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Case Report

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Staged surgical excision of a premaxillary plexiform schwannoma causing nasal obstruction and deformity

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Abstract

While schwannomas of the head and neck are not uncommon, plexiform type schwannomas are rare with very few nasal cases described in the literature. We present a case of plexiform type schwannoma of the nasal tip and premaxilla that caused functional impairment of nasal breathing, upper lip dysmotility, and cosmetic deformity. Due to concern for preserving both form and function of the nose and lip, a staged surgical approach was used to ensure complete excision and maximize functional and aesthetic outcome.

Introduction

Schwannomas are a type of encapsulated neurogenic tumor composed of the cells that form the myelin sheath of nerves. Given that these benign tumors can occur in any central or peripheral nerve with associated Schwann cells, they can implicate any region of the body, but are most commonly encountered in the head and neck due to the high density of neural structures [1,2]. Patients typically present in middle age with a slow-growing tumor with site-dependent symptomatology resulting from compression or obstruction of surrounding structures. Given their resistance to radiotherapy, surgical excision is the primary treatment for these lesions [3].

In most cases, schwannomas are solitary and sporadic. If multiple schwannomas occur in an individual patient, consideration should be given to the syndromic causes of schwannomas including neurofibromatosis type II, schwannomatosis, and Carney complex [4]. Although these tumors show an exceedingly low likelihood of malignant degeneration, they can be locally disfiguring and occasionally cause functional impairment [5].

Plexiform type schwannomas affecting the nose are exceedingly rare. Few schwannomas of the nasal tip have been described in the literature [6-10]. We present a unique case of a nasal tip schwannoma causing nasal obstruction and cosmetic deformity, which was excised in a staged fashion to maximize both functional and cosmetic nasal outcomes despite a technically challenging tumor location.

Case presentation

An otherwise healthy 55-year-old woman presented with longstanding nasal obstruction and deformity, as well as several months of worsening upper lip motor control. She had a remote history of an unspecified nasal tip mass which was excised via an unknown endonasal surgery at age 18. On exam there was a soft, mobile mass at the base of her columella, which was deviated to the right (Figure 1). The mass extended superiorly into the left ala and inferiorly into the upper lip. A weakened left lower lateral cartilage had led to static and dynamic nasal valve collapse. At this time, differential diagnoses included benign nasal masses such as neurofibroma, papilloma, granuloma, dermoid, leiomyoma, chondroma, hemangioma, and schwannoma. To maximize functional and aesthetic outcomes, the decision was made to proceed with excision in a staged fashion. This allowed the surgeons to address the nasal obstruction and lip dysmotility meticulously and effectively while allowing adequate cosmetic improvement.

Stage one of the excision included a sublabial approach with submucosal dissection off the premaxilla and nasal spine. The

mass had infiltrated the deep side of orbicularis oris, causing expansion and ptosis of the white lip. The muscle had thinned due to this expansion, but no surgical interventions were performed at this stage so as to wait until the defect was finalized after complete excision during the second stage. Final pathology demonstrated a 3 x 2.5 cm plexiform schwannoma extending to the specimen edge where it had been truncated.

A second stage surgery three months later included open septorhinoplasty with gross total removal of the residual 5 x 1 cm plexiform schwannoma from the columella and left ala (Figure 2). Lateral osteotomies were performed for dorsal repositioning after removal of the mass. Revision septoplasty and bilateral inferior turbinate reduction were performed to maximize nasal breathing. Caudal septal extension and alar batten grafts were utilized with cadaveric rib cartilage, as the patient did not have adequate septal cartilage for grafts following the endonasal surgery she had as a child. A lip-lift myectomy was also performed at this stage, with excision of the ptotic portion of orbicularis oris and local advancement to resuspend the white lip. The patient had resolution of nasal obstruction, improvement in upper lip movement with less lip laxity, and a satisfactory cosmetic outcome. At one-year post-operative follow up there was no evidence of recurrence (Figures 2 & 3).

Discussion

Schwannomas are the most common benign peripheral nerve sheath tumor and occur with increased frequency in regions of the body with a high density of neural tissue, such as the head and neck [1]. Subtypes of schwannomas include: conventional, cellular, melanotic, and plexiform [11]. First described in 1978, the plexiform type schwannoma is rare, comprising approximately 4.3% of all schwannomas and most commonly affecting the soft tissues of the scalp and neck [1]. Plexiform type schwannomas are differentiated from other subtypes by their multinodular, intra-neural growth pattern. They are diagnosed based on histopathology, which shows diffusely S-100 immunoreactivity, a multinodular plexiform growth pattern of Schwann cells, and a predominance of Antoni Type A areas with hypercellularity resembling a plexus or “bag of worms” [1].

