Supernumerary nostril: Congenital adrenal hyperplasia with a rare congenital anomaly

Access this article online
Website: www.amsjournal.com
DOI: 10.4103/2231-0746.147133
Quick Response Code:

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ABSTRACT

Multiple or supernumerary nostril is a rare congenital anomaly with unknown etiology. The first case was reported by Lindsay as bilateral supernumerary nostrils. Supernumerary nostril cases are mostly unilateral and isolated. They are also reported with other congenital malformations like facial clefts and congenital anomalies like congenital auricular hypoplasia, congenital cataracts, eusophageal atresia and patent ductus arteriosus. Here, we report a case of supernumerary nostril with congenital adrenal hyperplasia

Keywords: Congenital adrenal hyperplasia, nostril, supernumerary

INTRODUCTION

Supernumerary nostril is one of the rarest congenital nasal deformities.[1] Supernumerary nostril cases are mostly unilateral and isolated. They are also reported with other congenital malformations such as facial clefts and congenital anomalies like congenital auricular hypoplasia, congenital cataracts, esophageal atresia and patent ductus arteriosus (PDA).[2,3] No case of an supernumerary nostril and congenital adrenal hyperplasia co-existence has been reported previously in English literature. Here, we report a case of supernumerary nostril with congenital adrenal hyperplasia.

CASE REPORT

A 4-year-old female patient with a nasal anomaly was admitted to our outpatient clinic. She was born 2300 g prematurely on her 37th gestational week from a 34-year-old mother with a history of five pregnancies, two still births and two abortions. On her physical examination, she had a left-sided supernumerary nostril and cliteromegaly [Figure 1]. Her laboratory studies revealed low levels of androstenedione and her history showed that she had medical therapy for 1 year for her congenital adrenal hyperplasia. No additional anomaly except patent foramen ovale was detected in her work-ups. Paranasal sinus computed tomography study revealed that her left-sided accessory nostril opened to the left nasal cavity. The patient was operated, the opening of the supernumerary nostril to the nasal cavity was obliterated and the widened nostril was narrowed by excisions from the alar ground and lateral side [Figure 2]. A revision toward the nostril asymmetry is planned 1 year after the operation.

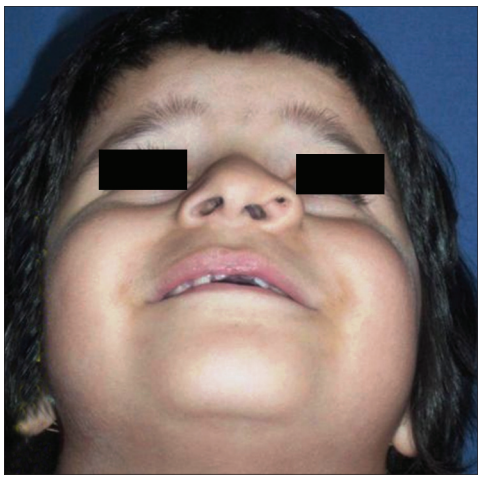

Figure 1: Preoperative view of the case

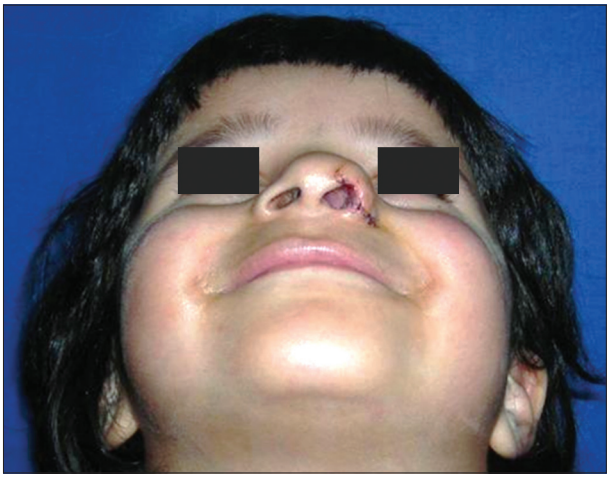

Figure 2: Perioperative view of the case

DISCUSSION

Multiple or supernumerary nostril is a rare congenital anomaly with unknown etiology.[1] The first case was reported by Lindsay as bilateral accessory nostrils. In this case, the accessory nostrils were placed over the normal nasal cavity and a relation between the accessory nasal cavity and the normal nasal cavity was observed. Tawse described the case of unilateral accessory nostril associated with the normal nasal cavity in 1920 and the third case was reported by Simonetta in 1936. A case associated with microcornea was reported in 2005 and another one together with esophagus atresia, imperforated anus and PDA was reported in 2009.[1-5] Several cases, which are not connected with the unilateral nasal cavity have also been reported. Duplication anomalies of the nose include polyhinia and supernumerary nostril. Both situations are congenital disorders arising from aberrant embryologic development. Differential diagnosis includes nasal glioma, encephalocele, meningocele, meningomyelocele, nasal dermoid, nasolacrimal canal duplication and midline defects. Supernumerary nostril cases always have two normal nostrils.[2] Even though the specific reason and development of the accessory nostril remains unknown, it is thought to be the result of the cleavage in the lateral nasal bud in early embryonic life.[1,2] Other congenital anomalies associated with accessory nostril include hypoplastic nose, cleft lip and nose, nasal-ocular cleft, congenital auricular hypoplasia, congenital cataracts, esophageal atresia, PDA and imperforated anus. In a recent study, Aslanabadi *et al*. have reviewed the 31 cases in the literature and reported that 12 of the accessory nostril cases (40%) were left-sided, 10 of them (33.3%) were right-sided and five cases (16.7%) bilateral, two cases were columellar origin. There was one case with no data.[2]

CONCLUSION

Supernumerary nostril is a very rare condition of unknown etiology. The possible existence of congenital adrenal hyperplasia must be kept in mind in patients with accessory nostril. Furthermore, timing of the surgery is as important as the planning of the incision, because excision performed at an early age avoids any serious impact on the nasal cartilages due to the fistula, deformation of the adjacent structures also undesirable psychological effects.

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Cite this article as: Ciloglu S, Duran A, Buyukdogan H, Yigit AK. Supernumerary nostril: Congenital adrenal hyperplasia with a rare congenital anomaly. Ann Maxillofac Surg 2014;4:193-4.

Source of Support: Nil, **Conflict of Interest**: None declared.