Trigeminal Ophthalmic Branch Schwannoma: Case Report and Literature Review

Schwannoma trigeminal de ramo oftálmico: Relato de caso e revisão da literatura

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Abstract

Schwannomas are the fourth most common primary neoplasms affecting the brain and cranial nerves. Central lesions commonly arise from sensory nerve roots, and a common intracranial site is the vestibular branch of the 8th nerve (>85%). We present the case report of a patient who has a schwannoma extending from the pterygopalatine fossa to the orbit, complaining about facial pain in the trajectory of the trigeminal ophthalmic branch. Schwannomas represent 1 to 2% of all neoplasms of the orbit, and trigeminal schwannomas are extremely rare, accounting for less than 0.5% of all intracranial tumors.

Keywords

- schwannoma
- orbital schwannoma
- ophthalmic schwannoma
- endoscopic transsphenoidal neurosurgery
- subciliary approach

Resumo

Schwannomas são a quarta neoplasia primária mais comum que afeta o cérebro e os nervos cranianos. As lesões centrais comumente surgem de raízes nervosas sensitivas, e um sítio intracraniano comum é o ramo vestibular do oitavo nervo (mais de 85% dos casos). Apresentamos o relato de caso de um paciente portador de schwannoma que se estende da fossa pterigopalatina até a órbita, com queixa de dor facial no trajeto do ramo oftálmico do nervo trigêmeo. Schwannomas representam 1–2% de todas as neoplasias da órbita e schwannomas trigeminais são extremamente raros, respondendo por menos de 0,5% de todos os tumores intracranianos.

Palavras-chave

- schwannoma
- schwannoma orbital
- schwannoma oftálmico
- neurocirurgia endoscópica Transesfenoidal
- abordagem subciliar

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Introduction

Schwannomas are the fourth most common primary neoplasms affecting the brain and cranial nerves. Central lesions commonly arise from sensory nerve roots, and a common intracranial site is the vestibular branch of the eighth nerve (> 85% of central schwannomas and 8–10% of all intracranial tumors.).[1–4] We present the case report of a patient who has a schwannoma extending from the pterygopalatine fossa to the orbit, complaining about facial pain in the trajectory of the trigeminal ophthalmic branch. Primary orbital tumors are very rare, with an overall incidence < 1 per 100,000/year;[5] schwannomas represent 1 to 2% of all neoplasms of the orbit,[4,6] and trigeminal schwannomas are extremely rare, accounting for 0.07 to 0.3% of all intracranial tumors.[7,8] The patient usually presents with facial pain, numbness, and paresthesia in the distribution of one or all the divisions of the trigeminal nerve depending on the location of the tumor.[7,9,10] Magnetic resonance imaging (MRI) is the gold standard for evaluation because of its multiplanar capabilities and better soft-tissue contrast, being useful for planning the surgical approach.[7,9]

Case Report

A 56-year-old female, diabetic, hypertense, who had a hemorrhagic stroke 7 years ago, sought the emergency service complaining about left hemicranial progressive headache, for 3 years. Neurological exam at the admission showed right spastic hemiparesis, Medical Research Council (MRC) grade III, right hyperreflexia and Wernicke-Mann posture. The patient underwent an MRI, which showed an expansive formation involving the left pterygopalatine fossa and the sphenoid bone, heterogeneously enhanced by gadolinium in T1 sequence, extending to the extraconal lateral region of the left orbit, with a rounded aspect measuring 1.9 cm in its largest diameter, hyperintense in T2 and without impregnation by gadolinium (►**Fig. 1**). The patient underwent a transsphenoidal surgery using an endonasal endoscopic

