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

Sajad Al-Helo, DLO, FICMS, *Ali S. Taghi,* DOHNS, MRCS, *Saad Mahboba,* DLO.

ABSTRACT

نستعرض في هذا المقال حالة غير اعتيادية ونادرة من حالات التشوه الولادي للأنف. حيث سجلنا حالة مضاعفة الأنف ومتلازمة انسداد المنخر الأنفي من الخلف. على الرغم من أن التشوهات الخلقية في الأنف ممكنة الحدوث إلا أن حالات مضاعفة الأنف تعتبر من الحالات النادرة جداً، وتستحق التبليغ عنها نظراً لندرتها.

We report here an extremely rare and unusual case of accessory nose associated with unilateral complete congenital choanal atresia. Although other types of nasal congenital nasal anomalies are not rare, however, extreme rarity of nose duplication is worth reporting, as it is an extremely rare type of nasal congenital deformity.

Saudi Med J 2008; Vol. 29 (9): 1342-1343

From the Department of Otolaryngology, Head and Neck Surgery, (Al-Helo, Mahboba), Assadar Teaching Hospital, Najaf, Iraq, and the Department of Otolaryngology, Head and Neck Surgery (Taghi), Gwynedd Hospital, North Wales, United Kingdom.

Received 15th April 2008. Accepted 3rd August 2008.

Address correspondence and reprint request to: Dr. Ali S. Taghi, 16 Saunderton Road, Wembley, Middlesex, HA0 2NE, United Kingdom. Tel. +1 (44) 7940187684. E-mail: taghi72@doctors.org.uk

Congenital anomalies of the face and nose are not rare in essence, but cases of accessory nose are of extreme rarity. Nasal dysplasia covers wide range of nose duplications, and these could range from a supernumerary nostril to a complete duplication of the nose (accessory nose). We report here an extremely rare and unusual case of accessory nose associated with unilateral complete congenital choanal atresia. Although other types of nasal congenital nasal anomalies are not rare, however, extreme rarity of nose duplication is worth reporting, as it is an extremely rare type of nasal congenital deformity.

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 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. The absence of one of the placodes leads to heminasal aplasia.[1] Based on previous study, it is possible to categorize nasal deformities into duplications and dysplasias. Nasal duplication as proposed by Erich[2] is

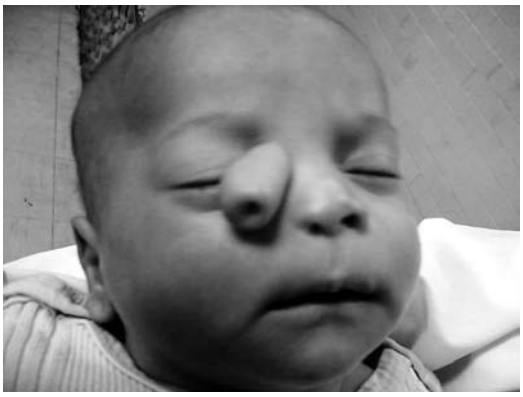

Figure 1 - The mass which is projected from the area just medial to the medial right canthus. Note the asymmetry in the nostril.

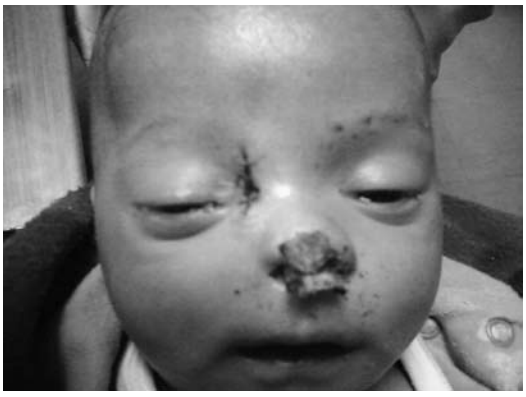

Figure 2 - Post-operative view, the mass excised simply with primary wound closure.

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[3] classification, and as cited by de Blécourt et al.[4] The classification is extensive and covers most deformities. They distinguished 4 types of nasal dysplasias. Type I is a nasal aplasia in which one nasal half is absent. This is frequently associated with other malformations such as cleft lip and palate, microphthalmia, and colobomata of the iris, and the eyelids. Type II 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 [4-9] however, on thorough reviewing of English literatures, we were only able to identified 2 cases. A very similar case was reported in Turkey with unilateral incomplete cleft palate.[10,11] 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

1. Mladina R, Manojlovic S, Markov-Glavas D, Subaric M. An Unusual Case of Heminasal Aplasia: Proboscis Lateralis or Cystic Teratoma? ***J Craniofac Surg*** 2003; 14: 41-45.
2. Erich JB. Nasal duplication. Report of case of patient with two noses. ***Plast Reconstr Surg Transplant Bull*** 1962; 29: 159-166.
3. van der Meulen JC. Classification of craniofacial malformations. In: Stricker M, van der Meulen, Raphael, Mazzola R. editors. Craniofacial malformations. Edinburgh (UK): Churchill Livingstone; 1990. p. 187-199.
4. Blecourt RA de, Roddi R, Berg JP, Bloem JJ. Nasal dysplasia. ***Indian J Plast Surg*** 1996; 28: 39-41.
5. Chen MT, Yeong EK. Supernumerary nostrils. ***Br J Plast Surg*** 1992; 45: 557-558.
6. Kawamoto HK Jr. Rare craniofacial clefts. In: McCarthy JG. editor. Plastic surgery. vol 4. Philadelphia (PA): Saunders; 1990. p. 2947.
7. Jian XC, Su HB, Chen XO. Triple nostrils associated with cleft lip and palate. ***Br J Plast Surg*** 1995; 48: 53.
8. Nakamura K, Onizuka T. A case of supernumerary nostril. ***Plast Reconstr Surg*** 1978; 80: 436-441.
9. Onizuka T, Tai Y. Supernumerary nostril: case report. ***Plast Reconstr Surg*** 1972; 50: 403-405.
10. Kurul S. Ozcan G. Cakalir C. Accessory nose associated with unilateral incomplete cleft lip. ***Eur Arch Otorhinolaryngol*** 1995; 252: 61-62.
11. Ghosh P, Dada K, Gupta S, Jamwal N, Daljit R. Double nose. ***J Laryngol Otol*** 1971; 85: 963-969.