These tumors must be distinguished from their counterpart: The plexiform neurofibroma. Whereas plexiform schwannomas are comprised exclusively of Schwann cells, show no malignant potential, and can be associated with neurofibromatosis type 2, the plexiform neurofibroma is comprised of multiple different cell types, can undergo degeneration to a malignant peripheral nerve sheath tumor, and is commonly associated with neurofibromatosis type 1 [1]. Despite no malignant transformation, surgical excision is the primary treatment modality of plexiform type schwannomas due to local mass effect and deformity.

The plexiform schwannoma involved in this case was at the nasal-lip junction, an area of great functional and aesthetic importance. Single-staged excision would have required extensive dissection of the nasal tip and resultant loss of tip and spine support. Additionally, it would require dissection into the upper lip and thinning of the orbicularis oris that could worsen the patient’s already poor lip movement. Instead, a staged surgical approach offered the benefit of ensuring complete excision of the tumor alongside optimization of functional and aesthetic outcomes by addressing the lip and nose individually.

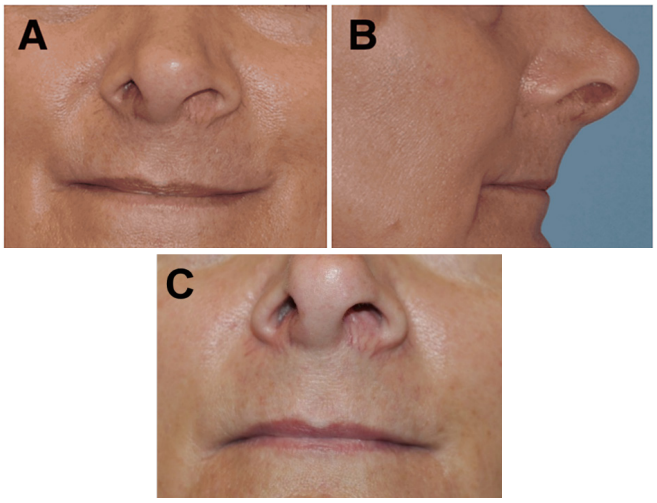

Figure 1: Pre-operative photographs.
(A, B, C) Pre-operative photographs demonstrate spongy mass of the premaxilla and nasal tip, with deformity of the nasal valves, columella, and upper lip.

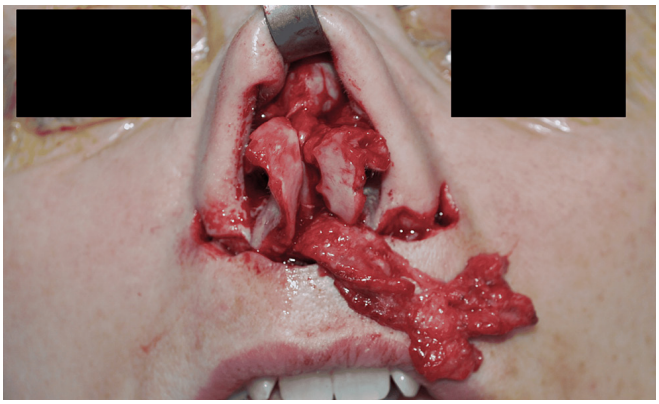

Figure 2: Intraoperative photograph during open septorhinoplasty. A large 5 x 1 cm mass extending from the premaxilla, which was dissected out of the columella, nasal tip, and left ala during open septorhinoplasty.

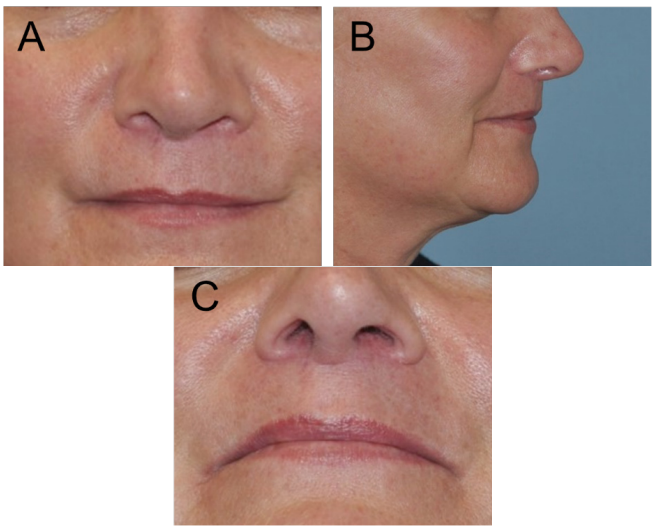

Figure 3: Post-operative photographs.
(A, B, C) One-year post-operative photographs demonstrate improvement in deformity of nasal tip, columella, external nasal valve, premaxilla, and upper lip/orbicularis oris.