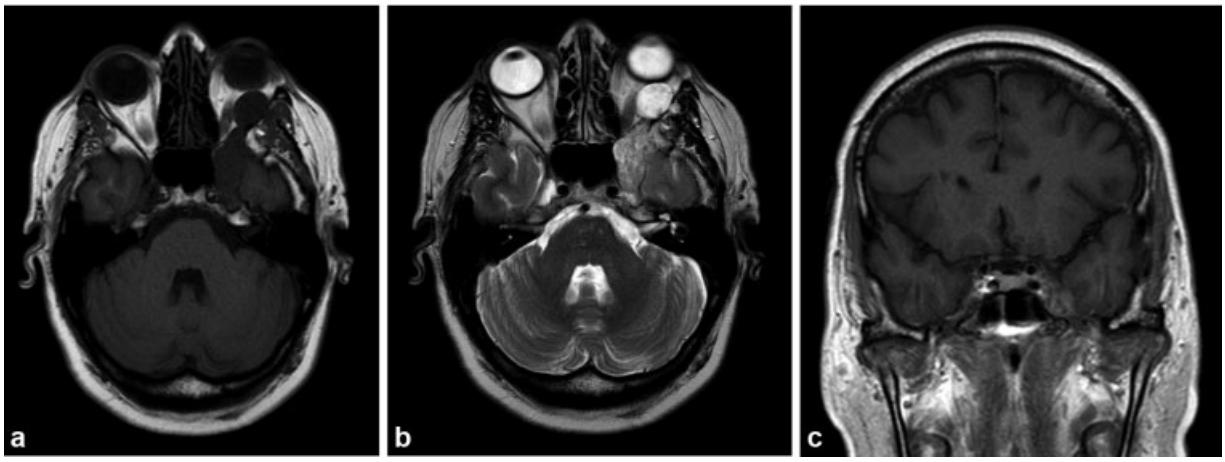

Fig. 1 Preoperative images. (**a**) Axial T1-weighted gadolinium-enhanced image with a rounded aspect measuring 1.9 cm in its largest diameter, without impregnation by the gadolinium. (**b**) Axial T2-weighted image, showing a hyperintense lesion. (**c**) Coronal T1-weighted gadolinium-enhanced image.

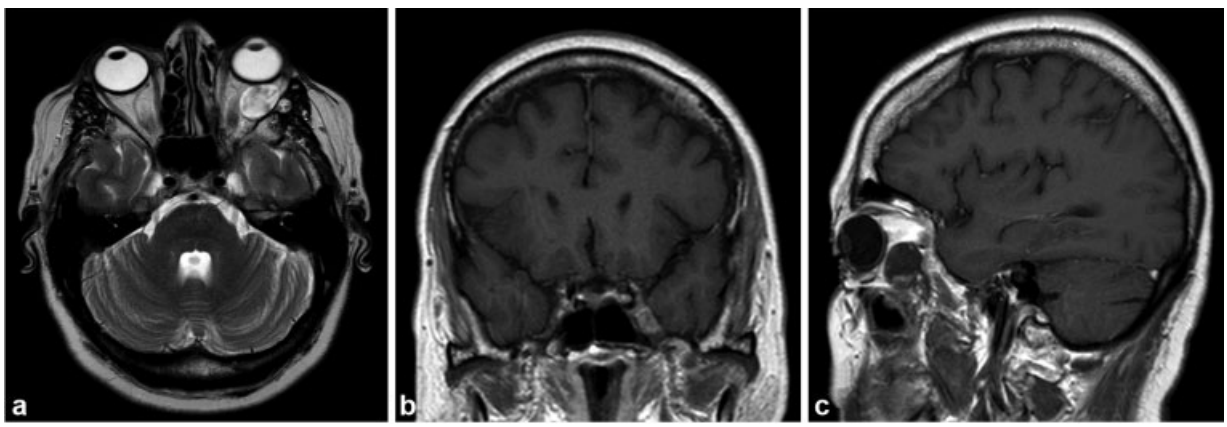

Fig. 2 Postoperative images, after the first surgery. Magnetic resonance imaging still shows an orbital lesion. (**a**) Axial T2-weighted image, still showing the hyperintense lesion in the extraconal lateral region of the left orbit. (**b**) Coronal T1-weighted gadolinium-enhanced image, showing the resection performed in the first surgery. (**c**) Sagittal T1-weighted gadolinium-enhanced image from tumor, measuring 1.9 cm in its largest diameter, without impregnation by the gadolinium.

