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# Orbital schwannomas. About a case and review of the literature

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## ABSTRACT

Orbital schwannomas are rare, benign neoplasms that develop from Schwann cells in the peripheral nerve sheath of the orbit. These slow-growing tumours are characterised by insidious proptosis, visual disturbances, diplopia, and occasionally pain. Following surgical resection, imaging modalities such as MRI and histopathological analysis are frequently used to make the diagnosis. The primary treatment modality remains surgical excision, with the goal of removing the tumour completely while preserving visual function and minimising postoperative complications. The prognosis for orbital schwannomas is favourable, with low recurrence rates after resection. However, long-term follow-up is required to detect any signs of recurrence or malignant transformation. More research is required to improve our understanding of the pathogenesis, optimal management strategies, and outcomes associated with orbital schwannomas, thus improving patient care and outcomes in this rare.

Keywords
orbital schwannoma, orbital tumours

## INTRODUCTION

Schwannomas are slow-growing, benign peripheral nerve sheath tumors that develop from Schwann cells. Orbital schwannomas are extremely rare, representing only 1% of all orbital neoplasms [1]. These tumors usually originate from sensory nerves that traverse the orbit, particularly the supraorbital and supratrochlear nerves. They are solitary tumors with distinct borders [2]. Clinically, orbital schwannomas usually occur in the second to fourth decade of life and develop within several months to years [3]. The main presentation of orbital schwannomas is slow increasing painless ocular proptosis caused by the indolent nature of their growth [4]. Orbital schwannomas are frequently asymptomatic when they are small. However, tumor growth may induce compression on the nerve from which the tumor originated, as well as to adjacent orbital structures. In this article, we report the case of an orbital schwannoma in a 45 years old woman.

## CASE REPORT

The patient, a 33-year-old man, presented with a chief complaint of right exophthalmia, which had been gradually worsening over the past year. He reported discomfort in his right eye, along with a sensation of

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pressure and mild pain. Ophthalmological examination revealed right exophthamia, reactive isocore pupils, visual acuity at 10/10 in both eyes.

The magnetic resonance imaging showed a benign extraconic mass, indistinguishable of the superior rectus muscle [img.1].

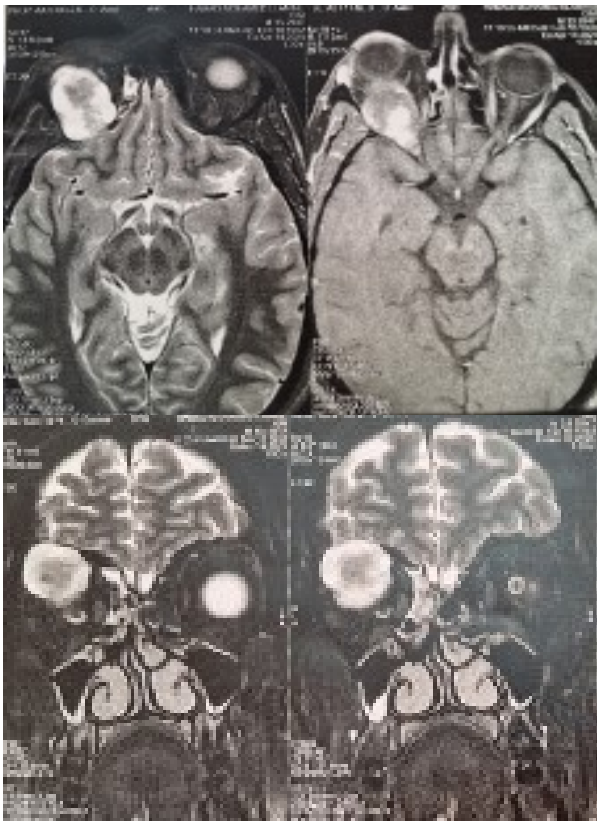

**Image 1.** MRI showing an intraorbital mass of the right orbit.

The patient went through surgery, a gross total removal of the tumor was processed using a unifrontal approach [img.2]. The surgical outcome was favorable.

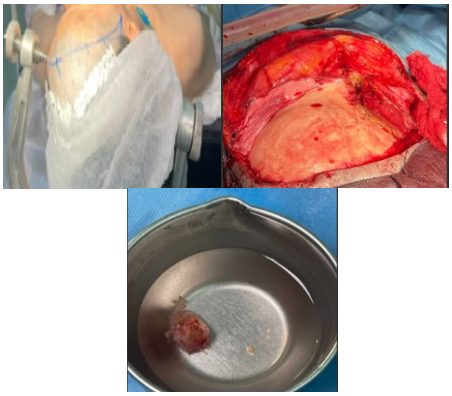

**Image 2.** Intraoperative images.

## Discussion

Orbital masses encompass a wide range of conditions, from benign growths to potentially fatal tumors, necessitating a thorough evaluation to distinguish and manage them effectively. These masses develop from a variety of sources, displaying a wide range of clinical symptoms that highlight the crucial importance of proper classification and precision treatments. [6]

