UNUSUAL CASE OF ACCESSORY NOSE ASSOCIATED WITH UNILATERAL COMPLETE CONGENITAL CHOANAL ATRESIA

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ABSTRACT

We describe here a very unusual case of 2 weeks Iraqi boy who was referred to our Otolaryngology department at the Assadar Teaching Hospital in Al - Najaf governorate in Iraq with history of cylindrical shaped mass projecting from area just medial to the medial canthus of the right eye. The mass was found since his birth and parents confirmed that the mass kept growing as the child grows.

Clinical and histological examination showed that the mass structure is consistent with an accessory nose. No other congenital abnormalities were detected apart from complete mixed bony- membranous choanal atresia on the same side of the mass in which contemporary treated with simple mass surgical excision and closure.

We regret that CT scan pictures are not available because the system was down during the surgery time.

KEYWORDS

Accessory nose, congenital choanal atresia, congenital anomalies, congenital nasal abnormality

INTRODUCTION:

Congenital deformities of the nose are rare, but we are describing here an extremely rare case of accessory nose associated with complete choanal atresia on the same side. This is the third case reported in English literature as far as we know. Diagnosis was based on clinical appearance and histological picture.

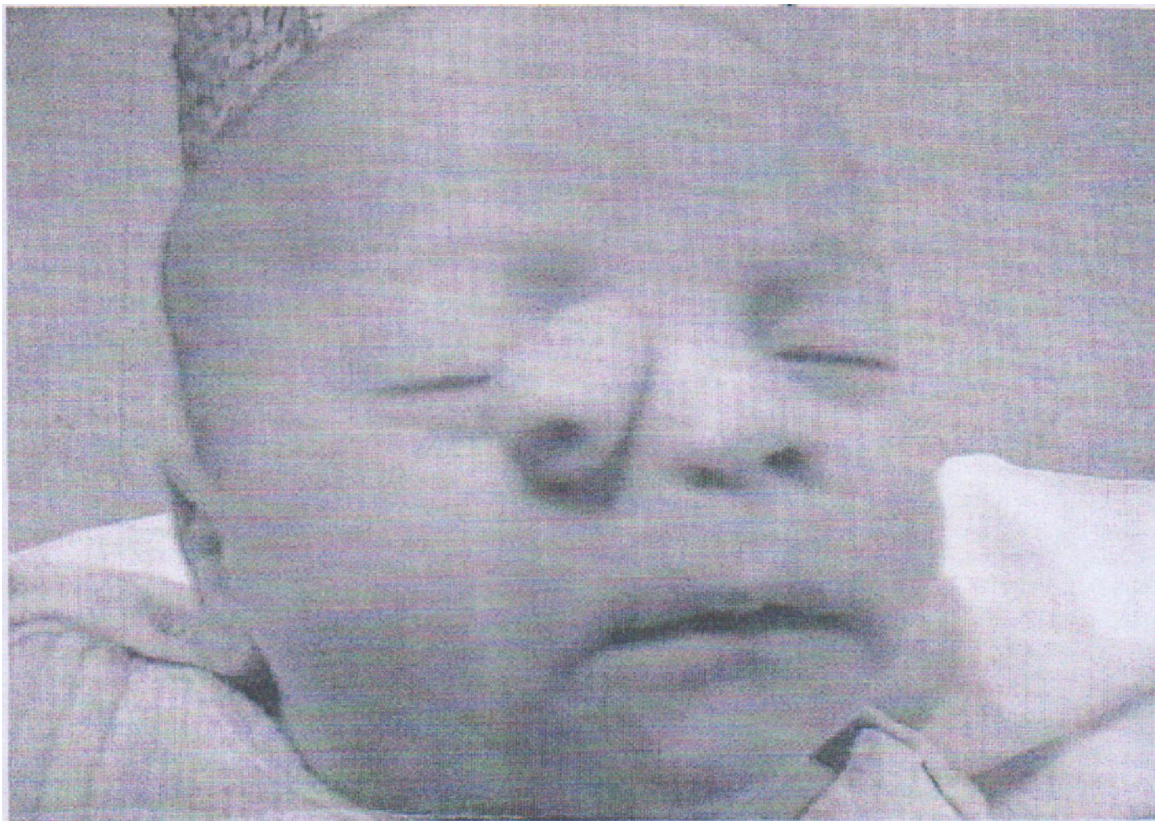
CASE HISTORY:

