Supernumerary nostril is an extremely rare congenital entity that results from aberrant embryological development. A review of the literature reveals that only 33 cases of supernumerary nostril have been reported. They can be associated with other congenital anomalies. The accessory nostril may or may not communicate with the ipsilateral nasal cavity, probably depending on the degree of embryological progression of the anomaly. A case of supernumerary left nostril with no nasal cavity communication is presented. The surgical treatment is described and different speculative theories related to the embryogenesis of supernumerary nostrils are also reviewed.

Key Words: Supernumerary nostrils; False nostril; True nostril

Supernumerary nostril is a very rare type of congenital anomaly. The first reported case was published in 1906 by Lindsay (1), who described a patient with bilateral supernumerary nostrils. In that case, the external openings of the supernumerary nostrils were situated above the normal nostrils, and the accessory nasal cavities communicated with the ipsilateral nasal cavities. In the report, Lindsay (1) proposed the theory of dichotomy by atavism or parallel evolution. In 1920, Tawse (2) reported a patient with a unilateral supernumerary nostril that communicated with the nasal cavity. In 1987, Reddy and Rao (3) reported a case of a third nostril that was situated below the left nostril; they hypothesized that the extra nostril arose as a result of an accessory placode or pit. An accessory nasal placode may be present either above or below the normal nasal placode. The present article describes a case in which the supernumerary nostril with a small accessory nasal cavity, which did not communicate with the normal nasal cavity on the same side, appeared in a six-month-old girl.

CASE PRESENTATION

A six-month-old girl was brought to the plastic surgery outpatient department by her parents for evaluation of an abnormal opening above the left nostril since birth. The patient’s parent did not give any history of discharge from the accessory opening. According to the history provided by the mother, her pregnancy had been uneventful and the child’s birth was normal. The girl had reached normal milestones of motor and intellectual development. No family history of such an anomaly was reported. Physical examination revealed the presence of a small cavity 8 mm above the left nostril. Nasal endoscopy of the accessory nasal cavity revealed that it was small and did not communicate with the ipsilateral normal nasal cavity. No anomalies were apparent in the normal nasal cavities. Careful examination of other systems did not detect any other abnormality (Figure 1).

At the four-week follow-up examination, the patient was doing well (Figure 5). It has been observed there is minor discrepancy in the septum showing marking for the incision (Figure 3). The de-epithelized alar base of the true nostril to reconstruct the nostril proper. An excess portion of the nasal composite tissue was excised on the alar rim of the accessory nasal cavity and the proper shape was maintained. A bolster suture was applied to the alar rim. The patient recovered uneventfully. The relatives were advised to keep the nostril retainer for maintaining nasal shape for eight weeks (Figure 4).

The patient underwent reconstruction of the left nostril. With the patient under general anesthesia, a peri alar incision was made on the left nostril (Figure 2). Septum or partition, which was connecting the apex of the columella to the alar base, was excised including a small part of alar base of false nostril (Figure 3). The de-epithelized alar base of false nostril attached to de-epithelized part of alar base of the true nostril to reconstruct the nostril proper. An excess portion of the nasal composite tissue was excised on the alar rim of the accessory nasal cavity and the proper shape was maintained. A bolster suture was applied to the alar rim. The patient recovered uneventfully. The relatives were advised to keep the nostril retainer for maintaining nasal shape for eight weeks (Figure 4).

At the four-week follow-up examination, the patient was doing well (Figure 5). It has been observed there is minor discrepancy in the position of the alar base. Therefore, correction of the discrepancy of alar rim with conchal composite graft and tip plasty will be planned in a later period.
DISCUSSION

The face develops from five facial primordia appearing as prominences around the stomodeum or primitive mouth. There is a single median front nasal prominence and paired maxillary and mandibular prominences. These prominences are produced by the neural crest cells migrating into the branchial arches during the fourth week of gestation. Facial development occurs mainly between the fourth and the eighth week. By the end of the fourth week, bilateral thickenings develop on the ventrolateral ectoderm of the front nasal prominence, known as nasal placodes.

The margins of the placodes proliferate into medial and lateral nasal prominences, resulting in the placodes lying in depressions known as nasal pits. The medial nasal prominences merge together and with the maxillary and lateral nasal prominences, resulting in separation of the nasal pits from the stomodeum. The nasal pits become nasal sacs and then develop into nostrils and nasal cavities (4).

 Supernumerary nostrils are exceedingly rare congenital anomalies of unclear etiology. In 1962, Erich (5) reported a case of double nose. He also supported Lindsay’s theory of dichotomy by atavism or parallel evolution, and he further speculated that if the accessory nasal pit is located too laterally, the fusion of the lamina is not affected, which leads to the formation of a supernumerary nostril. In 1972, Onizuka and Tai (6) reported the case of a single accessory nostril that had developed above the nasal ala. In 1987, Nakamura and Onizuka (7) reported a similar case; they hypothesized that the cause was probably a localized defect in the lateral nasal process. They hypothesized that during the proliferation of mesenchymal cells in the lateral nasal process, a concavity or fissure appears in this area accidentally and, thus, this lateral nasal process is divided into two segments, resulting in two nostrils and two alae on one side. This hypothesis can extrapolate the appearance of accessory nostril either above or lateral (as in our case) to the natural nostril or medially, depending on the position of change in the lateral nasal process. According to Erich (5), during the course of the evolution of the nasal placode, four nasal pits appeared horizontally, each became a nasal sac, and the medial two, which were interposed between the two nasal laminae, prevented the laminae from fusing into one nasal septum. This resulted in double nose. Supernumerary nostril is formed when the accessory nasal pit is located so laterally to the nasal lamina that the accessory nostrils are formed above the natural nostril and, thus, do not disturb the fusion of the nasal laminae (8), reported a case of supernumerary nostril with extra lower lateral cartilage and also supported the theory embryological fissuring of the lateral nasal process. Sinha et al (9) reported a case of supernumerary nostril with microcormea and congenital cataract and speculated that anomaly in development of the nasal placodes is the cause.

The presence of alar cartilage in present case report describes the embryological fissuring of the lateral nasal process and formation of supernumerary nostril. This theory has been supported by various authors in past.

In 1992, Chen and Yeong (10) described a case of bilateral supernumerary nostrils that were situated below the normal nasal openings, and they proposed treating such anomalies by staged corrective surgery. In 2001, Hallak et al (11) reported a case of supernumerary nostril in which a blind cavity was present in a normally developed nose. They advocated that corrective surgery be performed at an early age to prevent any possible alar deformity. The present case is the 34th case of supernumerary nostril reported. Sah et al (12) reviewed 33 cases of supernumerary nostril that had been reported worldwide.

Most reported cases of supernumerary nostrils have been unilateral, and most were associated with other craniofacial malformations, such as a facial cleft. A supernumerary nostril may or may not communicate with the ipsilateral normal nasal cavity, depending on the extent of the anomaly’s embryological progression (11). Our case of supernumerary nostril was not communicating with the nasal cavity and it is a large deformity of the nose.

CONCLUSION

Because of the extreme rarity of this congenital anomaly, its cause and development remain largely hypothetical. One common observation noted is that all authors advise early surgery, including excision of the fistulous or blind tract, or performing a fistulorhinostomy when the proximal portion is not accessible. Surgery at an early age is recommended; it will prevent possible subsequent alar deformity and will provide a more normal appearance, which is essential for normal psychosocial development (13). Because it is rare, genetic study of this anomaly can be suggested to determine the cause. Aesthetic outcome may be achieved better in a large series.

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REFERENCES

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