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## Case Report

# An Unusual Differential Diagnosis of Orbital Cavernous Hemangioma: Ancient Schwannoma

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## Keywords

Schwannoma · Orbit · Ancient schwannoma · Neurilemmoma · Orbital tumors · Peripheral nerve sheath tumor

## Abstract

Schwannomas are rare lesions of the orbit that can be confused with cavernous hemangioma on imaging studies. We report the case of an 84-year-old woman with a 9-year history of a tumoral lesion in the inferolateral left orbit. The imaging studies did not reveal specific characteristics, only bone remodeling due to the long evolution of the tumor. The patient underwent complete excision of the tumor by anterior orbitotomy via the inferior conjunctival fornix. The histopathological examination revealed an ancient schwannoma, a variant of schwannoma with uncommon histological features. The follow-up was uneventful. The present case emphasizes the importance of considering neural tumors in the differential diagno-

sis of orbital masses with bone changes and degenerative alterations such as hemorrhagic areas, cysts, and/or calcifications.

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## Introduction

Schwannomas of the orbit are rare lesions comprising approximately 1% of orbital tumors [1]. The ancient schwannoma is a rare variant of schwannoma with a typical slow-growing benign course and particular histopathological features. We detected only 6 cases of these tumors in the literature [2–7] (Table 1). The literature search was performed on the MEDLINE database using the keywords “schwannoma,” “ancient schwannoma,” “neurilemmoma,” and “orbit.” Only articles in English were included.

We report here an unusual case of orbital ancient schwannoma mimicking an orbital cavernous hemangioma. Note its particular features, namely the presence of bone remodeling and nuclear and hemorrhagic degeneration.

## Results/Case Report

An 84-year-old woman presented with a 9-year history of progressive protrusion, deterioration of visual acuity, redness, watering, and lower eyelid malposition of her left eye. Upon inspection, the left globe was displaced superiorly and anteriorly and a mechanical lower eyelid ectropion was present. Palpation showed a firm, easily mobile mass in the inferolateral left orbit. The mass was nonpulsatile and did not vary with posture, ocular movements, or Valsalva maneuver. Examination revealed normal pupil reactions and diplopia in all eye positions. The best-corrected visual acuities were 9/10 in the right eye and 4/10 in the left eye. Exophthalmometry (Hertel exophthalmometer) was 14 mm in the right eye and 22 mm in the left eye. Biomicroscopy, intraocular pressure, and fundoscopy were normal. The right eye and systemic evaluation were unremarkable. Computed tomography (CT) scans revealed a large, well-demarcated, oval mass in the intraconal space of the left inferolateral orbit, which was isodense compared to the extraocular muscles (Fig. 1). There was discrete bone remodeling. Magnetic resonance imaging (MRI) showed an intraconal lesion in the left inferolateral orbit with well-defined margins and an oval shape (Fig. 1). The major axis measured 33 × 24 mm. On T1-weighted images, the lesion showed isointensity in relation to muscle and a heterogeneous but mostly hyperintense signal on T2-weighted images. T2 gradient-echo images showed a central hypointense component suggesting the presence of micro-hemorrhages. On contrast-enhanced images, the lesion showed a progressive and heterogeneous enhancement with a more vivid central region. A diagnosis of cavernous hemangioma was made based on the clinical and imaging features. The patient underwent an anterior orbitotomy via the inferior conjunctival fornix. The mass was purple-yellowish, with a smooth surface and well encapsulated, with no infiltration of surrounding soft tissues (Fig. 2). The tumor was completely excised without damaging the capsule. The histopathological diagnosis of the lesion was an ancient schwannoma (Fig. 2). The mass

measured 35 mm in its largest dimension. Microscopically, the tumor was composed of hypercellular (Antoni A, predominant) and hypocellular (Antoni B) areas. The lesion presented degenerative changes such as nuclear atypia (pleomorphic, hyperchromatic nuclei and nuclear pseudoinclusions) and hemorrhagic areas. Immunohistochemical study revealed strong positive staining for S-100 protein in tumor cells (Fig. 2). On follow-up at 3 months, the patient was asymptomatic, with no evidence of tumor recurrence. The mechanical lower eyelid inferior ectropion resolved spontaneously.

## Discussion

Orbital schwannomas are benign, often unilateral and originate from Schwann cells. They usually arise from sensory nerves, although some cases have been reported to derive from motor nerves. In the orbit, they usually arise from branches of the supraorbital or supratrochlear nerves and less commonly from the infraorbital, ciliary, oculomotor, trochlear, abducens, or optic nerves. In about 50% of cases, its origin remains obscure [2, 4, 6, 8–10]. The ancient schwannoma is an uncommon histological variant of schwannoma. The term “ancient schwannoma” was coined by Ackerman in 1951 to describe these tumors in the thorax [11].