2 weeks old child referred to Assadar teaching hospital with congenital anomaly of the face. Child was first to born of second-degree relative marriage. Baby delivered normally and uneventfully. Examination showed that there is a 2 cm cylindrical shape fleshy mass projecting just medial to the medial canthus of the right eye, which in turn normal with no evidence of any coloboma of vision abnormalities and both ears were normal clinically and audiologically.

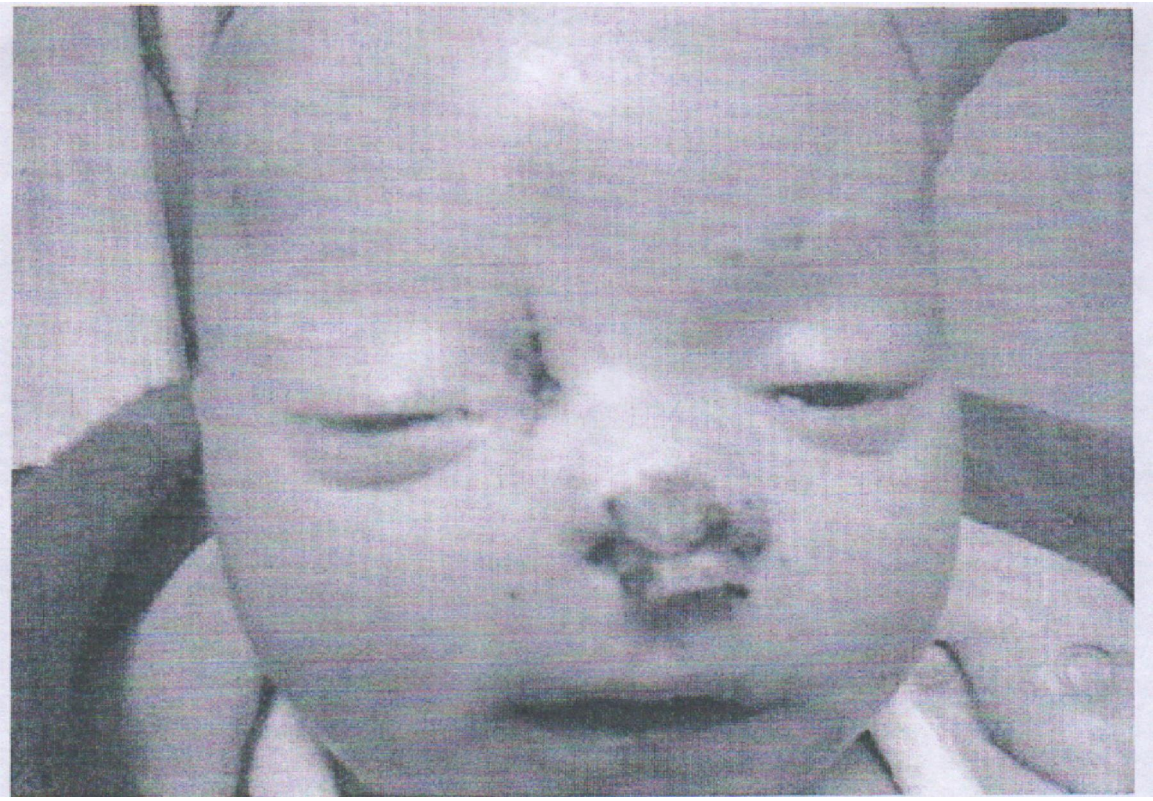
Further ENT examination showed that the child has unilateral complete choanal atresia on the same side of the mass with asymmetry in the size of external nostril as can been seen in Fig 1.

