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Supernumerary Nostril in a 15-Year-Old Girl

ABSTRACT

Objectives: To describe a rare case of a supernumerary nostril and its management and to discuss the different theories pertaining to this type of deformity.

Methods:

Design: Case Report
Setting: Tertiary Government Hospital
Patient: One

Results: A 15-year-old girl was born with one extra nasal opening. Nasal endoscopy after 15 years revealed a mucosa-lined cavity that ended blindly. Computed tomography scan showed a small opening lateral to the right nasal ala which did not communicate with the right nasal cavity. A crescent incision was made along the inner circumference of the supernumerary nostril and the blind ending nasal tract was excised. A portion of the ala was removed and a Z-plasty repair was performed.

Conclusion: Treatment has to be tailored as to the presentation of a supernumerary nostril and any other associated deformity. Though various surgical interventions exist, the goal of repair is to create a functioning and normal-appearing nose.

Keywords: *Supernumerary Nostril, Accessory Nostril*

A supernumerary nostril is a congenital malformation characterized by an accessory nasal cavity that usually manifests as a small nasal orifice with surrounding redundant soft tissue.[1] The accessory nostril is similar to a normal nostril which has vibrissae and an intranasal opening, can be unilateral or bilateral and may have a communication with the ipsilateral nostril.[2] The first reported case was published in 1906 by Lindsay who described a patient with bilateral supernumerary nostrils.[2] In the English literature, there have been about 40 reported cases of supernumerary nostrils.[2-12] To the best of our knowledge based on a search of HERDIN using the keywords "supernumerary" "accessory" and " nostril," there has been no locally-reported case of a supernumerary nostril.

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We describe a rare case of a supernumerary nostril and its management and discuss the different theories pertaining to this type of deformity.

CASE REPORT

A 15-year-old girl presented to the Ear Nose Throat (ENT) outpatient department due to an accessory nostril on the right side of her nose since birth. *(Figure 1)* There was no history of bleeding or discharge (although she reported recurrent yellowish discharge from the other nostrils). She had no previous medical consultations due to financial constraints. She was vaginally born to a then 22-year-old gravida 1 mother. The prenatal history was unremarkable with no history of exposure to alcohol, drugs or ionizing radiation. There was no history of consanguineous marriage or family history of birth defects and the antenatal and neonatal history were also uneventful.

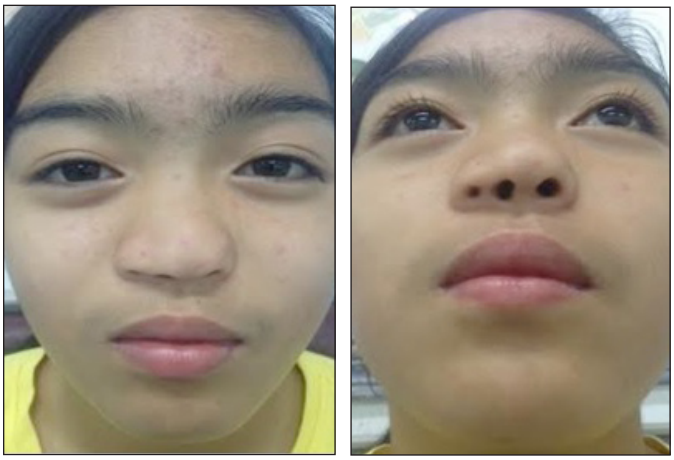

Figure 1. Preoperative frontal and basal view (published in full with permission)

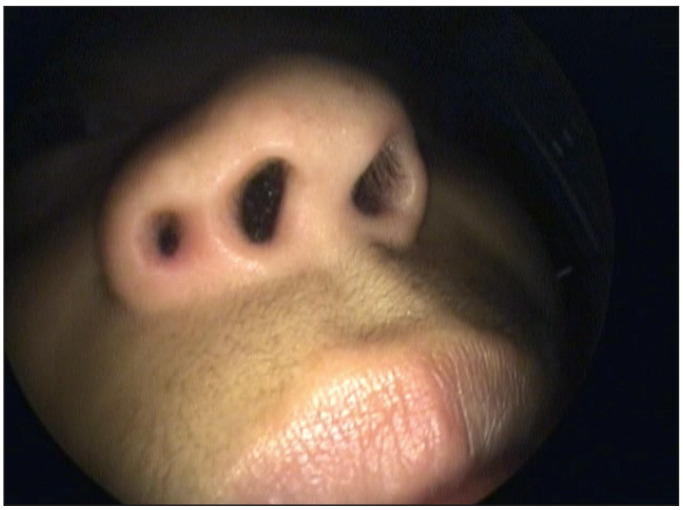

Figure 2. Small blind mucous membrane-lined pocket with vibrissae located lateral to and at the same level as the right nostril

On physical examination, the left nasal opening was wider compared to the right with two nasal openings. The smaller supernumerary nostril had an internal diameter of about 5 mm located lateral to and at the same level as the right nostril. *(Figure 2)* Anterior rhinoscopy revealed that the accessory nostril was lined with hair follicles. The septum between the right and left nasal cavities was midline. The inferior turbinates were not congested. However, there was yellowish discharge in both right and left nostrils. Nasal endoscopy revealed no communication between the normal and accessory nasal cavity which ended blindly and was lined by mucous membrane. A polypoid mass was noted within the left middle meatus.

