HEMINASAL APLASIA : A REPORT OF TWO CASES.

Kingsly Paul,* Prema Dhanraj, ** Ashish Kumar Gupta,* Sunil Babu.******

Abstract: Heminasal aplasia is a rare congenital malformation. It is often associated with other malformations of the facial region, including abnormalities of the eye and lacrimal system, facial bone malformations and proboscis lateralis. We present two cases of heminasal aplasia, discuss their embryological basis and nasal reconstruction of the same.

Keywords: heminasal aplasia; proboscis lateralis; reconstruction

INTRODUCTION:

Heminasal aplasia is a very rare congenital anomaly presenting with varied appearances. We present two case reports, one of which was associated with lateral nasal proboscis.

CASE HISTORY :

Case: 1 : A five-month-old male child was brought to our clinic with multiple congenital anomalies of the face. He was the first child in the family, born of second-degree consanguineous marriage. There was no complication during pregnancy. Examination showed a complete cleft of the left side of upper lip and cleft of palate. Right nostril and right nasal cavity was present and normal. Left nostril was absent. There was total absence of left side of external nose. There was a fleshy cylindrical mass seen attached near the medial canthus of the left eye. Lacrimal duct was absent on the left side (Photo 1). Examination of the eye showed choroidal coloboma with tilted disc, optic pit and optic disc coloboma. Right eye was normal. Both ears were normal. CT scan of the face showed hypoplastic left maxillary sinus, significant narrowing of left nasal cavity, left sided deviated bony nasal septum and soft tissue mass seen over the left fronto nasal region. Ethmoidal sinuses were absent on the left side. Sphenoid sinus and frontal sinus were not pneumatized.

The child underwent cleft lip repair at five months of age. Palatoplasty was done at fourteen months of age. At the age of two years, the proboscis was used to reconstruct the left half of

the external nose. Left nostril was also created and communication established with the left nasal cavity and maintained for a month with a hollow splint. At the age of seven years he presented with a minimal swelling of the proboscis tissue. Debulking of the tissues over the nasal dorsum and z-plasty at the left nasolabial region was done. (Photo 2)

Case: 2 : A twelve-year-old boy presented to our clinic with total absence of left side of the nose. He had microphthalmia, opaque cornea and dacryocystitis on the left side (Photo 3). X-ray of the paranasal sinuses showed narrow left nasal cavity and hypoplastic left maxillary sinus. Left side of the nose was reconstructed with midline forehead flap, and flap division and inset done after three weeks. Nasal dorsal augmentation was done one year later with an iliac bone graft. He underwent scar revision and z-plasty of the nasal tip in the subsequent stages (Photo 4).

DISCUSSION:

The development of the nose starts around the twenty-eighth embryological day and originates in the bilateral nasal placodes. The nasal placodes invaginate to form the nasal pits which are widely spaced on the anterolateral sides of the developing head of the embryo. Around the nasal pit on either side, three processes grow out: the medial nasal, lateral nasal and the maxillary processes. The medial nasal processes fuse together to form the ridge, tip and columella of the nose, the philtrum and the medial part of the upper lip. The maxillary processes fuse with the medial nasal processes and separates the nasal and oral cavities. The nasal pit invaginates further and breaks through the oral cavity. Absence of one of the placodes leads to heminasal aplasia.¹

Mazzola has divided congenital fronto nasal malformations according to the area of their appearance: the upper face, midcephalic borderline, and lower lateral face region. Nasal aplasia belongs to the first group and is divided into three subgroups: arhinia (Total aplasia of the nose), half nose (Heminasal aplasia) and half nose with proboscis lateralis²

Our first case belonged to the third subgroup, and the second case to the second subgroup. The maxillary process acts as a key stone between the nose, the eyes and the ears. Failure of maxillary growth leads to a bifid nose, or proboscis lateralis, cleft lip and cleft alveolus³. Bremer believes the proboscis is formed from the lateral and medial nasal processes⁴.

Case 1: showing the accessory nose

Case I: Post-operative photograph

Case II: Pre-operative photograph

Case II: Post-operative photograph

Several reconstructive methods are described using the proboscis as a donor, which gives the ideal texture and color^{5,6,7,8}. The proboscis may be split ventromedially to attach on the raw surface created on the left side of the nose, or it may be deepithelialised and passed down along a subcutaneous tunnel⁹. When the proboscis is absent, forehead flap is a good alternative for the nose reconstruction. To achieve symmetry and contour, further procedures including insertion of bone graft, z-plasty and debulking may be performed. Suitable cases like ours may necessitate creation of the nasal passages when a nasal cavity is present. The reconstructive modality must be individualized for each patient depending upon the associated abnormalities.

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Address for correspondence

Dr. Kingsly Paul, M., M.S. (ENT), M.Ch. (Plast),
Senior Lecturer,
Department of Plastic Surgery,
Christian Medical College,
Vellore, India.
Email: kingslypaul@gmail.com