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Uncommon presentation of orbital schwannoma: A case report



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ABSTRACT

INTRODUCTION: Schwannomas are well-differentiated solitary benign tumors that originate from the schwann cells of the nerve sheath, constitute 1–8% of all head and neck tumors and 1–4% of the orbital tumors.

SUMMARY OF THE CLINICAL CASE: A 57-year-old female patient visited our department, because she has a blindness of the right eye associated with an irreducible exophthalmia classified grade III, 4 years ago. Radiological exploration showed a mass in the orbital cone in relation to a probable optic nerve schwannoma confirmed by biopsy. The affected eye was exenterated because of delayed diagnosis.

DISCUSSION AND CONCLUSION: In this review we discuss the pertinent clinical findings of this rare lesion and review the literature relative to optic nerve and solitary orbital schwannomas and insist that aggressive surgery with total mass removal should be warned by early diagnosis.

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1. Introduction

Intra orbital schwannomas and solitary neurofibromas account for 2–5% of all operated intra orbital tumors. Without any known neurofibromatosis, their diagnosis is only suggested with the MRI data. Their treatment is solely surgical.

Clinical presentation ranges from insidious proptosis, visual field loss, retro-orbital pain and headaches, to in rare cases blindness. Because the optic nerve is myelinated by central nervous system oligodendrocytes rather than Schwann cells, it is not surprising optic schwannomas are exceedingly rare lesions with, to our knowledge, only six previously reported cases [1]. The present case aims to highlight the diagnosis, investigation and management of schwannoma occurring in an unusual orbital location.

2. Clinical case

A 57-year-old female patient visited our ENT department because it has a blindness of the right eye associated with exophthalmia grade III for the past 4 years, where the patient has followed a traditional herbal treatment (Fig. 1).

She reported that she feels severe periorbital pain and discomfort, the lesion gradually increases in size and visual acuity has gradually decreases and she has no light perception since 1 year. On

examination, orbital movements were restricted associated with nonreducible exophthalmia. Visual acuity was 0/10 (right) and 5/10 (left). The rest of the clinical examination was unremarkable and there was no significant past medical history. There was no paresis or anesthesia of the face, no facial paralysis, no cervical lymphadenopathy.

A brain CT scan and Magnetic resonance imaging (MRI) revealed an intraconal process centered on the right optic nerve caused exophthalmia grade III (Fig. 2).

An anterior orbitotomy was performed after biopsy of the mass with radical excision of the tumor. The affected eye was exenterated because of delayed diagnosis and poor cooperation of the patient, the orbit was very infected and is no longer functional. An orbital epithesis is carried out.

Histopathological exam revealed a schwannoma with Antoni type A, following immunohistochemistry demonstrated strong reactivity to vimentin C and strong reactivity to S-100 protein (Fig. 3).

Two years later there was no evidence of local intra-orbital recurrence.

3. Discussion

Schwannomas account for 8% of all primary intracranial tumors. The vestibulo-cochlear nerve and the trigeminal nerves are the most common sites of origin [2,3].

It is well known that the optic nerves are myelinated by oligodendrocytes since their cell bodies arise centrally within the lateral geniculate nuclei. Because of this basic cellular anatomy, optic schwannomas should theoretically not exist [4]. It is possible, however, these rare lesions stem from small sympathetic fibers

Abbreviations: CT, computed tomography; MRI, magnetic resonance imaging.

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Fig. 1. Enormous proptosis of the right eye.

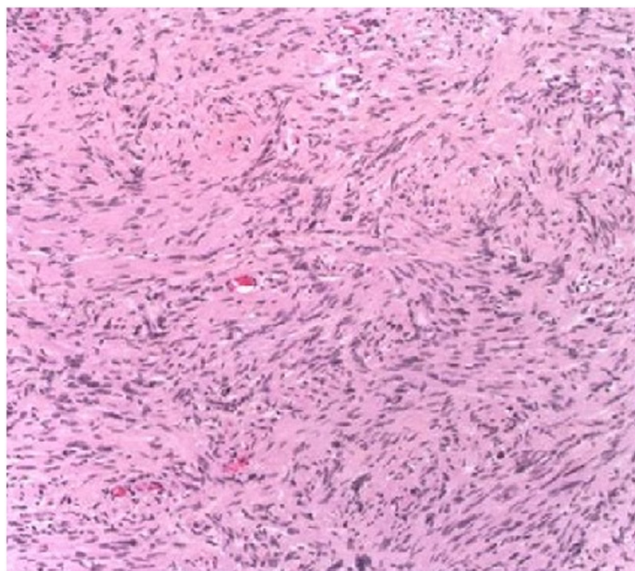


Fig. 3. Haematoxylin–eosin stain shows a highly cellular tumor with Antoni A areas (H&E*400).

that innervate the vasculature surrounding the optic nerve and its sheath [5].

