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CASE REPORT
Craniofacial/Pediatric

Congenital Nostril Stenosis with a Horseshoe-like Shape

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Summary: We report a rare case of congenital nostril stenosis because it is very interesting from the perspective of human embryo development. As we were not able to find a similar congenital case in the literature, we would like to describe it here. The patient is a 36-year-old woman who had bilateral congenital stenotic nostrils with horseshoe-like shape. Particularly, the bilateral medial crura protruded prominently. When she inhaled strongly, she could not breathe further because the action was analogous to the closure of a valve. Her elder sister had similar stenosis in the right nostril only, and parents did not have any stenotic nostrils. During the surgery, the prominent webs of the bilateral medial crura were resected, and Y-V advancement flap was inserted bilaterally to avoid restenosis. Histopathological examination revealed that the resected tissue contained cartilaginous and muscular tissue. A Koken retainer was worn throughout each entire day without washing the face or taking a bath for 3 months after surgery and was washed for keeping hygiene many times every day. After surgery, the patient can breathe well when she inhales strongly and is very satisfied with the operative results. Stenosis has not recurred 6 months after operation. Pathogenesis of this case is thought to occur at the early phase of embryo development when the medial and lateral nasal placodes form a downward-facing "horseshoe." *(Plast Reconstr Surg Glob Open 2023; 11:e5453; doi: 10.1097/GOX.0000000000005453; Published online 27 November 2023.)*

Cases of nasal nostril stenosis with bilateral choanal atresia have been reported.[1–3] Such a case requires surgical repair for the stenotic site because nasal airway narrowing induces various degrees of respiratory distress. A congenital accessory skin appendage of the columella and nostril sill[4,5] and accessory nostril,[6,7] a rare congenital nasal anomaly, have also been reported to cause nostril stenosis.

We report a rare case of congenital nostril stenosis because it is very interesting from the perspective of human embryo development. The cause of the stenosis is generally thought to be the malposition of bilateral footplates of the medial crura.[8] The causality of our case is thought to differ because it did not involve malposition of bilateral footplates. Because we could not find a similar congenital case in the literature, we would like to describe it here.

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PATIENT AND METHODS

The patient is a 36-year-old woman who had bilateral congenital stenotic nostrils with a horseshoe-like shape. Particularly, the bilateral medial crura protruded prominently (Fig. 1). When she inhaled strongly, she could not breathe further because the action was analogous to the closure of a valve. Her elder sister had similar stenosis in the right nostril only, and parents did not have any stenotic nostrils (Fig. 1).

During the surgery, the prominent webs of the bilateral medial crura were resected, and a Y-V advancement flap was inserted bilaterally to avoid restenosis after surgery due to scar contracture (Figs. 2 and 3). Almost all parts of the resected webs had cartilaginous tissue. (**See figure, Supplemental Digital Content 1**, which shows that the resected web had cartilaginous tissue. **http://links.lww.com/PRSGO/C908**.) The cartilage had spread parallel to the stenotic web. Histopathological examination revealed resected tissue containing cartilaginous and muscular tissue (Figs. 2 and 3). **[See figure, Supplemental Digital Content 2,** which shows that the resected tissue had

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Related Digital Media are available in the full-text version of the article on www.PRSGlobalOpen.com.

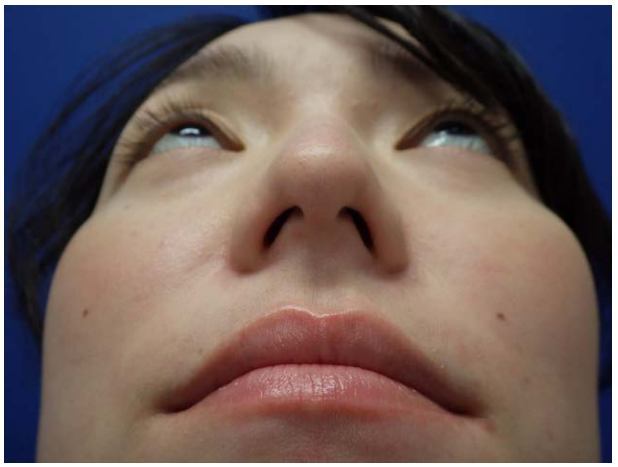

Fig. 1. A 36-year-old woman with bilateral congenital stenotic nostrils with a horseshoe-like shape.

cartilaginous tissue (H&E stain). **http://links.lww.com/PRSGO/C909**.]

RESULTS

A Koken retainer was worn through the entire day without washing the face or taking a bath for 3 months after surgery. After surgery, the patient is able to breathe well when she inhales strongly and is very satisfied with the operative results. Stenosis has not recurred 6 months after operation (Fig. 4).

DISCUSSION

In the early fifth week of embryonic development, the medial and lateral nasal placodes start to sink. They form a downward-facing "horseshoe" with an open end.[9] As the medial and lateral processes develop and become more prominent, the nasal grooves deepen and become the nasal pits. By the late fifth week, these pits continue to actively deepen, forming the nasal sacs. The medial nasal process is more prominent than the lateral nasal process, extending farther ventrally and caudally. The medial crura of the patient retained this characteristic, and the nostril continued to exhibit a horseshoe shape after birth.

