Supernumerary nostril with congenital cataract

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Abstract
Supernumerary nostril is a very rare congenital anomaly. It can be unilateral or bilateral, and it sometimes occurs in the presence of other congenital deformities. Behind the external opening of a supernumerary nostril is a small accessory nasal cavity, which may or may not communicate with the normal nasal cavity on the same side. We describe a new 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 young girl who also had microcornea and congenital cataract. The accessory nasal cavity was successfully removed surgically. We believe that this case may represent the first reported case of a supernumerary nostril with a congenital cataract on the same side. We also discuss the hypotheses that have been proposed to explain supernumerary nostrils.

Introduction
Supernumerary nostril is a very rare type of congenital anomaly. The first reported case was published in 1906 by Lindsay, 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 his report, Lindsay proposed the theory of dichotomy by atavism or parallel evolution. In 1920, Tawse reported a patient with a unilateral supernumerary nostril that communicated with the nasal cavity. In 1987, Reddy and Rao 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.

In this article, we describe a new 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 young girl who also had microcornea and congenital cataract. After an extensive search of the literature, we arrived at the conclusion that ours is the first reported case of a supernumerary nostril and congenital cataract on the same side.

Case report
A 2-year-old girl was brought to the ENT Department for evaluation of an opening above the left nostril and a white spot in the left eye; both anomalies had been present since her birth (figure 1). A persistent, thick, whitish fluid discharged from the opening. The patient’s mother explained that the opening would swell intermittently. According to the history provided by the mother, her pregnancy had been uneventful and the child’s birth was normal. The child 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 3 mm above the left nostril. The cavity was lined with mucous membrane and filled with mucoid discharge. Nasal endoscopy of the accessory nasal cavity revealed that it was small and did not communicate with the ipsilateral normal nasal cavity, a finding that was later confirmed by sinography. No anomalies were seen in the normal nasal cavities. Examination of the eyes revealed microcornea and congenital zonular cataract of the left eye. The left eyelid contained fewer eyelashes than did the right eyelid. The right eye was normal. Careful examination of other systems did not detect any other abnormality.

The patient underwent surgical excision of the accessory nostril followed by reconstruction of the defect. With the patient under general anesthesia, a circular incision was made along the margin of the accessory nasal cavity. The mucous membrane and an almost equally thick section of submucous tissue in the accessory nasal cavity were then excised (figure 2). Extreme care was taken to avoid damage to the cartilaginous frame of the accessory nasal cavity and...
the nasal cavity on the left side. The raw roof and the raw floor of the accessory nasal cavity were sutured together to obliterate the cavity, and the skin incision was closed with 6-0 polypropylene suture material. The patient recovered uneventfully, and she was referred to the ophthalmology clinic for evaluation of her microcornea and cataract.

Histopathologic examination of the excised tract from the accessory nasal cavity revealed that the respiratory mucosa was lined with pseudostratified ciliated columnar epithelium. The specimen also contained numerous goblet cells and a few islands of squamous epithelium.

At the 6-month follow-up examination, the patient was doing well. The functional outcome was excellent and the cosmetic result was satisfactory (figure 3).

Discussion
Embryologically, by week 4 of intrauterine life, precursors of nasal cavities and the lenses of the eyes are formed. The nasal placodes (olfactory placodes) arise from the medial aspect of the lower portion of the frontal prominence, and the lens placodes arise from the lateral aspect of the lower portion of the frontal prominence. At week 5, the mesenchyme, which covers the caudal surface of the forebrain, proliferates with the surface ectoderm to form the frontonasal process. Two ectodermal thickenings (the nasal placodes) arise on each side of the dependent part of the frontonasal process. Subsequently, a depression develops in the surrounding mesenchyme on each side of the two nasal placodes, thereby forming two olfactory pits or gutters. These olfactory pits separate the frontonasal process into a medial nasal process and two lateral nasal processes. The lateral nasal processes subsequently form the alae of the nose. As the olfactory pits progress deeper to form the primitive nasal cavity, the medial nasal process thins out gradually to form the primitive nasal septum. At this stage, the maxillary process develops from the cephalic side of the dorsal part of the mandibular arch. Each maxillary process grows ventromedially to meet and fuse with the lateral nasal processes, and each ultimately fuses with the medial nasal process, thus forming the external opening of the primitive nasal cavity and the upper lip.

Supernumerary nostrils are exceedingly rare congenital anomalies of unclear etiology. In 1962, Erich 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 reported the case of a single accessory nostril that had developed above the nasal ala. In 1987, Nakamura and Onizuka reported a similar case, and they hypothesized that the cause was probably a localized defect in the lateral nasal process. In 1992, Chen and Yeong 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. Finally, in 2001, Hallak et al. 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.

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 embryologic progression.

In our patient, the probable explanation for the embryogenesis of the supernumerary nostril, accessory nasal cavity, and congenital cataract involves a defect in the migration of cells from the frontal prominence, which led to a simultaneous anomaly in the development of the nasal and lens placodes. In this case, the additional olfactory pit that formed was clearly smaller than the normal olfactory pits. At the time that the primitive nasal cavity had formed, the additional olfactory pit developed into an incomplete, smaller accessory nasal cavity whose anatomy resembled a cul-de-sac; the accessory cavity did not communicate with the rest of the nasal cavity or with the nasopharynx.

The accepted treatment of a supernumerary nostril is excision of the accessory nasal cavity followed by reconstructive surgery. Surgery should be performed as early as possible.

References