Surgical management of masses located in the nasal tip relies on thoughtful consideration of the tip support structures and the way they interact with each other to form the nasal contour, projection, and rotation. The major support structures of the nasal tip include the scroll region, the inherent strength of the lower lateral cartilages, and the paired medial crura where they attach to the caudal septum [12]. Minor support structures include the dorsal cartilaginous septum, interdomal ligaments, membranous septum, nasal spine, skin and soft tissue, and the alar sidewalls. Failure to maintain nasal tip structural support by a combination of these mechanisms can result in tip ptosis, one of the leading adverse outcomes in rhinoplasty [13]. This patient had a deformity of the soft tissues of the nasal tip, with resection of the columellar and alar portion of her mass causing notable tip ptosis and de-rotation that was addressed with a caudal septal extension graft. The nasal tip and external nasal valves were further remedied with placement of alar batten grafts, which provided strength to the lower lateral cartilages and improved the inherent strength of the structural framework.

Staging the excision of this tumor made it possible to address the aesthetic subunits of the nasal tip, ala, and upper lip individually and to optimize function of the nose and lip, ensuring appropriate nasal breathing and oral competence. The patient had resolution of nasal obstruction, improvement in upper lip movement, and a satisfactory cosmetic outcome. This case is made noteworthy by the uniqueness of the pathology, the aesthetic and functional considerations in surgical management, and the unique staged surgical approach.

Conclusion

Benign masses of the nose can cause significant deformity and dysfunction, especially if located at the nasal-lip junction. In such instances, a staged surgical approach should be considered to ensure maximal excision and reconstruction, with attention to both functional and aesthetic nasal outcomes. We present a unique case of a rare schwannoma subtype that caused nasal obstruction, lip dysmotility, and cosmetic deformity. This case highlights the utility of a staged approach to management in form and function.

References

1. Berg J, Scheithauer B, Spinner R, Allen C, Koutlas L. Plexiform schwannoma: A clinicopathologic overview with emphasis on the head and neck region. Human Pathology. 2008; 39: 633-640.
2. Guerrissi J. Solitary Benign Schwannomas in Major Nerve Systems of the Head and Neck. Journal of Craniofacial Surgery. 2009; 20: 957-961.
3. Wang B, Yuan J, Chen X, Xu H, Zhou Y, et al. Extracranial nonvestibular head and neck schwannomas. Saudi Medical Journal. 2015; 36: 1363-1366.
4. Pathmanaban O, Sadler K, Kamaly-Asl I, King A, Rutherford S, et al. Association of Genetic Predisposition With Solitary Schwannoma or Meningioma in Children and Young Adults. JAMA Neurol. 2017; 74: 1123-1129.
5. Nayler S, Leiman G, Omar T, Cooper K. Malignant transformation in a schwannoma. Histopathology. 1996; 29: 189-192.
6. Mohan D, Krishna A, Ramakristnan V. Schwannoma presenting as a nasal tip deformity. Ann Plast Surg. 1997; 38: 83-84.
7. Lemmerling M, Moerman M, Govaere F, Praet M, Kunnen M, et al. Schwannoma of the tip of the nose: MRI. Neuroradiology. 1998; 40: 264-266.
8. Fujita K, Yuzuriha S, Noguchi M. 'Cyrano de Bergerac' nose deformity caused by the schwannoma of the nasal tip. J Plast Reconstr Aesthet Surg. 2006; 59: 1252-1254.
9. Skouras A, Skouras G, Diab A, Asimakopoulou F, Dimitiradi K, et al. Schwannoma of the nasal tip: Diagnosis and treatment. Aesthetic Plast Surg. 2011; 35: 657-661.
10. Ferraz R, Duarte J, Fujita R, Pignatari S. Plexiform Schwannoma of the nasal tip: surgical approach. Brazilian Journal of Otorhinolaryngology. 2012; 78: 134-134.
11. Kurtkaya Y, Scheithauer B, Woodruff J. The pathobiologic spectrum of schwannomas. Histology and Histopathology. 2003; 18: 925-934.
12. Han S, Lee D, Kim J, Kim W. An anatomic study of nasal tip supporting structures. Ann Plast Surg. 2004; 52: 134-139.
13. Margulis A, Harel M. Management of severe tip ptosis in closed rhinoplasty: The horizontal columellar strut. J Plast Reconstr Aesthet Surg. 2007; 60: 400-406.