However, by the mid-seventh week, the anterior lumen of each primitive nasal cavity becomes tightly filled with a proliferation of epithelial cells that form an epithelial plug.[10] By the 13th to 15th week, the plug dissolves. A case of congenital nostril atresia has been reported.[2] The report indicated that pathogenesis of the atresia may be due to the persistence of the temporary nasal epithelial plug during the early stages of fetal life.[3] If the plug is not completely absorbed and does not disappear, this leads to congenital anterior nostril atresia. However, our resected tissue of the stenotic nostril was not only epithelium characteristic of a nasal plug but also consisted of cartilaginous and muscular tissue. The origin of the cartilaginous tissue was thought to come from the nasal septum and the muscular tissue from the fibers toward the septum from the musculus nasalis. Pathogenesis of this case is thought to occur at the early phase of embryo development when the medial and lateral nasal placodes form a downward-facing horseshoe. The bilateral footplates of the medial crura were generally thought to spread laterally in this case.[8] We thought that the anomaly was accompanied with abnormality of the position in the footplates of the medial crura because almost all parts of the web contained the cartilage spread parallel to the stenotic web. The footplates should be situated at right angles to the web.[8]

A congenital accessory skin appendage of the columella and nostril sill[4,5] and accessory nostril,[6,7] a rare

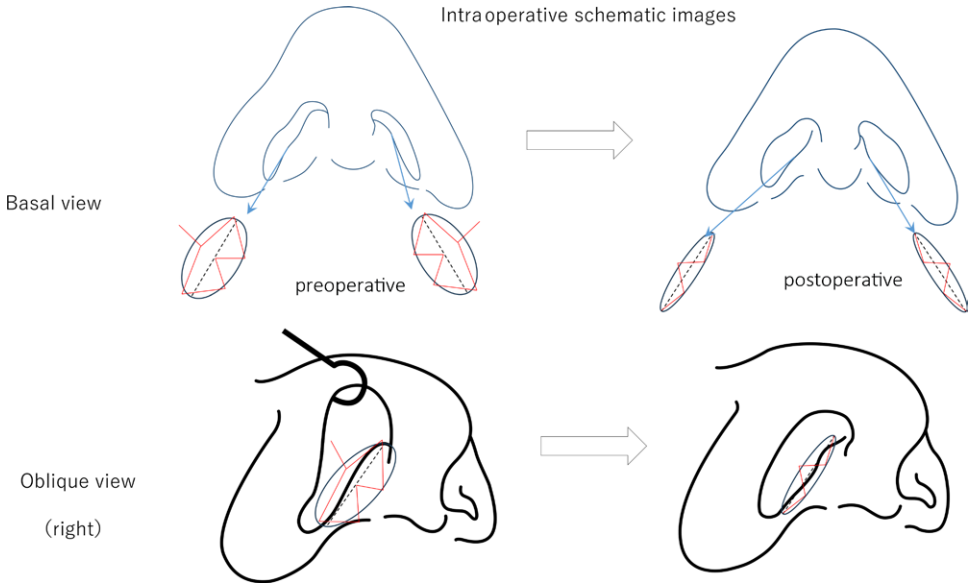

Fig. 2. A schema of the operation. Web resection and Y-V advancement flap were performed.

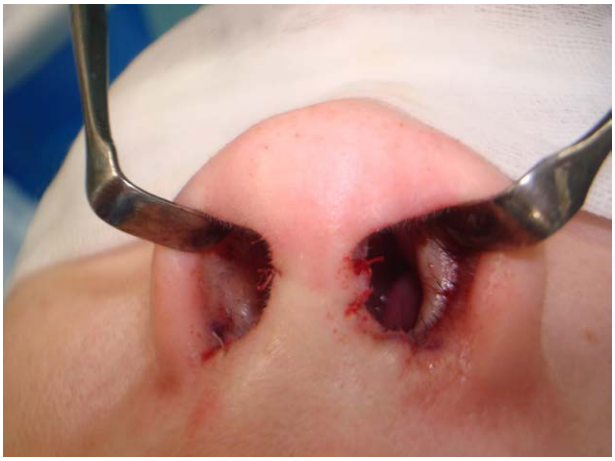

Fig. 3. The condition just after operation.

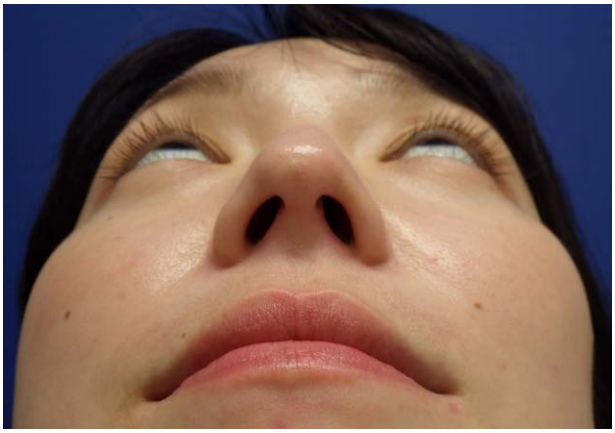

Fig. 4. The condition 6 months after operation.

congenital nasal anomaly, were also reported to cause nostril stenosis. It is hypothesized that an obstacle or injury during medial migration of the medial nasal process results in an accessory skin appendage of the collumella.[4] Accessory nostril is a very rare congenital anomaly, which is still unclear in etiology.[6] However, our case differs from those.

For treatment, a modified method of running Y-V plasty has been reported to correct bilateral nostril stenosis in an acquired case with a circular, linear contracture. However, for our case, only one Y-V plasty was performed because the web of the medial crura was prominent. No stenosis has recurred after our operation.

CONCLUSIONS

We report a rare case of bilateral congenital nostril stenosis and discuss the etiology from the perspective of human embryo development.

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DISCLOSURE

The authors have no financial interest to declare in relation to the content of this article.

PATIENT CONSENT

The patient provided written consent for the use of her image.

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