Reconstruction of Congenital Absent Columella
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A case of congenital absence of the nasal columella accompanying a facial hemangioma is presented. The nose is an important aesthetic unit of the face, and its projection is mainly supported by the columella. The etiology of the absent columella is unclear, and the clinical feature is rare. Reconstruction of the columella remains a challenging problem in plastic surgery, and numerous techniques have been described. The authors discuss several techniques aimed at the correction of the problem and describe a method of reconstruction of the absent nasal columella that was used in the case reported.

Key Words: Congenital absent columella, columella reconstruction

The nasal columella is an important aesthetic unit of the face that gives support and projection to the tip of the nose. Defects of the nasal columella, result in significant cosmetic and functional deformities. Such defects usually are caused by tumor resection,1-4 trauma,2,5,6 or infection.7,8 Congenital absence of the nasal columella is extremely rare, and only five cases have been reported in the literature.9-11 The authors present the sixth case of congenital absent columella accompanying hemangioma of the upper lip, left fronto-orbital region, and the left ear.

CASE REPORT

A 2-year-old girl was referred to our clinic with nasal deformity and hemangiomatous lesions on her face. Physical examination revealed a 1- x 1.5-cm defect of the columella and the caudal portion of the septum (Fig 1). This resulted in retroplacement of the dome and shrinkage of the airway. She also had involuting hemangioma involving the full thickness of the upper medial lip, the left fronto-orbital region, and the left ear. The patient was born in a normal delivery at term after a normal pregnancy. The birth weight was 3300 g. Careful questioning revealed the columella was absent at birth. The progression of the hemangiomas started 2 months after birth, and involution started at 18 months. There was no consanguinity in the parents. Family history was negative for other congenital anomalies. Results of blood analyses were within the normal limits. No other pathologic features were observed in the radiologic examinations of the cranium.

With the patient under general anesthesia, after local anesthetics were administered, an open rhinoplasty incision was performed. Because the inferior border of the septum had a cover of stratified squamous epithelium, the authors decided to use it as the columellar cover. Upper and lower caudal septal flaps were dissected from the septal cartilage. Thereafter, bilateral medially based, vestibular labial mucosal flaps with dimension of 10 × 30 mm were elevated. They were transported to the nostril bases through submucosal tunnels. The proximal parts of these flaps remaining submucosally were de-epithelized, and oral incisions were closed with 5/0 absorbable sutures. To achieve columellar support and adequate tip projection, a cartilage graft of 1 × 2 cm was harvested from the right auricular concha. The donor site was closed primarily with 4/0 non-absorbable sutures. The cartilage graft was anchored in a “T” shape to be used as a strut graft and placed caudally to the septal cartilage. Vestibular labial mucosal flaps were used to cover the lateral mucosal sides of the cartilage graft and sutured with 5/0 absorbable sutures. Caudal septal flaps were used to cover the caudal side of the columella and sutured with 5/0 nonabsorbable sutures (Fig 2).

DISCUSSION

The formation of the face is preceded by migrations of cranial neural crest cells from the region of the trigeminal nerves to the face.12 These cells establish the mesodermal elements that later develop into the face. Development of the nose occurs between 3 and 10 weeks of gestation.12 The nasal alae are formed by fusion of the nasal lateral and medial processes.13 The medial nasal processes fuse in the
midline with the frontal prominence and result in the formation of the frontonasal process that gives origin to the columella, philtrum, upper lip, nasal bones, cartilaginous nasal capsule, and superior alveolar ridge.\textsuperscript{13} The nasal placodes, which are local thickenings of surface ectoderm, invaginate to form the nasal pits during the 5th week of gestation. Cells within the nasal pits continue to migrate posteriorly to form the primitive nasal cavities.\textsuperscript{14} By the 9th week of gestation, the cartilaginous nasal septum, which results from persistence of neural crest cells between the nasal cavities, directly overlies the buccal cavity.\textsuperscript{14} By the 10th week of gestation, the palatal shelves and the inferior septum fuse to form the secondary palate.\textsuperscript{12}