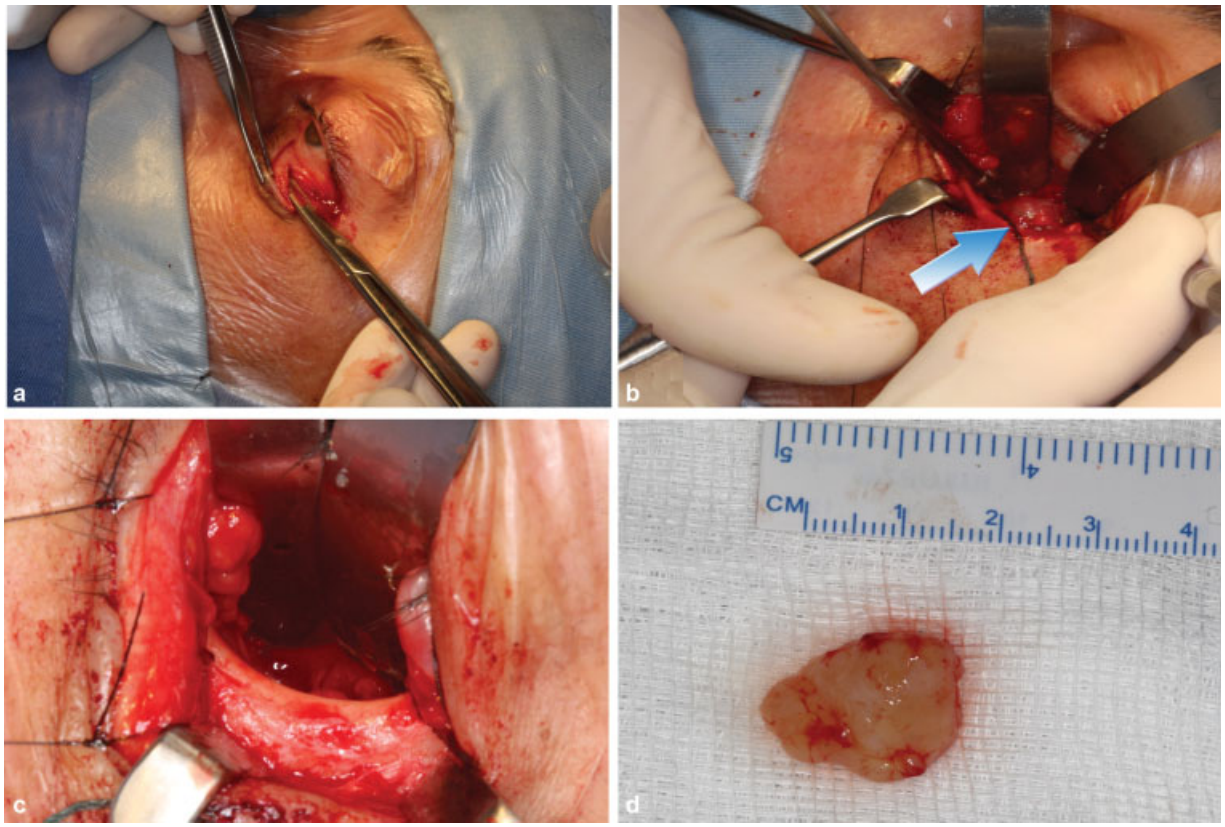

Fig. 3 Intraoperative images, from the second surgical procedure for resection of the orbital portion of the tumor. (**a**) Transconjunctival approach. (**b**) Lesion exposure (blue arrow) with partial resection (**c**) Revision of the tumor cavity evidencing complete resection of the tumor. (**d**) Surgical specimen measuring 1.8 cm × 1.2 cm × 0.5 cm.

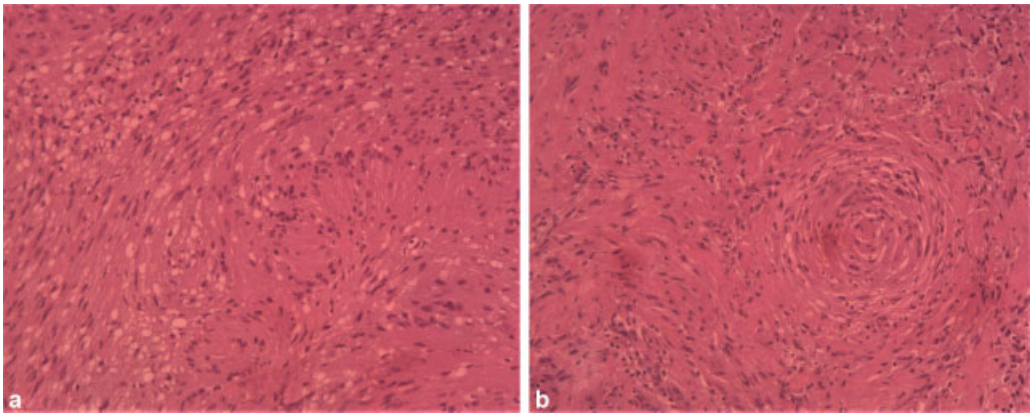

Fig. 4 Second histopathological examination, also conclusive for schwannoma. (**a**) Hematoxylin & Eosin stain (5x) – neoplasia consisting of fusocellular cells, Antoni A and Antoni B areas. (**b**) Hematoxylin & Eosin stain (10x) – neoplasia consisting of fusocellular and monomorphic cells, without atypia and without presence of mitosis.

approach for total resection of the sphenoidal portion of this lesion. The histopathologic examination was conclusive for schwannoma. The postoperative MRI showed the total resection of this portion, leaving just the orbital portion (►**Fig. 2**). After 2 years, the patient started to complain about pain in the trajectory of the ophthalmic branch of the trigeminal root and was submitted to a transconjunctival approach for resection of the orbital lesion, combined with an orbital reconstruction (►**Fig. 3**). The histopathologic examination was also conclusive for schwannoma (►**Fig. 4**). After this procedure, the patient became asymptomatic, with no referred facial pain, and without new deficits.

