ORBITAL SCHWANNOMA, A RARE ENTITY. CASE REPORT

Schwannoma orbitario, una entidad poco frecuente. Reporte de caso

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ABSTRACT

Introduction: Orbital schwannoma is a rare pathology, which constitutes approximately 1 to 6.5% of orbital tumors, and can originate from the ophthalmic branch of the 5th cranial nerve or from perioptic sympathetic nerves. Its diagnosis is made by magnetic resonance imaging (MRI) with contrast. The first-line treatment is surgery, and total resection provides a good prognosis. The time of illness is used to evaluate the visual prognosis in these patients.
Clinical Case: A 12-year-old woman, with a 9-year illness, characterized by a progressive decrease in right visual acuity until reaching amaurosis. Brain MRI with contrast shows an isointense tumor on T1, adhered to the medial aspect of the optic nerve sheath, which captures contrast, slightly hyperintense on T2. Total resection of the lesion is performed, and the diagnosis of schwannoma is confirmed by pathological anatomy. A month after surgery, the patient had slightly recovered her vision, without presenting other complications.
Conclusion: Orbital schwannoma is a rare pathology that must be treated surgically as soon as possible to achieve a better visual prognosis for the patient.

Keywords: *Neurilemmoma, Optic Nerve, Orbital Neoplasms, Cranial Nerves, (source: MeSH NLM)*

RESUMEN

Introducción: El schwannoma orbitario es una patología poco frecuente, que constituye aproximadamente el 1 al 6.5% de los tumores orbitarios pudiéndose originar de la rama oftálmica del V par o de nervios simpáticos periópticos. Su diagnóstico se realiza mediante resonancia magnética (RMN) con contraste. El tratamiento de primera línea es la cirugía, y la resección total otorga un buen pronóstico. El tiempo de enfermedad sirve para evaluar el pronóstico visual en estos pacientes.
Caso Clínico: Mujer de 12 años, con tiempo de enfermedad de 9 años, caracterizada por disminución progresiva de agudeza visual derecha hasta llegar a la amaurosis. La RMN cerebral con contraste muestra una tumoración isointensa en T1, adherida a cara medial de vaina del nervio óptico, que capta contraste, ligeramente hiperintensa en T2. Se realiza resección total de la lesión y el diagnóstico de schwannoma es confirmado mediante anatomía patológica. Al mes de cirugía, la paciente había recuperado ligeramente la visión, sin presentar otra complicación.
Conclusión: El schwannoma orbitario es una patología poco frecuente, que debe ser tratado quirúrgicamente lo antes posible para lograr un mejor pronóstico visual del paciente.

Palabras Clave: *Neurilemoma, Nervio Óptico, Neoplasias Orbitales, Nervios Craneales. (fuente: DeCS Bireme)*

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Schwannoma is a benign lesion that originates from Schwann cells and has been reported in 1-6.5% of orbital tumors, its main origins being the sensory nerves (from the ophthalmic division of the trigeminal nerve) and the sympathetic nerves (from perioptic vessels). [1, 2, 4, 6]

The Gold standard within the diagnosis is brain MRI with contrast, where it is identified as an isointense lesion on T1 and hyperintense on T2, with intense contrast enhancement.[2] Surgery by means of a craniotomy or an orbitotomy is the treatment of choice, being complete, curative excision without risk of recurrence. [1, 2, 4, 6]

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Theoretically, it is impossible for a schwannoma to arise from the optic nerve sheath because it does not have Schwann cells, although some exceptional cases have been described, [1,4] The case of a patient with an orbital Schwannoma treated successfully in the Hospital is presented. Almenara, Lima Peru.

CLINICAL CASE

History and examination: A 12-year-old female patient from Piura (northern Peru) has a medical history of toxoplasmosis without obvious sequelae. She has a 9-year history of a disease characterized by minimal visual alteration of the right eye, almost imperceptible, attributed to a refractive disorder. Right visual disorder worsens rapidly in the last 2 years until reaching a total loss of vision in the right eye. The patient begins medical evaluation in 2021 (due to pandemics). On examination: she is conscious, Glasgow Coma Scale (GCS): 15 points, without motor deficit, without sensory deficit, amaurosis of the right eye, without gait or balance alteration.

Brain MRI with contrast showed the presence of a tumor in the right anterior clinoid entering the optic canal, with regular edges, a round shape, isointense on T1, slightly hyperintense on T2, avidly capturing contrast and with the appearance of being adhered to the medial aspect of the right optic nerve, without restricting diffusion *(Fig. 1)*. A digital visual campimetry was performed which confirmed the amaurosis of the right eye.