Lack of mesenchymal migration and fusion of ectodermal and mesodermal elements results in clefts of this region. Absence of the septum is a component of Tessier 0 cleft.\textsuperscript{15} Clinical features suggest that these defects result from genetic or environmental factors.\textsuperscript{16} Congenital absence of the nose is rare, and the etiology of this condition is unknown.\textsuperscript{17} Some postulations suggest that lack of development of the nose results from failure of the medial and lateral nasal processes to grow, lack of resorption of the nasal epithelial plugs during the 13th to 15th weeks of gestation, abnormal migration of neural crest cells to this region, or aberrant flow of the multiple mesodermal structures required to establish the nose and its cavities normally.\textsuperscript{18}

Congenital absence of the nasal columella is extremely rare, and only five cases have been reported in the literature.\textsuperscript{9,11} Some authors have emphasized that the reason for the rarity might be that absence of columella seems inconspicuous.\textsuperscript{9,10} The pathogenesis of isolated absence of the columella has not yet been understood.\textsuperscript{14} It has been suggested that a teratogen introduced at the time of columellar development might selectively arrest the cellular penetration and chondrification of the nasal columella.\textsuperscript{9,11}

Our patient had another pathologic entity—hemangioma involving the upper lip, left frontoorbital region, and the left ear. Hemangiomas are the most common tumors of infancy; they usually appear after birth, grow rapidly, and involute with time.\textsuperscript{19} Congenital hemangioma is a distinct lesion that is fully grown at birth, and the anatomic distribution is the same as for common infantile hemangioma.\textsuperscript{20} The most common complication of hemangioma is ulceration,\textsuperscript{21,22} and it often involves the perioral and perianal lesions.\textsuperscript{23} Nasal hemangiomas are particularly slow to regress, compared with those of other sites, and remaining fibro-fatty deposits cause contour deformities.\textsuperscript{24} The authors found no reported case of nasal columellar atrophy after an ulcerating or involuting congenital hemangioma. Questioning of the parents confirmed that the columella was absent at birth.

The nasal columella is an important aesthetic unit of the face that gives support and projection to the tip of the nose. It extends from the tip of the nose to the upper lip, joining the lip at the upper portion of the philtrum and separating the external nares. Its loss results in a severe aesthetic and functional deformity. Ultimate necessities for nasal columella reconstruction should contain skin color, subcutaneous bulk, width, and transition zones at the nasal tip, base of the nasal columella, and nasal floor.\textsuperscript{1,9}

Techniques for reconstruction of nasal deformities continue to evolve as plastic surgeons strive to achieve the most natural nasal configuration. Numerous methods have been described for reconstructing the columella, especially in the manage-

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**Fig 1** Preoperative view of the patient.

**Fig 2** View of the patient 3 months after surgery.
ment of cleft lip nose deformity. The most common and striking feature of the cleft lip nose deformities is a short columella, and many techniques have been used to lengthen the columella. These techniques can be summarized as the flying buttress graft, which is made up of a single or paired spreader grafts secured to a columellar strut; caudal septal grafts and tip grafts of various shapes; radix grafts placed to elevate the nasion; and interposition grafts placed between the upper and lower lateral cartilages.

The absence of nasal columella is a more challenging situation than is short columella. Numerous methods have been described for reconstruction of the columellar defects. Skin grafts, composite grafts, forehead flaps, tube pedicle flaps, nasolabial flaps, nasomalar flaps, nasal septal flaps, randomized myocutaneous alar island flap, upper lip flaps, Abbe flaps, and composite free flaps from the ear have been used in repairing defects of the nasal columella.

The survival of composite ear grafts is a matter of concern. They are best applied in partial reconstruction of the columella. Forehead and tube pedicle flaps require several procedures, and the result generally is far from ideal. A forehead flap can be a good choice in the presence of a larger defect including the nasal tip. Local flaps are used frequently,
and they offer good color match, close proximity, and excellent blood supply. However, they often result in facial scars and bulky structures that require secondary revisions. Composite free flaps from the ear offer excellent color match and do not produce a scar in the central portion of the face. However, such flaps require microsurgical proficiency and longer operation times for relatively small defects.

In the current case, vestibular labial mucosal flaps were elevated from the vestibular side of the upper lip, leaving an invisible scar at the donor site (Fig 3). It is an appropriate tissue to replace the nasal septum mucosa, which is thin-walled and consists less subcutaneous fat. Another advantage is that this region has excellent blood supply, so it can easily nourish a cartilage graft inserted under the flap. Partial de-epithelization of the flap allowed avoidance of the second surgical step for pedicle division described by other authors. Using caudal septal flaps helped to avoid the use of an additional donor area to cover the cartilage strut graft.

REFERENCES