No other cross congenital anomalies were detected apart from the above mentioned.

Child underwent simple excision of the mass and conventional endoscopic repair of the congenital choanal atresia.



Figure(1)



Figure(2)

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, 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]

Based on available literature, it's possible to categorize nasal deformities into *duplications* and *dysplasias*

Nasal duplication as proposed by Erich [2] , is basically a complete formation of two sets of nostrils (two septae, four nostrils, and four nasal cavities). He proposed the theory of dichotomy by atavism to explain the formation of a double nose.

Dysplasias, on the other hand, cover a wide range of deformities. The van der Meulen classification [3] **and** as cited by de Blécourt et al[4]

It's extensive and covers most deformities. They distinguished four types of nasal dysplasias.

Type I is 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 1 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 this classification, our case would be a type IV nasal dysplasia

Supernumerary nostrils are uncommon. Both unilateral and bilateral variants have been described [4, 5, 6, 7, 8, 9, 10, and 11] but on thorough reviewing of English literature we were only able to identified two cases. A very similar case was reported in Turkey with unilateral incomplete cleft palate [12, 13, 14]

CONCLUSION:

An interesting case of an accessory nose with complete unilateral congenital choanal atresia is presented. Similar cases are reviewed. A simple surgical technique for correction of the accessory nostril is also described. Accessory nose is extremely rare case and only one case has reported in literature.

REFERENCE:

1.◌̲◌̲ Mladina R, Manojlovic S, Markov-Glavas D, Subaric M. Anunusual case of heminasal aplasia: proboscis lateralis or cystic teratoma? J Craniofac Surg. 2003 Jan;14(1):41-5.9.

2.Erich JB (1962) Nasal duplication: report of a case of a patient with two noses. Plast Reconstr Surg 29:159

3.Meulen JC van der (1990) Classification of craniofacial malformations. In: Stricker M, van der Meulen JC, Raphael B, et al (eds) Craniofacial malformations. Churchill Livingstone, Edinburgh, pp187–199

4.Blécourt RA de, Roddi R, Berg JP, Bloem JJ (1996) Nasal dysplasia. Ann Plast Surg 37:633–637

5.Chen MT, Yeong EK (1992) Supernumerary nostrils. Br J Plast Surg 45:557–558

6.Kawamoto HK Jr (1990) Rare craniofacial clefts. In: McCarthy JG (ed) Plastic surgery, vol 4. Saunders, Philadelphia, p 2947

7.Jian XC, Su HB, Chen XO (1995) Triple nostrils associated with cleft lip and palate. Br J Plast Surg 48:53–55

8.Nakamura. K, Onizuka T (1987) A case of supernumerary nostril. Plast Reconstr Surg 80:436–441

9.Onizuka T, Tai Y (1972) Supernumerary nostril: case report. Plast Reconstr Surg 50:403–405

10.Reddy KA, Rao AK (1987) Triple nostrils: a case report and review. Br J Plast Surg 40:651–652

11.Sharma LK (1975) Supernumerary nostril. Indian J Plast Surg 8:90–92

12.Kurul S. Ozcan G. Cakalir C. Accessory nose associated with unilateral incomplete cleft lip. [Case Reports. Journal Article] *European Archives of Oto-Rhino-Laryngology. 252(1):61-2, 1995.*

13.Ghosh P. Dada K. Gupta S. Jamwal N. Daljit R. Double nose. [Journal Article] *Journal of Laryngology & Otology. 85(9):963-9, 1971 Sep.*

14.Williams A. Pizzuto M. Brodsky L. Perry R. Supernumerary nostril: a rare congenital deformity.[CaseReports.JournalArticle] *International Journal of Pediatric Otorhinolaryngology. 44(2):161-7, 1998 Jul 10*