

A COLUMELLAR DEFORMITY CAUSED BY A CONGENITAL SCHWANNOMA

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Abstract

Schwannoma is a benign neoplasm arising from Schwann cells of the peripheral nerve. It very rarely located in the nasal tip. We report two cases of congenital schwannoma of the nasal columella and discuss the surgical approach of such tumor.

Case report: Case 1: An 18 year-old female was referred to us for a very slow growing tumor of the columella. The deformity had been present since the birth. The patient underwent an excision of its tumor using an open rhinoplasty approach. The histological examination revealed a schwannoma. No recurrence was found within 2 years of follow up.

Case 2: A 4 month male baby presenting a congenital tumor of his columella. He underwent an excision using open rhinoplasty approach. The histological examination showed a plexiform schwannoma.

Discussion: Schwannoma of the nasal tip is a benign tumor that gradually causes aesthetic and functional disorders. Congenital schwannoma of columella is an extremely rare clinical situation. Its diagnosis and treatment can pose certain challenges. The treatment is surgical excision and histological analysis of the specimen. Open rhinoplasty approach provided a good surgical exploration and a good cosmetic result on this nasal tip tumor.

Key words: nose neoplasms; neurilemmoma; rhinoplasty

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Introduction

Schwannoma is very rare in the nasal tip [1] and to the best of our knowledge congenital schwannoma of the nasal columella has never been reported. The diagnosis of such rare tumor is challenging and even if it is a benign neoplasm excision biopsy is essential to exclude sinister pathology [2]. We report a case of columellar schwannoma.

Case Report

Case 1:

A 18 year-old female, came to our service for a slow growing deformity of her columellar. The deformity had been present from the birth (Fig. 1).

Physical examination revealed a well-defined elastic tumor in of the anterior part of the columella measuring 1cm. The overlying skin was thin.

The patient underwent an excision of its tumor using an open rhinoplasty approach and a "V" shaped incision (Fig. 2). A superficial subcutaneous undermining allowed tumor removal. The medial crus of greater alar cartilage were slightly distorted. No nerve of origin was seen during the dissection. The limits of the excision were free of tumor, and histologic analysis of the tumor showed the characteristics of a

schwannoma. The patient remains having no recurrence within 2 years of postoperative follow up.

Case 2:

A 4 month-old baby was referred to us for a congenital rapidly growing tumor of the columella (Fig. 3). On examination he presents an oval firm, mobile, globular swelling of the columella measuring 15 mm by 5 mm. The tumor had extended to the upper part of the philtrum and had caused a partial obstruction of the right nostril. Excision of the tumor was performed using an open rhinoplasty approach. Pathological examination revealed the diagnosis of plexiform schwannoma.

Discussion

Developmental midline nasal masses in children are rare, with a reported annual incidence of one in every 20,000–40,000 live births [2]. They result from a failure of embryologic separation of neuroectodermal and ectodermal tissues during the development of the nose and frontobasal region. Many such lesions may include an intracranial extension or connection. Their differential diagnosis includes abscesses, hemangiomas, fibromas, lipomas, granulomas, and mucocoeles.



Figure 1. Tumor of the nasal columella



Figure 2. A "V" incision and open rhinoplasty approach to excise the tumor



Figure 3. Tumor of the nasal columella in 4 months baby.

Schwannomas are slightly less common than neurofibromas, but, like the latter, constitute about 5% of all benign soft-tissue tumors. Nasal tip location of schwannoma was first described by Bingham et al [3]. Seven cases of nasal tip Schwannoma have been reported in the literature and none of involving the only columella.

Schwannoma is a common tumor that can develop at any age. It grows slowly, expanding and applying pressure to the surrounding tissues, causing aesthetic and sensory problems [4]. Even if nasal tip schwannoma is a benign tumor it should be removed as soon as possible to avoid cartilage deformities. Radiologic examinations can assist in its diagnosis [5].

Magnetic resonance imaging (MRI) is especially useful for the diagnosis and can help also to evaluate the local effect of the tumor on the surrounding structures.

The open rhinoplasty approach is ideal for excision of the tumor in the nasal tip [6-8]. Schwannomas have a true capsule composed of epineurium, which allows their successful surgical resection [9]. The increased exposure makes it easier to perform certain technical manoeuvres for resection, and gives the surgeon the ability to diagnose the deformity of the osseocartilaginous framework.

Recurring tumors are rare (2%) when removal is complete, and are particularly associated with neurofibromatosis [10].

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