Discussion

According to the 2016 World Health Organization Classification of Tumors of the Central Nervous System, schwannomas include cellular, plexiform, and melanotic schwannomas, neurofibromas, perineuriomas, hybrid nerve sheath tumors

and malignant peripheral nerve sheath tumors.[11] They affect adults in the 3rd to the 7th decades of life[12] but are more commonly found in patients in their 4th decade, without sex predilection.[13] This type of tumor comprises the fourth most common primary neoplasm affecting the brain and cranial nerves.[14] The majority ($\sim$ 90%) of cranial nerve schwannomas arise from the vestibulocochlear nerve, with the next most commonly involved nerves being the trigeminal and facial nerves, followed by the lower cranial nerves.[15] The orbital nerves more commonly involved are the supratrochlear and supraorbital nerves.[16]

Schwannomas constitute 1 to 8% of all head and neck tumors.[17] When intraorbital, it is a rare tumor that accounts for $\sim$ 1 to 2% of all neoplasms of the orbit.[4,6] They are known to originate from sympathetic and parasympathetic fibers, as well as from branches of the oculomotor, trochlear, trigeminal, and abducens nerves, with the ophthalmic division of the trigeminal nerve being the most common.[18] Trigeminal schwannomas are particularly rare, accounting for 0.07 to 0.3% of all intracranial tumors and 0.8 to 5% of intracranial schwannomas. Orbital schwannomas, most commonly, arise from the sensory branches of the trigeminal nerve.[4] They may arise from the cisternal segment (most commonly), the trigeminal/Gasserian ganglion in Meckel's cave, or from one of the three branches of the nerve. They tend to occur predominantly in the 4th to 6th decades of life and are slightly more common in females.[7] The oculomotor, ciliary, lacrimal, and zygomaticotemporal nerves have been reported as the nerves that the orbital schwannomas most commonly arise from.[17] The frontal nerve, in particular, is frequently affected due to the predominance of sensory nerve branches, namely the supraorbital and supratrochlear nerve.[17,18] Based on the work of Erdogmus et al, Irace et al proposed that the entry point of nerve fibers in the muscle (e.g., area nervosa or myoneural junction) is the zone where intraorbital schwannomas truly arise.[4] Motor root and sympathetic chain involvement is uncommon, and involvement of the brain or cord parenchyma is rare.[1] Also, it is unusual for orbital schwannomas to arise from extraocular muscles, with only a few reports in the literature to date.[12]

Schwannomas are benign tumors consisting of a clonal population of Schwann cells, which often undergo cystic and degenerative change.[1] They originate in a zone of transition of peripheral central myelin (Obersteiner-Redlich zone)[19] and are usually isolated, solitary, slow-growing, and well-encapsulated lesions, except when they are associated with neurofibromatosis. In the latter case, the patients can develop multiple schwannomas, in which case the condition is termed schwannomatosis.[13] Persons with neurofibromatosis type 2 characteristically develop bilateral vestibular schwannomas, but approximately half of them also have nonvestibular schwannomas, which most commonly involve the oculomotor and trigeminal nerves.[15] Although hereditary disorders predispose to nerve sheath tumors, the specific etiologies of schwannoma remain unknown. However, sporadic reports have suggested an association of formation of schwannomas with previous events of trauma.[13] Postradiation schwannomas, including acoustic schwannomas, appear with a latency of up to 50 years following treatment.[1] Besides, they are generally known to be slow-growing, benign masses. There have been clinical reports of rapidly growing schwannomas in pregnant women, attributed to progesterone receptors or intratumoral hemorrhage.[12]

When the imaging features and clinical signs are compatible with schwannoma, the presence of end organ compromise, such as denervation-induced muscle atrophy or sensory deficit, may aid in the identification of the cranial nerve (CN) of origin.[15] Since most of the tumors originate from the sensory nerves, they do not interfere with the eye movements or vision unless they are located in the orbital apex or compress the optic nerve.[17] Clinically, trigeminal schwannomas usually present with facial pain, numbness, and paresthesia in the distribution of one or all the divisions of the trigeminal nerve depending on the location of the tumor,[7,9,10] what seems to fit in our case report. Headache and diplopia can also be present.[20] Long-standing tumors may also present with motor symptoms like difficulty in chewing and deviation of the jaw.[7] Yoshida and Kawase showed 6.3% of asymptomatic patients (found incidentally).[21]

