Case Report

Proboscis lateralis- A 17 years follow-up, a case report

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ABSTRACT

Proboscis lateralis is a rare congenital anomaly for which no embryological basis has been established. Besides heminasal aplasia or hypoplasia, various other craniofacial anomalies are also associated with it. It is only uncommonly free of other anomalies. Evaluation of a patient should include CT scan examination to look for growth of facial and skull bones. Reconstruction should start at an early age. Proboscis itself is the best option for heminose formation. Cartilaginous or bony support can be planned later in the late teens to give good aesthetic result.

KEY WORDS

Proboscis lateralis, Heminasal aplasia, Congenital craniofacial anomaly.

INTRODUCTION

Proboscis lateralis is a very rare congenital anomaly. Forester made first mention of this anomaly in 1861 and later Selenkoff in 1884 described in detail the autopsy findings of a farmer with proboscis.[1] Khoo Boo-Chai collected 34 cases from the literature, classified them into four groups, discussed various management options, and advised dilatation of the proboscis before starting the reconstruction.

Developmental basis of Proboscis Lateralis is still not clear.[1-4] It has been related to the craniofacial clefts. The condition is caused by the developmental failure or absence of medial and lateral nasal processes, resulting in fusion of the maxillary process with the contra lateral frontal process.[5-7]

It is a fleshy club like structure with a narrow epithelial lined tract in the center. Its proximal end is attached in the area of medial canthus.[1,8-10] According to associated anomalies, Khoo Boo-Chai has divided proboscis into four groups.

GROUPS

1 Proboscis with normal nose
2 Proboscis with nasal defect only
3 Proboscis with nasal defect plus defect of eye and adnexa
4 Proboscis with nasal defect plus abnormality of eye and its adnexa plus cleft lip or palate, or both

The nasolacrimal duct ends blindly.[1,3] There may be associated CNS anomalies.

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 development.[1,11]

Management should start early in childhood to avoid psychosocial consequences related to this deformity. Complete aesthetic outcome is delayed until late teens when growth of nasal skeleton is almost complete.[1,12,13]

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 the proboscis with normal heminose.[1,8,14,15]

CASE REPORT

We are reporting a case of Proboscis Lateralis, seen by us for the first time in 1985. Now we have a 17- year follow-up of this patient. The patient was a male child first in the order among four children of his farmer father. He was from Bhavnagar district of Gujarat state. He was of one-year when he had presented for the first time.

All the siblings were normal and there was no history of any congenital anomaly in the family. The mother was absolutely well during pregnancy and delivery was normal and was at full term. Growth of the child was adequate for his age. There was no history of consanguineous marriage in the parents.

The child was normal in his behavior and intelligence. He attended school and was interested in studies.

Our patient had following abnormalities: (Figures 1-6)

1. A right proboscis lateralis.
2. Heminasal aplasia on the ipsilateral side.

Patient had no other associated anomaly specially mentioning about absence of any ocular deformity. There was a small opening on the affected side leading to the nasal cavity.

The patient was of the group-II according to the description of Khoo Boo-Chai.

CT scan showed (Figures 7 & 8) absence of pneumatization of right maxillary, ethmoid, and sphenoid sinuses. There was lack of development of nasal bone on right side. There was lack of development of superior, middle, and inferior turbinates on right side. Septum was deviated towards right side. On the left side, turbinates are hypertrophied and maxillary sinus is enlarged.

Both orbits, zygomatic bones, mandible, hard palate, frontal bones, and zygomatic arches appeared normal.

TREATMENT

We submitted the child for reconstruction at an age of 1 year. We opted for staged reconstruction. In the first step, we splitted the proboscis and attached it to the side of the normal opposite heminose (Figure 2). In the second stage we transferred the upper end and

Figure 1: At the time of presentation during infancyTubular structure arising from the medial canthus area and absence of heminose