The key-feature of an orbital tumor is exophthalmoses [6]. Pain is also a frequent feature of neoplastic disease in the eye. Sudden onset of pain is suggestive of intra-orbital hemorrhage [7]. Visual acuity disturbances vary. Diplopia is also a common complaint. Pupillary size alterations suggest invasion or compression of parasympathetic fibers through the ciliary ganglion [5]. Fundus changes such as optic nerve atrophy or papilloedema and choroidal folds are also among the usual findings [4].

On CT scan, schwannomas appear as smooth, ovoid, solitary, orbital retro bulbar masses, most commonly in the superior orbit with the long axis in the direction of the nerve, which is generally the anteroposterior direction [4,6]. The lesion can be seen in the extraconal or intraconal space. Rarely, it may present as an intra-muscular mass, however [8]. The tumor mass is usually isodense or slightly hyperdense when compared with the brain, and after injection of intravenous contrast medium, it often demonstrates homogeneous or heterogeneous moderate to marked contrast enhancement [9].

Intraorbital schwannomas commonly give low signal intensity in T1 and high signal intensity in T2-weighted images on MRI. They also demonstrate homogeneous contrast enhancement and generally appear as well-defined lesions with mixed components [10].

Even if the patient has not been the MRI, it is important to evoke because it is a key consideration in this type of pathology.

Histopathology examination should be performed to confirm the diagnosis, two patterns have been described, Antoni A and Antoni B [11].

Antoni type A areas consist of well-differentiated spindle cells with ovoid nuclei and fine chromatin stippling [11]. In Antoni type B areas, bipolar and multipolar cells are suspended in a loose myxoid matrix. On immunohistochemical stains, the tumors showed a negative reaction to cytokeratin and desmin. They were highly reactive to S-100 antibody and weakly reactive to vimentin, indicating that these lesions originated from Schwann cells of the peripheral nervous system [12].

Surgical excision is indicated as definitive therapy for orbital schwannoma [9,11]. The surgical approaches, whether anterior, lateral, or combined lateral orbitotomy and frontal craniotomy, depend on the location of the tumor in the orbit [9,6]. In our case orbitotomy allowed a total control of tumor, but the severe

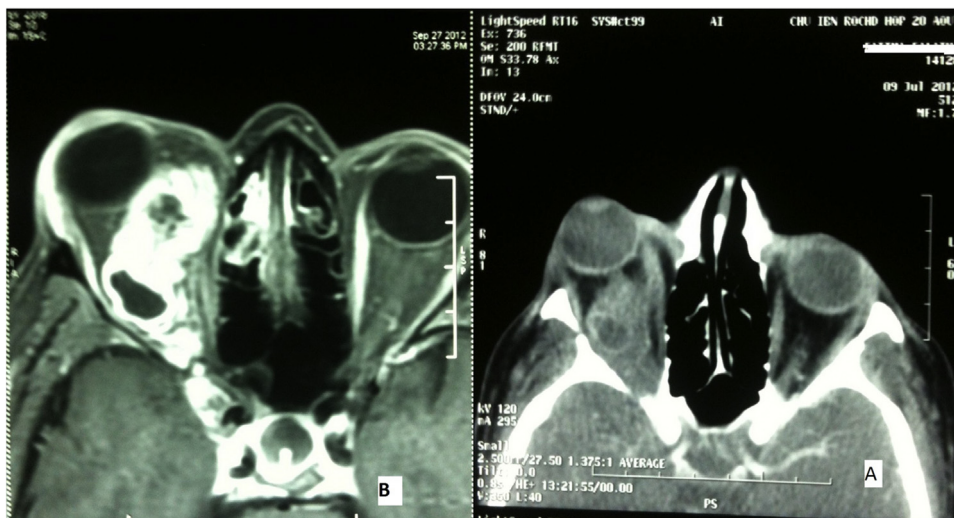


Fig. 2. (A) CT scan showing well-defined, hyperdense, heterogeneous tumor process in retro-orbit centered on the optic nerve. (B) Pre-operative T1-weighted MRI with contrast Axial view showing an enhancing lesion along the optic nerve at the retro-orbit.

infection of the orbit, we had no choice and we regret to do an exenteration and implement an orbital epithesis.

4. Conclusion

Aggressive surgery with total mass removal should be warned by early diagnosis. Although it may be difficult to differentiate these benign masses from other orbital tumours on radiologic imaging, the MRI characteristics can sometimes point to the diagnosis of a nerve sheath tumor. A definitive diagnosis can be made through correlation with histopathologic findings, however. In most cases, schwannomas have low malignant potential, and with total excision, recurrence is rare. An exophthalmia with decreased visual acuity should not delay diagnosis of this kind of tumor and quickly take care.

Conflicts of interest

All the authors have no personal or financial conflicts of interest regard this case report.

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Ethical approval

Not applicable for case reports.

Consent

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

Author contribution

Dr. Taoufiq Adouly was involved in the case writing and data collection for the case report. Dr. Chouaib Adnane was involved in critical review and making corrections to the manuscript.

Guarantor

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The work has been reported in accordance with the recommended criteria CARE.

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