Both MR and CT images show evidence of the slow growth of schwannomas, including smooth expansion of the neural foramina, osseous remodeling, and/or deformation of adjacent brain tissue, with a disproportionately small amount of edema, given the size of the lesion.[15] Schwannomas have a characteristic appearance on MR imaging: on T2-weighted images, schwannomas appear heterogeneously hyperintense.[4,15,22,23] This heterogeneity is attributed to regions of compactly arranged cells (Antoni type A) mixed with regions of loosely arranged cells (Antoni type B), with variable cellularity and water content.[15] Usually, the central enhancement seems to represent the presence of hypercellular Antoni A-type cells in the central part of the tumor and hypocellular Antoni B-type cells in the periphery.[24] On T1-weighted images, these lesions have low or intermediate signal intensity and demonstrate avid enhancement after contrast material administration, with or without nonenhancing cystic spaces. Larger lesions commonly have heterogeneous enhancement, cystic spaces, and foci of hemosiderin due to internal hemorrhage.[15] On CT scan, they usually appear as uniformly enhancing masses with remodeling of the adjacent bone.[7,10]

Several classification systems have been proposed for a systematic approach in selecting the optimal surgical strategy. The most apt system was proposed by Wanibuchi et al based on 4 anatomical categories of tumor: peripheral, ganglion, root, and dumbbell. The peripheral type refers to tumor along V1 in the orbit, V2 in the pterygopalatine fossa (PPF) and maxilla, and V3 into the infratemporal fossa.[20,25] The Gasserian ganglion was the most frequent site described by Wanibuchi et al, and the most common type of extracranial extension (possibly corresponding peripheral or dumbbell by this classification) was from the third division described by Goel et al.[20,26]

Standard treatment for schwannomas consists of complete surgical resection, whereas subtotal resection is linked to an increased risk of recurrence,[25] which could reach 13-

fold higher.[23,27] Nevertheless, total tumor removal is not always feasible without neurological complications. In this circumstance, stereotactic radiation technique has emerged as an alternative treatment to surgical resection.[25,28] This has been shown to provide 5-year progression-free survival of 95% in selected patients.[25] With improvement of advanced surgical technique, total tumor removal is accomplished in 40 to 80% (Sharma et al, 2008; Zhang et al, 2009) of the cases, with tumor control rate of 81 to 100% (Lee et al, 2001; Al-Mefty et al, 2002; Goel et al, 2003; Kadri et al, 2004; Bulsara et al, 2008).[28] Surgical resection is also the best treatment for orbital schwannomas. Subciliary approach is the most preferred method for the masses inferior and medial to the optic nerve.[17] To access the medial inferior quadrant of the orbit, one option is endoscopic access, which is less invasive than the transcranial route. For small lesions located in the anterior half of the orbit, approaches without osteotomies, like eyelid superiorly or subciliary and orbital rim inferiorly can be tried, but anterior approaches give a restricted vision field, which promotes difficulties in locating and preserve the anatomical structures.[2,5,29]

Even so, the surgical approach must be determined case by case. Raza et al showed results suggesting that endoscopic transpterygoid approaches can help to achieve optimal resection rates with limited CN morbidity for tumors isolated to the Meckel's cave, with combined Meckel's cave peripheral extension, or primarily extracranial in location. According the Raza et al[25], endoscopic transpterygoid approaches provide safe access to Meckel's cave and disease extending along V2 into the Pterygopalatine fossa (PPF) and V3 into the infratemporal fossa without risking additional CN morbidity.[25] When complete resection is not possible, adjuvant radiotherapy can be considered.[2] The authors also reported significant rates of dry eye and corneal neurotrophic keratopathy that should be considered in those patients with either preoperative V1 neuropathy or tumor extending along this division. The presumed mechanism of these complications would be attributable to either vidian nerve injury (if the vidian is transected) or a V1 neuropathy. While an endoscopic transpterygoid approach to Meckel's cave is typically supravidian and does not require the nerve's sacrifice, transposition of the PPF contents could theoretically result in vidian nerve injury.[25] Pain ($>$ 90% of the patients), dysesthesia, and diplopia may be relieved after surgery; however, hypesthesia frequently remains or may be worsened by surgery.[20]

Conclusion

Schwannomas are benign and highly treatable lesions. When trigeminal, from the ophthalmic branch, they are extremely rare lesions, usually obeying the symptomatology of the nerve path. The approach should be individualized according to the morphology of the lesions and nerve involvement, always looking for total resection or subtotal in association with radiation therapy.

Conflict of Interests

The authors declare that there is no conflict of interests.

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