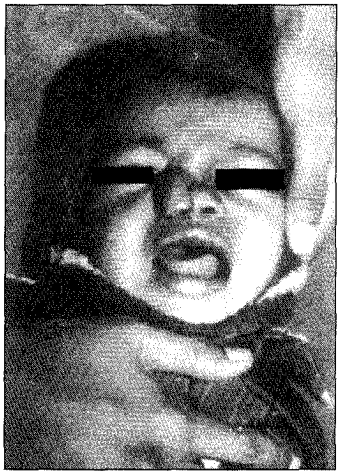

Figure 2: First stage: lower end of the proboscis inseted Age 1 year

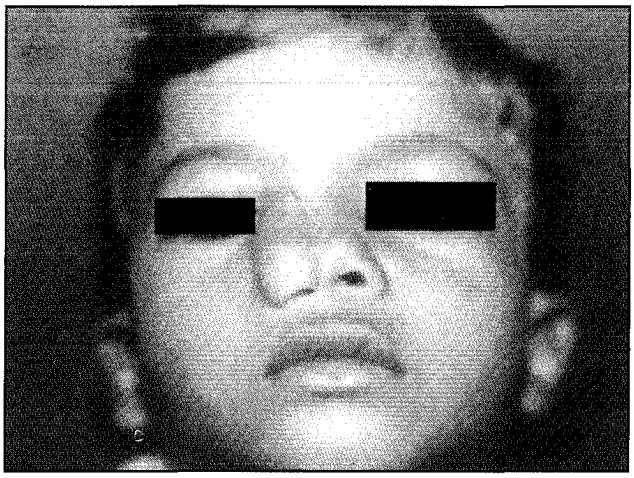

Figure 3: Second stage: upper end also inseted Age 4 years

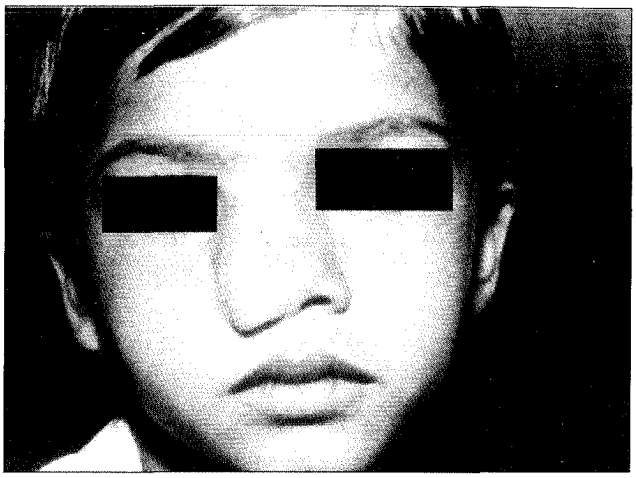

Figure 4: During follow-up Age 7 years

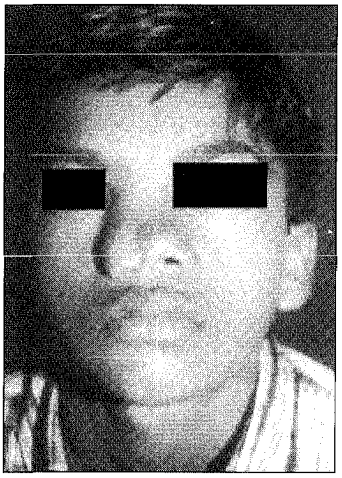

Figure 5: Follow-up at the age of 15 years Overgrown proboscis causing longer nostril on the affected side, also flat and broad dorsum is seen

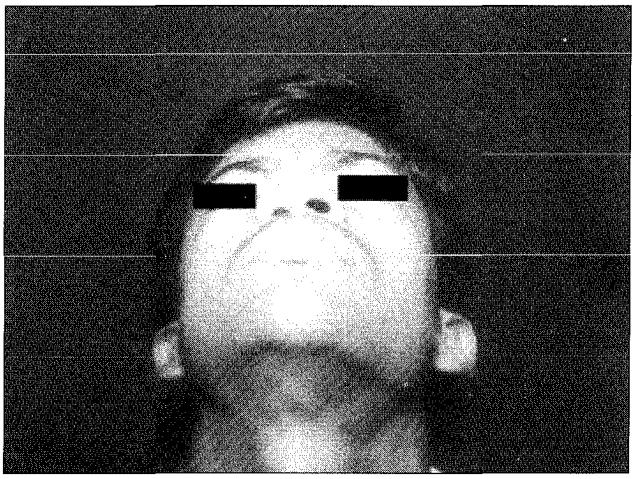

Figure 6: Recent photograph at the age of 18 years, groove between the two heminose is visible

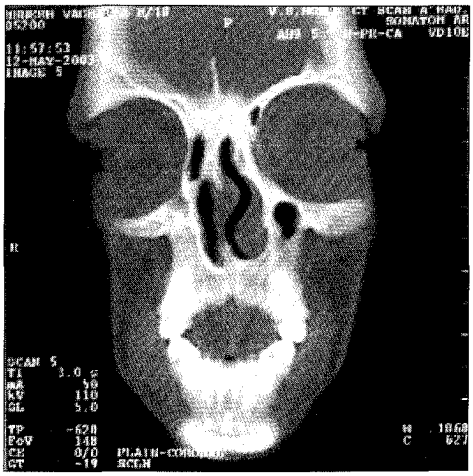

Figure 7: CT Scan: Absence of pneumatization of right maxillary, ethmoid, and sphenoid sinuses

