Supernumerary nostril: Report of a rare case and a review of the literature

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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).

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 collumella 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.
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nostrils and two alae on one side. This hypothesis can extrapolate
this lateral nasal process is divided into two segments, resulting in two
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
already 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 embry-
ological fissuring of the lateral nasal process. Sinha et al (9) reported
a case of supernumerary nostril with microcornea 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 super-
numerary nostrils that were situated below the normal nasal openings,
and they proposed treating such anomalies by staged corrective sur-
gery. In 2001, Hallak et al (11) reported a case of supernumerary nos-
tril 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 unilat-
eral, and most were associated with other craniofacial malforma-
tions, 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 cav-
ity 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 recom-
mented; 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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