Clinical features may include progressive proptosis, optic neuropathy, diplopia, numbness, and pain. There are no clinical or radiographic pathognomonic features differentiating schwannomas from other well-circumscribed orbital lesions such as cavernous hemangioma, fibrous histiocytoma, neurofibroma, hemangiopericytoma, solitary fibrous tumor, and others. A definitive diagnosis can only be made on histopathology [4–10]. Histopathological examination reveals that schwannomas are characterized by hypercellular (Antoni A) and hypocellular (Antoni B) areas. Areas of increased cellularity with nuclear atypia (pleomorphic, hyperchromatic, multilobulated nuclei) alternate with hypocellular areas with considerable fibrosis. Despite nuclear atypia, mitotic figures are absent. Immunohistochemical testing is usually positive for vimentin, neuron-specific enolase, and S-100 protein and negative for cytokeratin [2, 4, 7, 10]. Degenerative findings such as hemorrhage, cyst formation, focal calcification, and perivascular hyalinization are indicative of the long duration of the tumor and are characteristic of the so-called “ancient schwannomas” [2–7, 10]. The hypercellularity and nuclear atypia frequently present in ancient schwannomas may lead to the erroneous interpretation of a malignant tumor [2, 10]. Complete surgical excision was curative in all reported cases and malignant change was never seen in ancient orbital schwannomas [2–7, 9].

We believe that this case improves the knowledge about orbital tumors. We report here for the first time bone remodeling in an orbital ancient schwannoma with 9 years of evolution and with a detailed MRI description. The present case shows that tumors of the peripheral nervous system should be considered in the differential diagnosis of orbital masses with bone changes.

### Statement of Ethics

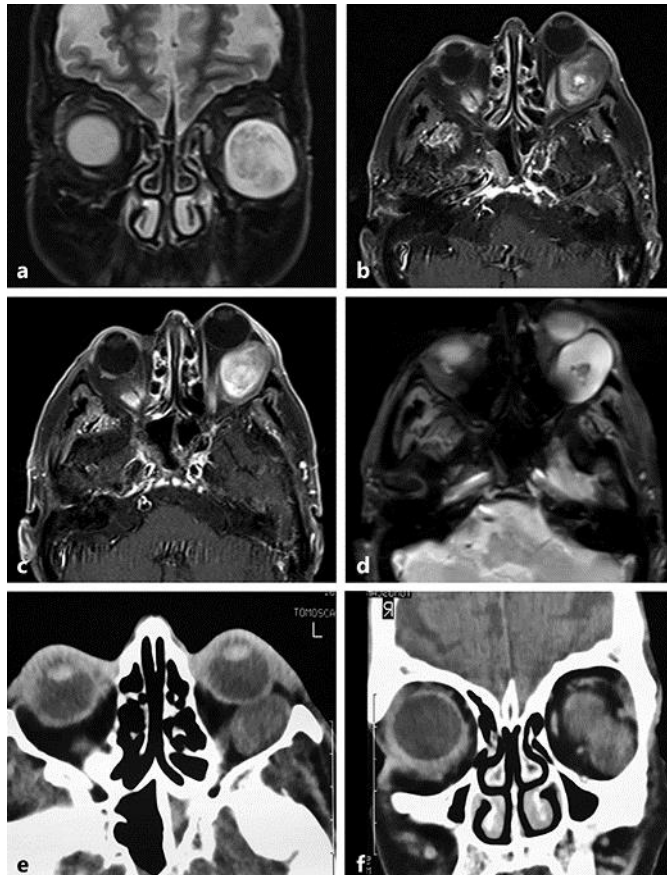
Written informed consent was obtained from the patient for publication of this case report and accompanying images.

### Disclosure Statement

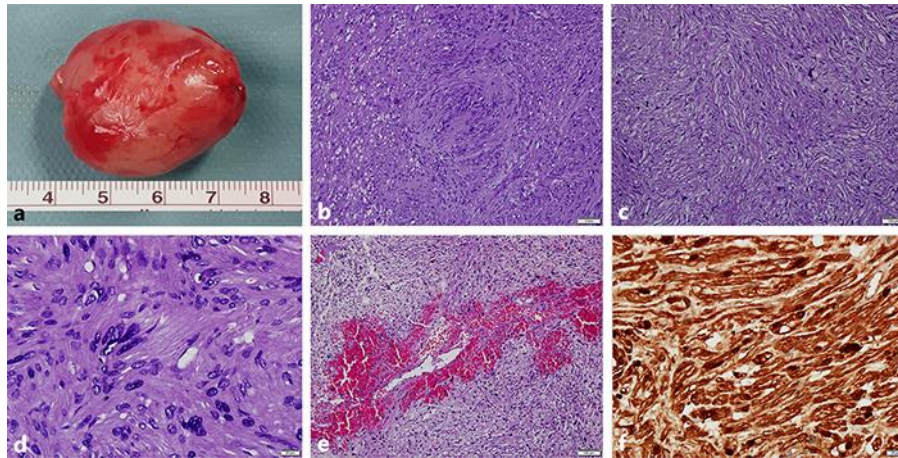
The authors have no financial disclosure.

