Case report

Bilateral congenital aplasia of nasal lower lateral cartilage: a rare anomaly

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SUMMARY

Congenital nasal anomalies are rare and occur in 1/20 000–1/40 000 newborns. An 8-year-old boy presented with developmental aplasia of bilateral nasal lower lateral cartilages, with excessive wrinkled and loose skin on the dorsum of the nose and with difficulty breathing through the nose. This is probably the first such case to be reported in the literature. The defect was reconstructed using conchal and septal cartilage grafting through an external rhinoplasty approach. At the end of the 12-month follow-up period, the patient was found to be satisfied with the functional and aesthetic results of the operation. Bilateral congenital aplasia of nasal lower lateral cartilages is extremely rare. Paediatric rhinoplasty is imperative in such cases.

BACKGROUND

Congenital nasal deformities are very rare and cause multiple issues including psychosocial trauma, leading to an impaired academic performance of a child. Congenital nasal anomalies occur in 1/20 000-1/40 000 newborns. Isolated congenital absence of the alar cartilages and nasal septum is a particularly rare entity.² It is important to take a detailed history of functional and aesthetic complaints due to nasal deformities. The quality of the skin, that is, thickness, elasticity and colour, and pathological condition are important in predicting surgical outcomes.³ Patients mostly present as adults due to the low priority given to the condition at an early age, especially in low-income countries. The ensuing management thus follows protocols for adults. We present a rare case of congenital aplasia of bilateral nasal lower lateral cartilages in an 8-year-old child. Our case presented at an early age, and despite the scarcity of literature available on this condition good outcome was achieved through timely and decisive management.

CASE PRESENTATION

An 8-year-old boy presented with complaints of external nasal deformity and nasal airway obstruction since birth. There was no history of any trauma, nasal surgery or any associated congenital abnormalities.

Examination revealed the septum to be in the midline. There was a complete collapse of the nasal alae as well as the nasal tip, with severely compromised external nasal valves. The patient had excessive loose and wrinkled skin on the dorsum of the nose, with a fibrotic band. There were no palpable



Figure 1 Preoperative frontal view of the face.

lower lateral cartilages. Lip and dental examination was normal (figures 1 and 2).

TREATMENT

Informed consent was obtained from the parents of the child and the patient was scheduled for surgery. Under general anaesthesia with full aseptic precautions and after endoscopically guided nasal evaluation, transcolumellar inverted-V incision at the base of the columella and marginal incision along the alae were performed. Elevation of the skin revealed rudimentary fibrous lower lateral cartilages. Dissection was carried out in the supra-perichondrial and subperiosteal planes. Intracorporeal septoplasty



Figure 2 Preoperative lateral view of the face.



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Figure 3 Intraoperative images.

was performed. A 1.5×3.0 cm strip of the septal cartilage was harvested. The integrity of the cartilaginous bony junction was maintained, preserving 1.5 cm of the dorsal and caudal strip of the cartilaginous septum. A 1.0×2.5 cm conchal cartilage was harvested from the right pinna, preserving the antihelical fold. A columellar strut was sculpted from the septal cartilage and was placed between the two hypoplastic medial crura of the lower lateral cartilages within a subperichondrial pocket. This was sutured to the periosteum of the anterior nasal spine and the caudal end of the septum. Two curved cartilage struts were sutured with the upper lateral cartilage and columellar strut using Ethilon 5-0 to create a dome and the lateral crura (figure 3). An additional small cartilaginous shield graft was kept. A dorsal cartilage onlay graft was placed. Lateral osteotomies were performed to narrow the lateral walls. Excess skin on the nose was paired and sutured. Both nasal cavities were packed with Merocel and dressing was done with steristrips. A plaster of Paris cast was placed. Tip projection and alar symmetry were maintained at 12-month follow-up (figures 4 and 5).



Figure 4 Postoperative frontal view of the face (after 12 months).



Figure 5 Postoperative lateral view of the face (after 12 months).

OUTCOME AND FOLLOW-UP

The patient and his parents were satisfied with the results of the surgery. The patient was doing well with his social life and academics after 12 months of follow-up.

DISCUSSION

Losee *et al*¹ devised a morphogenic classification system classifying congenital nasal deformities into four categories. Type I, or hypoplasia and atrophy, represents atrophy or underdevelopment of the skin, subcutaneous tissue, muscle, cartilage and/or bone. Type II, or hyperplasia and duplications, represents anomalies of excess tissue. In type III, or clefts, the comprehensive and widely used Tessier classification of craniofacial clefts is applied. Type IV includes neoplasms and vascular anomalies. According to this classification, our case was of type I anomaly.¹

The external nasal valve consists of the ala, the skin of the vestibule, the nasal sill and the contour of the medial crus of the lower lateral cartilages.⁴ In cases with severe deformity of the nasal tip, which needs extensive reconstruction, open (external) approach is preferable.⁵ We employed open rhinoplasty approach due to the severe deformity of the lower lateral cartilages and tip.

Depending on the operative technique and extent of correction, cartilaginous grafts from the concha, septum or a combination of these can be used. We employed a combination technique in our patient. The auricle can provide a useful amount of cartilage for nasal reconstruction when septal cartilage is insufficient. Shield graft, popularised by Sheen,⁶ is used in nose that lacks tip projection in the under projected tip with a short columella

Learning points

- ► Bilateral congenital aplasia of nasal lower lateral cartilages is extremely rare.
- ► Paediatric rhinoplasty is imperative in such cases.
- ► Timely surgical intervention can prevent psychosocial trauma in a child.

and weak lower lateral cartilages.⁷ We used shield graft in our patient.

In minor tip deformities, when the lateral crura is collapsed or mildly deformed, septal or auricular cartilage can be used for the lateral crural strut graft to re-establish the shape and stability of the lateral legs of the tripod.⁵ Where lateral crura are not usable, although the medial crura and domes are intact, a columellar strut is placed and sutured between the medial crura to strengthen the caudal leg of the tripod. The lateral crural strut grafts are sutured to the undersurface of each dome to replace the missing lateral crura.

Conventional wisdom suggests delaying rhinoplasty in children until they are about 18 years of age; however, it may be performed for anatomical, physiological, social and psychological reasons, as in the present case. There are only few case series of lower lateral cartilage aplasia, and this is the first reported case of a bilateral nasal lower lateral cartilage aplasia with overlying skin deformity in a child.⁸

Contributors KS is the primary surgeon responsible for the conception and design of the study. DK and BC are the assistant surgeons responsible for drafting the article and providing the images. RKS was responsible for proof-reading the article and finalising the manuscript. All authors have read and approved the final version of the manuscript.

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