CASE REPORT

A patient with periorbital pain: A case of isolated schwannoma of the eyebrow

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Abstract: Schwannomas rarely occur around the orbit and they commonly arise in patients between ages 20 and 70 as unilateral masses with well-defined borders and with a rather slow growth rate. There are only a few case reports with lower and upper eyelid involvement in the English-language literature. In this report, an isolated schwannoma case with eyebrow involvement on the trajectory of the supraorbital nerve in a 24-year-old male patient is presented.

Keywords: Schwannoma; eyebrow; supraorbital nerve


Introduction

Schwannomas, first described by Uruguayan neuropathologist Jose Verocay in 1908, are benign tumors arising from the neuroectodermal cranial, intraspinal, peripheral or autonomous nerve sheaths. Schwannomas rarely occur around the orbit and they commonly arise in patients between ages 20 and 70 as unilateral masses with well-defined borders and with a rather slow growth rate¹. Although these tumors can develop in any part of the orbit, they most commonly involve the ophthalmic branch of the trigeminal nerve. However, schwannomas can also arise from the 3rd, 4th and 6th cranial nerves as well². It has been reported that some hereditary diseases such as neurofibromatosis, schwannomatosis and Carney complex are associated with multiple schwannomas³. There are only a few case reports with lower and upper eyelid involvement in the English-language literature⁴–¹⁵. In this report, an isolated schwannoma case with eyebrow involvement on the trajectory of the supraorbital nerve in a 24-year-old male patient is presented.

Case report

A 24-year-old male patient was admitted to our outpatient clinic with complaint of a painful mass at the medial part of the eyebrow. In his medical history, he stated that the mass had grown slowly over the past four years. Physical examination revealed a mobile, hard-nodular lesion measuring 4 × 2 cm. No café au lait spots, axillary or inguinal freckles, and neurofibromas suggesting neurofibromatosis were observed during the dermatological examination of the patient. Ophthalmological examination revealed a 20/20 visual acuity and normal eye movements in each eye. During fundus and anterior segment examinations, no Lisch nodules — which are among the diagnostic criteria of neurofibromatosis — were detected. In computerized tomography and orbital magnetic resonance imaging, a mass located on the anterior of supra-orbital rim was detected. The mass was...
reported to be extraorbital, homogenous, having distinct borders and a capsule, and measuring at 39 × 21 mm. It was isodense to muscle tissue. No sphenoid wing dysplasia, pseudarthrosis or thinning of long bone cortex was detected.

Total excision of the mass was performed in a plastic surgery clinic, preserving the supraorbital and supratrochlear nerve, levator palpebre superioris and orbicularis oculi muscle. There was no functional and sensory impairment of the eyelid and eye brow movements (Figure 1). Histopathological examination of the mass reported that it was consistent with schwannoma, containing compact hypercellular Antoni A areas and myxoid hypocellular Antoni B areas. Intense positivity was observed for S-100 protein immunohistochemical staining. It was detected that the cells were narrow, elongated and wavy, and Verocay bodies were observed in cellular areas (Figures 2A and 2B). There was no complication post-operation, and no relapse developed after one-year follow-up of the patient.

Discussion

Ninety percent of the schwannomas are solitary and isolated. Common clinicopathologic variants of schwannomas include conventional, cellular, cystic and melanocytic schwannoma. Schwannomas arise from the proliferation of periaxial or endoneural Schwann cells. Although a defect or loss in type-2 NF gene located in the chromosome 22 has been implicated for the Schwann cell proliferation, its real cause remains unknown.

Schwannomas have the appearance of solitary, soft, pink-yellow, smooth nodule or tumor localizing in deep dermis or subcutaneous tissue. They are almost always found within a capsule. Their dimensions can range between 0.5 cm and 3 cm. They most commonly involve the flexor surfaces of the extremities, followed by the wide nerve bodies along the head and neck region. The tumor cells have a cylindrical or spindle-shaped appearance, and they are usually wavy. They consist of hypercellular (Antoni A) and hypocellular (Antoni B) areas. Hard schwannomas appear tightly organized in under macroscopic examination and contain intense Antoni A areas. Their nuclei comprise palisades of basophilic Schwann cells divided by bright eosinophilic areas (Verocay bodies). Soft schwannomas are edematus; they are composed mostly or completely of Antoni B tissue characterized by loose connective tissue and ectatic blood vessels. Schwannoma is stained positive for S-100 and negative for epidermal membrane antigen (EMA) in immunohistochemical examination.

Differential diagnosis of schwannoma includes lipoma, fibroma, xanthoma, hemangioma, hemangiopericytoma, leiomyoma, fibrosarcoma, leiomyosarcoma, solitary fibrous tumor, adnexal tumors and meningiomas. Differentiation from these tumors can be challenging because schwannomas are also painless, mobile and soft. When cystic degeneration is present, schwannomas can be confused with mucocele, dermoid cyst, pyocele, meningoencephalocele, hematocoele, epidermal inclusion cyst, ganglione and teratoma.

Detection of the origin of the tumor can be difficult due to its complex orbital anatomy. Nerve origin can be found in only 32% to 47% of orbital schwannomas. The tumor is mobile on the horizontal plane of the nerve line, while it is frequently fixated on a vertical plane. Although it is commonly manifested by painless proptosis, it can cause complaints of numbness or findings imitating sinusitis, as in our case. When found around the eye globe, the localization of the tumor becomes important because of the movement of the globe. Di-

Figure 1. Perioperative view of the case

Figure 2. (A) Compact hypercellular Antoni A areas and myxoid hypocellular Antoni B areas. Cells are narrow, elongate, and wavy with tapered ends interspersed with collagen fibers. Nuclear palisading around fibrillary process (Verocay bodies) are often seen in cellular areas (Hematoxylin and eosin (H&E), 200X); (B) S-100 staining of the lesion.
plopia and eccentric proptosis are among the common findings in intraconal lesions. Similar to that of our case, however, extraorbital tumors result in minimal changes in orbital structures because they are slow-growing masses. Since most cases with schwannomas exhibit supraorbital and supratrochlear nerve localization, the globe is displaced downwards. The tumor rarely arises from infraorbital nerve, and pushes the globe upwards.

The treatment of extraorbital schwannomas involves total excision of the mass with its capsule. In some cases, adhesion of the mass to adjacent tissues does not permit total resection and tumor relapse can be encountered. Prognosis is good and relapse rate is low in patients who undergo complete surgical excision.

**Conclusion**

Schwannomas are uncommon tumors. Preoperative diagnosis can be challenging because of the variability of locations and the absence of pathognomonic characteristics. It is important to perform early diagnosis with CT scan and orbital MRI followed by early intervention in suspected lesions during early stage in order to prevent serious complications.

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**Conflict of interest**

The authors declare no potential conflict of interest with respect to the research, authorship, and/or publication of this article.

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