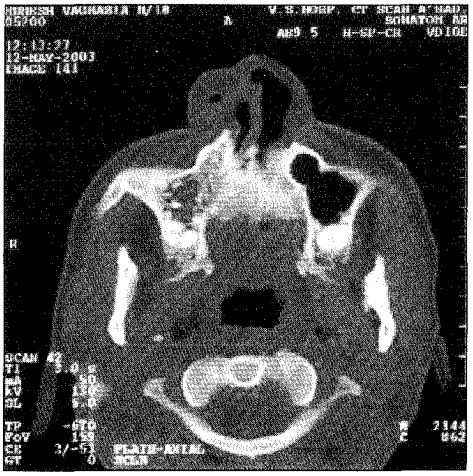

Figure 8: CT Scan: Deviated nasal septum; absent right turbinates; normal orbit, zygomatic, and frontal bones

attached it near the root of the nose (Figure 3).

At present, he needs the correction of the groove and saddle deformity (Figure 6). As the patient is now 18, we have planned aesthetic corrections in recent future. We have planned multiple Z-plasty for correction of groove. A bone graft will be required to augment the saddle deformity. We may also plan cartilaginous support to the reconstructed nose.

DISCUSSION

We would like to highlight few things about this anomaly, which others and we have observed. The proboscis is a ready tube pedicle, which can be used for absent heminose reconstruction.[1] It shows the best color and texture match with the opposite normal heminose. Many authors have suggested extirpation of the proboscis and later reconstruction with various tube pedicle flaps.[1] But this option seems very much illogical as we are losing a very good resource for heminose reconstruction. The tube has been used in various ways. Decorticated tube is passed through subcutaneous tunnel made in the region of absent heminose. Rounded and constricted appearance of the new opening is the problem with this method. It requires further corrections of nostril with z-plasty. Khoo Boo-Chai has suggested dilatation of proboscis before starting the reconstruction to avoid narrow opening and size discrepancies. But this will require repeated dilatations at regular intervals and proboscis will loose its suppleness. So we find opening the tube and attaching directly to normal heminose in staged fashion is the best option. Aesthetic correction and framework formation can be delayed till late teens when growth of nose has completed.

We have also noted that the proboscis grows with age (Figures 5 & 6). In fact in our case the proboscis has grown slightly more than normal side and it is hanging. So idea of some authors in past to start reconstruction late is not legible.

REFERENCES

1. Khoo Boo-Chai. The Proboscis lateralis- A 14 year follow-up. Plast Reconstr Surg 1985;569
2. Biber JJ. Proboscsis lateralis: A rare malformation of the nose and its genesis and treatment, J Laryngol Otol 1949;63:734.
3. Rontal M, Duritz G. Proboscis Latralis: Case report and embryologic analysis. Laryngoscope 1974;87:996.
4. D'assumpcao EA. Proboscsi lateralis. Plast Reconstr Surg 1975;55:494.
5. Tessier P. Anatomical classification of facial, cranio-facial and laterofacial clefts. Journal of Maxillofacial Surgery 1976;4:69.
6. Kirkpatrick TJ. Lateral nasal proboscis. J Laryngol Otol 1970;84:83.
7. Bishop BWF. Lateral nasal proboscis. Br J Plast Surg 1964;17:18
8. Binns JH. Congenital tubular nostril (Proboscis lateralis). Br J Plast Surg 1969;22:265.
9. Dasgupta G, Kacker K, Bhan AK. Proboscis. J Laryngol Otol 1971;85:401.
10. McLAren LR. A case of cleft lip and palate with polypoid nasal tubercle. Br J Plast Surg 1955;8:57.
11. Gupta VP, Singh RP. Unilateral absence of nose with proboscis. Indian J Plast Surg 1991;24:13.
12. Rao PB. Proboscis lateralis. J Laryngol Otol 1963;77:1028.
13. Shoujing W, Yuwen W, Hampton R. Proboscis lateralis, Microphthalmos, and Cystic degeneration of the optic nerve. Ann Ophthalmology 1983;15:756.
14. Rosen Z, Gitlin G. Proboscis lateralis. Arch Otolaryngol 1959;70:545.
15. Mahindra S, Daljit R, Jamwal. Lateral nasal proboscis, J Laryngol Otol 1971;85:963.