Proboscis Lateralis

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Abstract

Presented here is a rare case of Proboscis Lateralis. The development of such a malformation in terms of embryological classification of the body is highlighted. The difficulty encountered in reconstruction with a small proboscis is discussed.

Key Words

Nose, abnormalities; Abnormalities, multiple; Proboscis lateralis.

The Proboscis Lateralis is a rare malformation which was reported for the first time in 1861 by Forster in his monograph on Congenital Malformations of the human body. This condition has been described to manifest either all alone with a normal nose or in association with the ipsilateral nasal deformities like heminasal aplasia or arrhinia and sometimes with the eye deformities like coloboma eyelid (upper and lower), microphthalmia, or with other facial deformities like cleft palate and cleft lip (Khoo Boo-Chai, 1985). Invariably, it is a trunk like appendage attached to anteromedial portion of the orbital roof.

Case Report

A 5 months old boy born as a second child to a perfectly healthy parents, presented in our outpatients department with the presence of trunk like appendage near left medial canthus for which the mother was very apprehensive. There was no consanguinity amongst parents and during the confinement the mother did not have any fever or infection and did not take any teratogenic drug. The first child was normal.

On examination the child was moderately nourished and had normal milestones. A hollow trunk like appendage of elastic consistency measuring 1.5 cm in length and about 1.5cm in diameter was seen arising 1 cm medial and above the medial canthus of the left eye showing no increase in size with the cry. There was some mucopurulent discharge from its distal end since 2 weeks after birth. A probe passed through the opening could not pass beyond the root of the trunk and the radiological dye study done on it failed to show any communication with cranial cavity or the nasal

cavity. Dacryocystography also did not show any communication of lacrimal apparatus with the proboscis.

Besides, there was absence of nares and nasal cavity on left side. and there existed coloboma of left eyelid

Fig. 1 Shows the Lateral proboscis with heminasal aplasia on the left side.

and iris and the absence of lacrimal sac on the left side. There was no other abnormal opening or groove on the affected side of the face. Nasal bone was found to be deficient on left side on palpation and on X-ray. There was no associated cleft lip or palate. The philtrum was normally developed.

Due to the small size and higher attachment of the proboscis in our case, the reconstruction of the left heminose using the proboscis as a donor material was not possible. Moreover, there was recurrent development of soft cystic swelling medial to the left medial canthus post-operatively which was due to the inadequate drainage of the discharge from the mucus lining of the malformation and also the paranasal sinuses. In our case we had to resort to the perforation of the nasal septum to overcome this recurrent problem. The case is still under serial reconstruction and follow-up after 4 years.

Discussion

The malformations developing in the first embryonal ~~month are usually~~ not visible whereas the regional malformations of second and third embryonal months are visible and are of surgical interest. In the region of head, they can involve prosencephalic, diacephalic, or rhombencephalic region. As per embryological classification of body applied to the human face, prosencephalic centre is responsible for the development of the dorsum nose, central part of upper lip, pre maxilla and the nasal septum. And the diacephalic border is responsible for the eyes, alae nasi and philtrum. Due to intensive metabolic needs and complicated morphologic process the embryonal tissue are very susceptible to the malformations and if this happens, it follows a 'malformation programme' as the malformation develops in specific embryonal phase (Pfeifer, 1991). The eyes can be affected in malformations that are primarily located in the prosencephalic region or diacephalic border.

The two symmetric proboscis may exist as separate appendages or form two separate halves of the nose. However, in asymmetric proboscis one may be missing thereby producing heminasal aplasia, or it may form

an appendage and heminasal aplasia on one side while the other half of nose is developed as seen in the present case.

As there is associated abnormality of the ipsilateral eye and the absence of side wall of ipsilateral nostril in many cases, it was suggested by Biber (1949) and Rao (1963) that the material for the proboscis is derived from the lateral nasal process. The actual cause for such a malformation is not known as yet but it seems to be due to some exogenic cause like traumatic noxious injury.

All the published cases so far revealed this appendage to be a blind trunk ending at the bone except for one case reported in 1906 by Kirchamyr, where he found it reaching dura. Therefore, the management of such cases is basically the reconstruction of the nose, preferably by using the proboscis as a donor material. The communication to the nasal cavity which is not completely developed is obtained by incising the different layers on the malformed lateral nasal wall. Then the proboscis is incised and sutured on to the lateral wall parallel to the bridge, thus forming the alae and lateral nasal wall.

The age for such a reconstruction is not fixed. It is considered by some workers that the reconstructive operation on such cases should be deferred till maturity as there is danger of the difference in the rate of growth of normal and operated side and also because the dimensions of the normal side and the proboscis are permanently established by maturity. However, in practical terms it is not always feasible to wait that long as was the case with Biber (1949) and also with us, because of the insistence from the parents to get the outgrowth removed immediately, Khoo Boo-Chai (1985) considered it reasonably safe to begin reconstruction of the heminose in infancy as he feels that the proboscis grows harmoniously with the normal side and the rest of the face. But, with the problems encountered in our case who had the proboscis of a short length, it is better to wait for the proboscis to grow and the face to develop before starting the reconstructive surgeries.

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