Computed tomography scans of the paranasal sinuses revealed soft tissue thickening of the right nasal ala with a small opening laterally. There were no bony or cartilaginous abnormalities. There were opacifications seen in the left frontal sinus, left anterior ethmoid, both maxillary sinuses and mucosal thickening of the right sphenoid sinus. *(Figure 3)*

The patient was admitted with an impression of right unilateral supernumerary nostril and chronic rhinosinusitis with left nasal polyp and underwent surgery under general anesthesia. A crescent incision

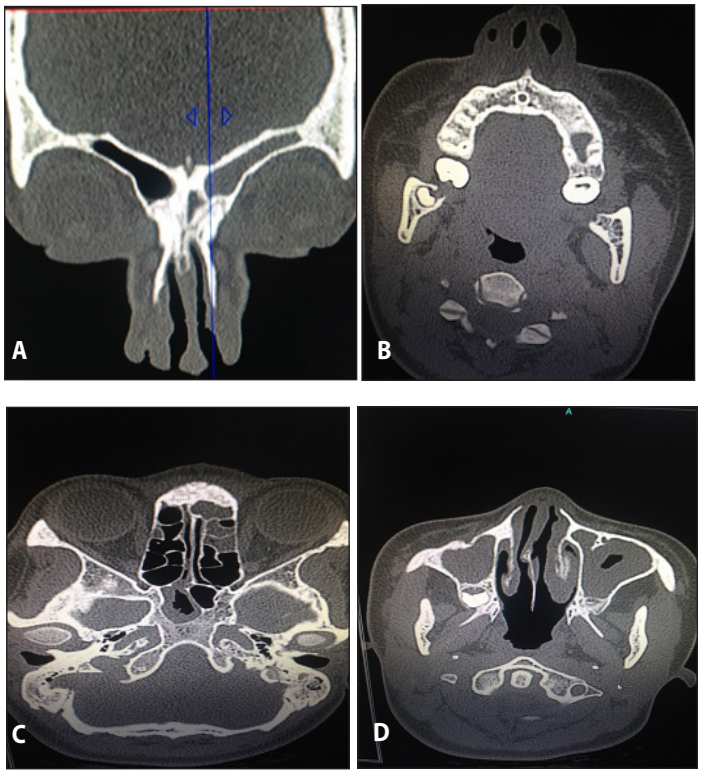

Figure 3. Paranasal Sinus CT scans. **A.** Coronal and **B.** Axial cut show a small opening lateral to the right nasal ala that does not communicate with the right nasal cavity. **C.** and **D.** Axial cuts reveal opacifications in the left frontal sinus, left anterior ethmoid, both maxillary sinuses and mucosal thickening of the right sphenoid sinus with deviated nasal septum to the right.

was made along the inner circumference of the supernumerary nostril and the blind ending nasal tract was excised. The result was a medial nostril and untouched ala with dead space in between. A portion of the ala was removed and a Z plasty was designed. Through-and-through sutures were placed to obliterate dead space. *(Figure 4)* Endoscopic sinus surgery was performed to remove the left nasal polyp and establish a functional osteomeatal complex.

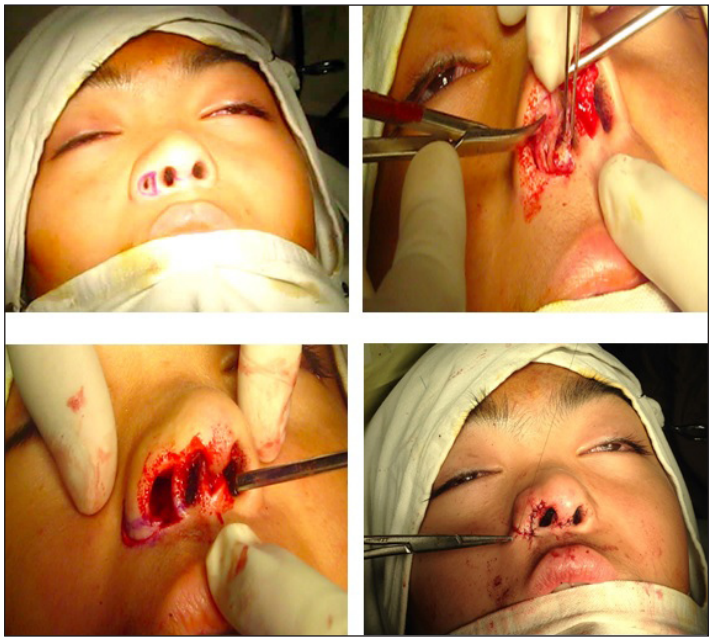

Figure 4. Intraoperative photos **A.** crescent incision **B.** blind ending nasal tract excised **C.** z-plasty and **D.** closure

