Proboscis Lateralis of Nose—A Case Report

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Abstract Lateral proboscis is a rare craniofacial malformation characterized by a rudimentary tubular, nose-like structure located off-center from the vertical midline of the face, and occur in association with a wide spectrum of other anomalies, including heminasal aplasia spectrum, mental retardation, callosal agenesis, heminasal hypoplasia, microphthalmia, and atypical clefting syndrome. 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. Esthetic and psychological problems are often concern of the patient and families. Here we presented a male infant of 2 months with Proboscis Lateralis. There was no e/o cleft lip or cleft palate. Staged repair was done at an early age to avoid anxiety of parents and to allow proportionate growth of nose and face.

Keywords Lateral proboscis · Heminasal proboscis · Lateral nasal proboscis

Introduction

Proboscis Lateralis is a very rare congenital anomaly characterized by a rudimentary tubular, nose-like structure most of the cases arising from the medial portion of the orbital roof [1]. Lateral proboscis is a rare anomaly, with a reported incidence to be less than 0.1:10,000 [2].

Incidence in male appears to be higher than in females, with a 3:1 male/female ratio. The reason for this sex difference is unknown [1]. The diagnosis was first described by Foster in his monograph entitled “Congenital Malformations of the Human body” in 1861. He described an 8-month-old girl with the lateral proboscis, heminasal aplasia, and cyclopean eye [3]. Later in 1884, Selenkoff reported this anomaly in the autopsy findings of a 34-year old Finnish farmer [4].

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. The classification of lateral proboscis suggested by Koo is the following: [5]

- 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.

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.

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 [5]. 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 [6].

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A Case Report

An infant male of 2 months an issue of nonconsanguineous marriage presented to us with a thick tubular structure arising from medial canthus of right eye and hypoplasia of right nostril. This was group II deformity. On examination infant was otherwise normal. There were no signs of mental delay.

Following findings were noted (Fig. 1),

- 1) A tubular structure hanging from medial canthus of right eye with dimple at tip with few drops of serosanguineous discharge.
- 2) Right heminasal hypoplasia.

Right eye was normal. No evidence of cleft lip and cleft palate. Scan was done s/o No e/o coanal atresia and soft tissue shadow attached in relation with right supraorbital margin (Fig. 2).

A staged surgical procedure was planned.

Stage I: The tubular structure was opened medially, was found to have three layers skin, subcutaneous tissue, mucosa (Fig. 3). The lateral wall of right hypoplastic nostril was opened and the proboscis was sutured with absorbable sutures (Fig. 4).

Stage II: procedure was planned after 6 months for correction of the saddle deformity at the tip of nose

Discussion

The references mention various surgeries done at later age probably to allow adequate growth of facial skeleton. However in our study early operation was done to avoid

Fig. 2 Scan showing No e/o coanal atresia and soft tissue shadow attached in relation with right supraorbital margin

anxiety of parents and also to allow proportionate growth of nose and face. Sims to be more helpful.

Proboscis is a typically obvious deformity of the facial structures, which can be diagnosed at birth or prenatally by ultrasound [7]. The treatment for lateral proboscis is surgical repair after complete facial growth. However, due to the complex nature of this malformation, achieving an esthetic result is often a challenge. A prosthetic is suitable management until final repair and reconstruction are possible. It shows the best colour and texture match with the opposite normal heminose. Many authors have suggested extirpation of the proboscis and later reconstruction with various tube pedicle flaps [5].

There are several nasal reconstruction methods using the proboscis as a donor [8]. The tube has been used in various ways. Decorticated tube is passed through subcutaneous tunnel made in the region of absent heminose. Rounded and

Fig. 1 Clinical photograph of the patient showing tubular structure hanging from medial canthus of right eye and Right heminasal hypoplasia

Fig. 3 Intraop photograph showing three layers skin, subcutaneous tissue, mucosa

Fig. 4 Postop photograph showing lateral wall of right hypoplastic nostril and the proboscis was sutured with absorbable sutures

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 lose 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.

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