UNILATERAL ABSENCE OF NOSE WITH LATERAL PROBOSCIS
(A CASE REPORT)

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SUMMARY

An extremely rare congenital malformation in a male child having complete absence of the nose on the right side with unilateral proboscis arising from the supracanthal region on the same side is being reported here for the purpose of documentation in the literature. The morphology and embryology of this malformation has also been discussed.

This is an extremely rare, congenital malformation. So far only very few cases have been reported in the literature (Converse 1977, Barsky 1979). The absence of nose can be partial or total. The condition may be associated with unilateral arrhinencephaly or unilateral proboscis.

Case History

A three years old male child, 3rd amongst the siblings was born full term at home, without any antenatal problems. No other child in the family has ever had any other known congenital deformity either on the maternal or paternal side.

On local examination, the child had a normal well developed nostril on the left side. The right half of the nose was completely absent. It was replaced by a tubular snout like structure attached to the right side of the nasal bridge just above the medial canthus (Fig. 1). The length of the proboscis was almost equal to the length of the normal nose. Its free end had a blind track measuring 1.5-2 cm in length. A cartilage like structure was felt in the distal part of the proboscis. The nasal pyramid was less prominent, the nose bridge was flat and depressed. The nasal septum was present. The child had almost normal breathing through the existing nose on the left side. There was epiphora on the right side, due to the absence of the right naso-lacrimal duct.

Embryology

The nose develops from the fronto-nasal process, which divides into a medial and a lateral nasal process at about the 3rd-4th week.

Fig. 1. Pre-operative photograph showing absence of right nose with lateral proboscis.
of the intrauterine life. The nasal placodes, an epithelial thickening, appearing on either side of the fronto-nasal process deepens and forms the nasal cavity. The final fusion of the medial, lateral nasal processes and the maxillary processes forms the anterior nares.

Discussion

The unilateral or bilateral absence of nose is a rare type of congenital defect. The arrest of the growth of the lateral nasal process is due to aplasia of its vascular network (Mustarde, 1979). The lateral proboscis could represent a morphological deviation of the fronto-nasal process under a noxious stimulus. The hemiabscence of nose with lateral proboscis is described as "Dystopia". The authors are of the opinion that lateral proboscis could be a result of nonfusion of lateral nasal process to medial nasal process and maxillary process. The shape of the proboscis appears to be like a distorted nose having the primitive residual structure of nose in it. These findings have also been confirmed by Routal and Duritz (1977).

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