and the

ethmoids and the frontal sinus may be absent as well. There may be associated CNS anomalies. For the complete evaluation of the anomaly CT or MRI is important as it allows for assessment of the growth of the facial and skull bones as well as the Central nervous system development.¹¹

This malformation is almost always associated with anomalies of the eye and its adnexa, and may also be rarely associated with a cleft lip or cleft palate. Khoo Boo Chai collected 34 cases from literature, classified them into four groups, discussed various management options and advised dilatation of proboscis before starting the reconstruction⁶. The four groups are as follows:

1. Group 1 consists of Proboscis with normal nose.
2. Group 2 consists of Proboscis with nasal defect only on the same side.
3. Group 3 consists of Proboscis with ipsilateral nasal defect is associated with eye or adnexal defects. It is the most common type.
4. Group 4 consists in addition to the nasal and ocular defect, a cleft of the lip or the palate.

Proboscis lateralis necessitates surgical treatment.

Management should start early in the childhood to avoid psychosocial consequences related to this deformity.

Complete aesthetic outcome may be delayed until late teens

when growth of the nasal skeleton is almost complete^{6,25}. For the hemi nose reconstruction, use of proboscis itself is the best option. Later secondary procedures may be required to correct skeletal deformities.

Although many surgical techniques have been described, there are two eminent techniques. According to Denecke and Meyer,¹ Young was the first to mention correction of proboscis in literature in 1949: the medial or distal half of the tube is deepithelised and sutured to the split lateral wall of the ipsilateral nose¹. The other frequently used technique is the tunneling method. According to Denecke and Meyer¹ this method was first reported at the French convention for plastic surgery in 1956 by Recaimer and Florentin. The tube is totally deepithelised except for the distal segment, which will form the new ala. It is then brought to the area of missing nostril through a subcutaneous tunnel formed on the lateral wall of the defect side of the nose.

In this report, we present a case of Group 3 Proboscis lateralis, encountered by us for the first time.

CASE REPORT

A 4-year-old female child from Sabarkatha district of Gujarat state presented to us with a 4.2 cm long, 1.2 cm diameter tubular structure attached to the left medial canthal region with a pedicle. She was the younger of the two siblings and there was no history of any congenital anomaly in the family. There was no history of any consanguineous marriages. The mother had a full term uncomplicated normal vaginal hospital delivery. The prenatal history was negative for exposure to alcohol, smoking, drugs or radiation. Physical examination revealed that there was clear mucoid secretion from the distal end of the tube. There was a hypoplastic left nostril with a normal right nasal framework. The nasal airway was patent on the right side, but not on the left. The maxillary area on the left side was hypoplastic. There was a coloboma on the medial third of the left lower eyelid causing epiphora, and the nasolacrimal canal was absent. However there were no visual disturbances (Figure 1). The patient was diagnosed as having Group 3 proboscis lateralis, according to the classification of Boo-Chai, because of the associated ocular anomalies.

Figure 1

Figure 1: Preoperative photographs



The CT scan showed absence of pneumatization of the left maxillary, ethmoids, and sphenoid sinuses. There was hypoplasia of the frontal sinus. There was lack of development of the superior, middle, and inferior turbinates on the left side. The maxillary sinus on the right side was normal. Both orbits, zygomatic bones, zygomatic arches appeared normal.

Procedure done was creation of left side hemi nose and filling of infraorbital soft tissue deficiency using proboscis. Under endotracheal intubation after painting and draping the measurements were made. The length of proboscis was 4.2 cm. The distance through the radix to the tip of the nose right side was 4.5 cm. The distal tip of the proboscis had hair and mucous discharge. The hole was dilated with urethral dilator. The nasal sill on the right side was 12.2mm and the ala length along the curve was 2.2cm. The flaps 1 and 2 were marked on both sides after the tube was opened posteriorly till close to the base through a horizontal incision over the medial canthal region and after creating the left side alar defect the skin between two incisions was tunneled widely beyond the right side dorsum and the infraorbital region left side leaving the alar rim area which was marked and the base area. The remaining area was deepithelised and demucosalised (Figure 2). The roof of the proboscis area, the mucosa was charred off with surface cautery for fear of hampering the vascularity of the proboscis. The proboscis had a similar skin texture and color match as that of the nose and had highly vascular subcutaneous tissue supplied by longitudinally running large sized vessels.

Figure 2

Figure 2: Markings and intraoperative photos after deepithelisation and flaps creation



The whole proboscis was tunneled through this subcutaneous tunnel and the ala was sutured to the left side hemi nose incision. The inner aspect blind sac mucosal incision line was sutured to the flap margins.

Through bolsters the flap 1 and 2 was sutured to respective places. Small area in the mucosal aspect of the nose was left unsutured and acted as drain in case of collection(Figure 3).

Figure 3

Figure 3: Flaps sutured with help of bolsters



The patient had continuous mucoid nasal discharge post op for 4 to 5 days which subsided spontaneously. Apart from that the post operative recovery was uneventful . All sutures were removed on postop day 7. Because of absence of nasolacrimal duct on the left side, epiphora was persistent. The patient was discharged on post op day 10.

Figure 4

Figure 4: Post operative photograph at four months follow up



The coloboma was left untouched for secondary surgery along with lacrimal sac surgery. Drain kept and patient extubated after dressing.

DISCUSSION

Proboscis lateralis is a rare congenital anomaly among the nasal malformations. The proboscis is a ready tube pedicle, which can be used for absent hemi nose reconstruction ^{6,25} . It shows the best color and texture match with the opposite normal hemi nose. Many authors have suggested extirpation of the proboscis and later reconstruction by various tubed flaps ⁶ According to Boo Chai, the proboscis lateralis used to be extirpated in the early 1900s . Excision of the proboscis may still be adequate for patients included in the group 1 of Boo Chai classification ^{7,17} . According to Denecke and Meyer ¹ , Young was the first to publish the description of correction of proboscis lateralis, in 1949. He used the proboscis as a donor. Basically, two raw surfaces that would face one another, both in the proboscis and the hypo plastic or aplastic nose, were formed and sutured. This method is considered simple and can possibly be completed in one stage. The main disadvantage of this method is shortage of nasal lining and long vertical scars ⁶ .

Another popular method is the tunneling method, described by Recaimer and Florentin in 1956. The deepithelised proboscis is passed under a transverse bipediced flap prepared from the lateral skin of the defective nose. The advantage of this method is the absence of vertical scars ¹ .

Surgical treatment thus generally consists of two parts: the nostril is formed in the first step, and the deformity caused by the pedicle is revised in the second stage.

In our patient, we used the flaps of the proboscis for maxillary hypoplasia augmentation. The excellent vascularity of the proboscis made this possible. However, minor corrections were left for second stage along with the management of coloboma and the lacrimal sac.

Almost all authors begin reconstruction as early as one year of life^{6,7,13,14,16}. Some have suggested that reconstruction should be delayed to 3 to 4 yrs of age to avoid retarding the development of the operated hemi nose. In 14 year follow-up of a patient operated at the age of 6 months, Boo Chai⁶ showed symmetrical growth of the proboscis with the healthy side. In our case, we intervened as soon as the patient reported to us.

Thus, reconstruction should be initiated as soon as the surgeon finds it comfortable to handle the tissues. Also the psychological trauma to the patient and the parents should also be considered. Various factors like the group of the proboscis lateralis as described by Boo Chai, the location of the proboscis and the characteristics of the proboscis must be considered while deciding the plan of surgery.

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