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**Fig. 1.** MRI showed a well-defined, oval, intraconal lesion in the left inferolateral orbit. Axial T1-weighted image shows isointensity in relation to muscle. Coronal T2-weighted image shows a heterogeneous but mostly hyperintense signal (a). Axial post-contrast fat saturated T1-weighted images (b, c axial scans) show a progressive and heterogeneous enhancement with a more vivid central region. Axial T2 gradient-echo image (d) shows a central hypointense component suggesting the presence of micro-hemorrhages. CT scans (e axial; f coronal) revealed a large, well-demarcated, oval mass, isodense compared to the extraocular muscles in the intraconal space of the left inferolateral orbit; there was discrete bone remodeling.



**Fig. 2.** Orbital ancient schwannoma. **a** Macroscopic aspect: purple-yellowish mass, with a smooth surface and well encapsulated. Histopathological examination: Antoni A area, hypercellular with a Verocay body (**b** HE.  $\times 100$ ); Antoni B area, hypocellular (**c** HE.  $\times 100$ ); nuclear atypia (**d** HE.  $\times 400$ ), and hemorrhagic degeneration (**e** HE.  $\times 100$ ). **f** Immunohistochemical staining revealing positivity for S-100 protein. S-100.  $\times 400$ .

**Table 1.** Cases of orbital ancient schwannoma reported in the literature

First author [Ref.], year	Gender/ age, years	Clinical presentation	CT	MRI	Localization	Hystological findings	Complete excision	Recurrence/ follow-up
Khwarg [2], 1999	F/52	Epiphora Exophthalmos	Well-demarcated, oval, nonenhancing mass	NS	Intraconal Inferolateral	Nuclear pleomorphism Hyperchromatism Nuclear atypia Areas of hyper- and hypocellularity	Yes	NS
Moloney [3], 2004	F/65	Discomfort Protrusion	Large, round mass	NS	Extraconal Superolateral	Cystic Haemosiderin pigment deposition Nuclear atypia	Yes	No/6 months
Sales-Sanz [4], 2007	F/49	Progressive inferior orbital fat prolapse	Bilateral, heterogeneous, well-demarcated masses	NS	Extraconal Inferior	Antoni A and Antoni B areas Cystic IH – S-100 protein positive	Yes	No/16 months
de Jong [5], 2010	M/44	Upper eyelid swelling Anesthesia of supraorbital region	Tumor located above the rectus superior and levator palpebrae muscles	T1: hypointense with a peripheral enhancing rim T2: fluid-fluid level, representing hemorrhage within a central cystic lesion	Extraconal Superior	Hemorrhage Hemosiderotic or foamy macrophages Cystic degeneration IH – S-100 protein positive	Yes	NS
Pecorella [6], 2012	F/32	Inferior eyelid swelling Proptosis	1 cm localized homogeneous oval mass below the globe at the level of the inferior orbital rim	T1: moderate signal intensity in the lesion; enhancement of the signal intensity after gadolinium injection	Inferolateral	Cystic spaces Diffuse hypocellular areas Verocay bodies	Yes	Lost to follow-up
Kulkarni [7], 2014	F/68	Protrusion Pain Redness Watering	Mass in the left orbit	NS	Superior	Antoni A and Antoni B areas Verocay bodies Hemorrhage IH – vimentin, neuron-specific enolase, S-100 protein positive	Yes	No/1 months
Present case	F/84	Protrusion Inferior ectropion	Well-demarcated, oval mass; isodense compared to muscle	T1: isointense T2: heterogeneous, mostly hyperintense T2 GE: central hypointense component T1 C+: progressive, heterogeneous enhancement; central enhancing component	Inferolateral Intraconal	Antoni A and Antoni B areas Verocay bodies Hemorrhage Nuclear atypia IH – S-100 protein positive	Yes	No/3 months

CT, orbital computed tomography; MRI, magnetic resonance imaging; T1, T1-weighted images; T2, T2-weighted images; GE, gradient-echo images; C+, contrast-enhanced images; IH, immunohistochemical investigation; NS, not stated.