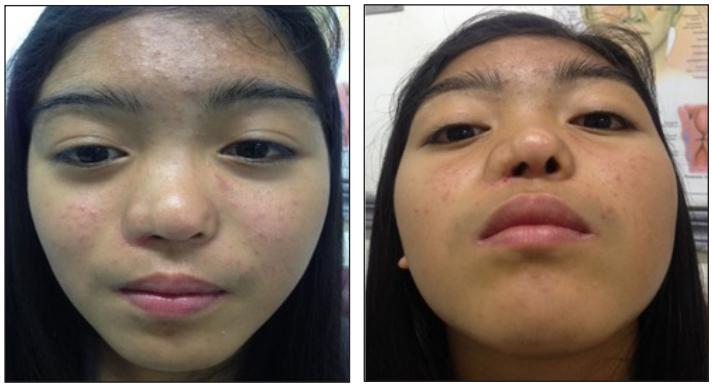

Figure 5. Postoperative frontal and basal view (published in full with permission)

The post-operative period was uneventful and she was discharged after 2 days with a slightly swollen right nostril. Results at 2 months showed improvement in the contour of the right nostril. *(Figure 5)*

DISCUSSION

Supernumerary nostrils are rare congenital anomalies with unclear etiology. A review of literature reveals that there are about 40 published cases since 1906.[2-12] Of these 40 cases, 23 were isolated supernumerary nostrils and 15 had associated anomalies; more than half of the reported cases were from the Asian continent of which 10 were from India.[2-12]

There is no known exact mechanism and stage of embryological development at which the error occurs and because of its rarity, different speculative theories exist related to its embryogenesis.[11] According to Erich, Lindsay described a case of bilateral supernumerary nostrils and proposed the theory of dichotomy by atavism or parallel evolution in 1906.[2] In biology, "an atavism is an evolutionary throwback, such as traits reappearing which had disappeared generations before… that can occur in several ways. One way is when genes for previously existing phenotypical features are preserved in DNA, and these become expressed through a mutation that either knock out the overriding genes for the new traits or make the old traits override the new one."[13]

In 1962, Erich reported a case of a patient with two noses and explained the development of that deformity and the accompanying four nostrils by the appearance of four nasal pits instead of two, horizontally, each forming a nasal sac.[2] He hypothesized that "the medial sacs are interposed between the nasal laminae, preventing their fusion into one septum and resulting in a double nose and suggested that when the accessory nasal pit is located too laterally, the fusion of the laminae is not disturbed and one nose with a supernumerary nostril is formed."[2]

Nakamura and Onizuka advanced a new theory when they reported a case of supernumerary nostril in 1987.[3] They suggested that "the supernumerary nostrils resulted from a localized abnormality of the lateral nasal process, with a fissure appearing accidentally and dividing the lateral nasal process in two, resulting in two nostrils on one side of the nose."[3] Also in 1987, Reddy and Rao reported a case of triple nostrils and assumed that the "deformity developed as a result of an accessory olfactory pit appearing either above or below the normal location of the placode."[4]

As for treatment, there is no generally accepted method of surgical repair. The goal of repair is to create a functioning and normal-appearing nose with minimal deformity.[5] My review of literature revealed a systematic classification of supernumerary nostril by Saiga and Matsua and its corresponding surgical technique.[6] The classification is based on the shape of the nasus externus. *(Figure 8)* Based on this classification, our patient belongs to Type 1 where the inner nostril is sufficiently large and symmetry with the unaffected side can be obtained. In this case, the suggested surgical technique is resection of the outer nostril.

Although various types and techniques exist for the management of a supernumerary nostril, most of the authors advocated the circumferential excision of the accessory cavity and resection of the entire fistulous tract.[2,5-8,11-12] As to the timing of surgery, it is advisable to operate in early childhood to prevent deformity of normal nostrils.[4] For adult patients, surgical treatment should be planned in stages to achieve a good outcome.

Because of the rarity of this congenital anomaly, the etiology and mechanism of developmental error remain hypothetical. An observation of a high incidence in the Asian continent calls for further investigation to identify common factors in this geographical region that contribute to the deformity.[7] As mentioned by Rout and Lath, there is no cookbook for the correction of this defect.[7] Treatment has to be tailored as to the presentation of a supernumerary nostril and any other associated deformity. Though various surgical interventions exist, the goal of repair is to create a functioning and normal-appearing nose essential for normal psycho-social development.

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