Middle-aged women are more likely to develop optic nerve sheath meningiomas, which are characterized by gradual, painless vision loss and proptosis. [3] Schwannomas are often found in people between the ages of 20 and 70. [4] These tumors may initially be asymptomatic when tiny, but when they grow within the orbit, they cause painless proptosis. The clinical appearance of schwannomas differs greatly depending on their origin and location. These tumors, which most usually arise from branches of the supraorbital or supratrochlear nerves, cause the globe to move lower.[5] Larger schwannomas, particularly those originating from the orbital section of the third, fourth, or sixth cranial nerves, may cause diplopia. Lesions at the orbital apex may resemble retrobulbar neuritis, confounding diagnosis. [7]

Orbital schwannomas are difficult to diagnose just through imaging because they have no key diagnostic indicators. As a result, many surgeons use an incisional biopsy to make a histopathology-based diagnosis. Given the significance of identifying orbital schwannomas from other orbital cancers such as cavernous hemangiomas and meningiomas, several studies have attempted to define the differential radiographic patterns found in these tumors.

Orbital schwannomas present on ultrasonography as well-defined round or oval masses with low to medium internal reflectivity.[2]

Computed tomography (CT) is commonly used to analyze orbital tumors.[8] Pre-contrast, orbital schwannomas present as iso- to hypodense. Chen et al found that 81% of ocular schwannomas had iso- to hypodense oval or rounded soft tissue lesions. [8] Another study discovered this trait in just 61.6% of participants. These tumors show mild to moderate contrast enhancement following contrast. In a study of 21 patients with orbital schwannomas, Chen et colleagues discovered that schwannomas have lower CT values, ranging from 19.6 to 40 Hounsfield Units (HU), compared to other orbital tumors such as

orbital cavernous venous malformation and meningiomas, which have greater CT values.[9]

Magnetic resonance imaging (MRI) is crucial for surgical planning because it provides excellent soft tissue contrast.5 Orbital schwannomas can be well defined, with morphologies ranging from oval to multilobulated, cone-shaped, and dumbbell.[7] T1 weighted (T1W) scans usually show a nonspecific pattern of uniform isodensity or tiny patches of hypointensity.23,31 In some situations, enhanced T1W images show peripheral enhancement.. Relying on the histological characteristic of the schwannoma, post-gadolinium (Gd) enhanced T1W images reveal homogeneous or heterogeneous enhancement.[10] T2 weighted (T2W) images often show heterogeneous iso- to hyperintensity with patches of hypointensity inside the schwannoma.

The use of dynamic contrast-enhanced (DCE) MRI to distinguish between orbital lesions has also been investigated; however, a recent triple-blind randomized trial found that adding DCE-MRI does not improve diagnostic performance for describing orbital abnormalities.[10] Other imaging findings that can be seen in orbital schwannomas include cystic degeneration.[7,9] Intratumoral hemorrhage can also be detected in ancient schwannomas, distinguishing them from neurofibromas. Also, a recent retrospective study by Bi et al suggest that artificial intelligence can be utilized to distinguish between cavernous hemangiomas and schwannomas, helping in the diagnosis and therapy of these tumors.[1]

Though the majority of the ophthalmological literature recommends tumor excision as the primary treatment for orbital schwannomas, close observation has been described for schwannomas in various regions of the body, including several cranial nerve schwannomas.

Many studies advocate for early surgical resection due to the possibility of progressive symptoms; nevertheless, investigations tracking clinical outcomes of extra-orbital schwannomas by observation alone have indicated that many remain stable in size and symptoms. Elmalem et al discovered no significant variation between the initial and follow-up sizes of the extra-orbital trochlear schwannomas. A further investigation on clinical outcomes of surveillance of extra-orbital trochlear nerve schwannomas alone reported that one of six patients clinically worsened after 15 months. [5]

However, the remaining five individuals remained clinically stable. Furthermore, a short retrospective case series of three patients who developed a superior oblique palsy as a result of an extra-orbital trochlear nerve schwannoma discovered that none of the individuals had any additional symptoms, supporting cautious care. While further research is needed to define the natural history of conservatively handled orbital schwannomas, the slow growth rate of schwannomas elsewhere suggests that tiny schwannomas should be observed with serial MRI scans as their primary care.

There is currently no widely established methodology for managing orbital schwannomas; nonetheless, adopting a patient-centered approach is critical for collaborative decision-making. Management of extra-orbital trochlear schwannomas should be tailored to each patient depending on tumor size, growth rate, and symptoms, similar to Torun et al's recommended approach. Those with small tumors or minor symptoms should be given conservative treatment and advised about the possibility of surgical resection or GKS if they develop substantial symptoms or tumor growth. In cases of diagnostic doubt based solely on imaging, a biopsy and debulking of the mass may be preferred over total surgical excision to limit harm to orbital components. [10]

## Conclusion

Orbital schwannomas are rare tumors. Preoperative diagnosis is challenging because to the varying presentation and location. To avoid severe problems, orbital tumors should be assessed early using CT or MRI, and treated promptly. Orbital schwannomas should thus be considered in the differential diagnosis of slow-growing orbital masses.

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