Case Reports

Unusual case of accessory nose associated with unilateral complete congenital choanal atresia

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Case Report. A 2-month-old child was referred to Assadar Teaching Hospital, Najaf, Iraq with congenital anomaly of the face. The child was first born to a second-degree relative marriage, and was delivered normally, and uneventfully. Examination showed that there is a 2 cm cylindrical shape fleshy mass projecting from the medial to the medial canthus of the right eye, which in turn normal with no evidence of coloboma, or vision abnormalities, and both ears were normal. Further, examination showed that the child has unilateral complete choanal atresia on the same side of the mass with asymmetry in the size of external nostril as could be seen in Figure 1. No other cross congenital anomalies were detected. The baby underwent simple surgical excision of the mass, and the wound was closed primarily without flap repair. The conventional endoscopic repair of congenital choanal atresia was carried out as well. An accessory nose is an extremely rare congenital anomaly. It had been reported twice in the medical (English) literature since the 1960’s. In this article, we described this anomaly, and reviewed international literature for similar cases. A brief embryological classification was explained as well. We think that reporting such extreme rarity of cases is worthwhile to enrich medical literature.

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 that are widely spaced on the anterolateral sides of the developing head of the embryo. Around the nasal pit on either side, 3 processes grow out; the medial nasal, lateral nasal, and the maxillary processes. The medial nasal processes fuse together to form the ridge, tip and columnella 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. The absence of one of the placodes leads to heminasal aplasia. Based on previous study, it is possible to categorize nasal deformities into duplications and dysplasias. Nasal duplication as proposed by Erich is...
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basically a complete formation of 2 sets of nostrils (2 septae, 4 nostrils, and 4 nasal cavities). He proposed the theory of dichotomy by atavism to explain the formation of a double nose. Dysplasias, on the other hand, covers a wide range of deformities as seen in the Van der Meulen's classification, and as cited by de Blécourt et al. Type I nasal dysplasia is characterized by aplasia with proboscis. Type III nasal dysplasia (nasoschisis) also known as Tessier cleft number one or lateral nasal clefts. Type IV nasal dysplasia covers the entire range of duplications. This malformation ranges from a supernumerary nostril to a complete duplication of the nose, and upper face, which is called diprosopia. Using the above classification, our case would be type IV nasal dysplasia. Supernumerary nostrils are uncommon both unilateral, and bilateral. Variants have been described however, on thorough reviewing of English literatures, we were only able to identify 2 cases. A very similar case was reported in Turkey with unilateral incomplete cleft palate. Accessory nose is extremely rare case, and only one or 2 cases have been reported in the English literature during the last 50 years.

References