Surgical treatment: A right pterional craniotomy is performed, with proximal opening of the Silvio valley and subfrontal arachnoid dissection until reaching the anterior clinoid process, the dura was opened, and a smooth white-pearly tumor is found. An anterior intradural clinoidectomy is performed, then a thick capsule is opened, followed by debulking and adequate hemostasis. The capsule is released, and the tumor is removed, the most difficult part being the tumor segment adhered to the medial part of the optic nerve sheath, where a very thin lamina remains that was coagulated with bipolar. *(Fig. 2)*

Clinical evolution: The patient has a good clinical evolution. In the immediate postoperative period, she was in GCS: 15, without motor or sensory deficits, photoreactive pupils, anopsia of the right eye (similar to the preoperative one). A brain tomography (CT) with contrast did not show a residual tumor or bleeding in the surgical bed *(Fig. 3)*. The pathological anatomy was reported as grade I Schwannoma, with positive S100 markers and positive CD56 in immunohistochemistry and negative EMA. These markers are compatible with neoplasms of the nerve sheath, ruling out the diagnosis of meningioma.

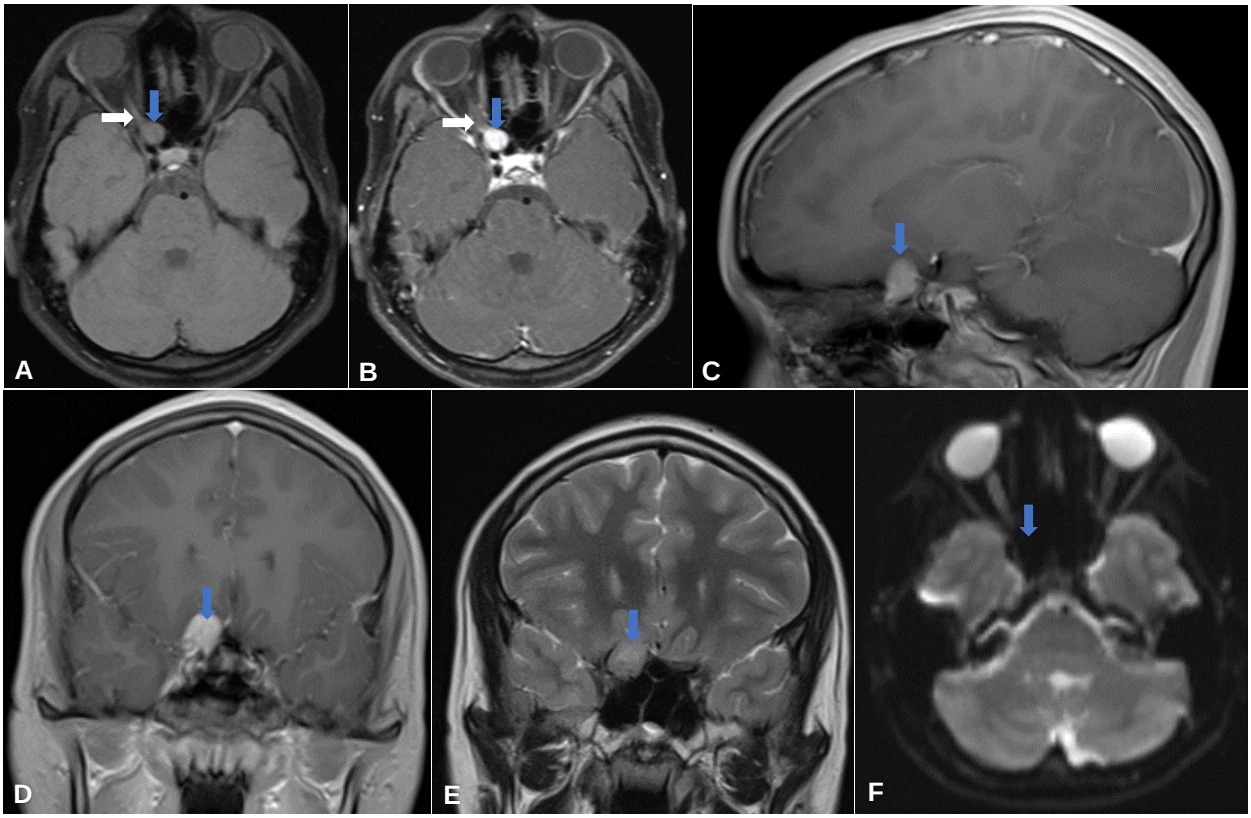

Fig 1. (A) Brain MRI without contrast, in axial section, in T1 sequence, showing an isointense expansive process *(blue arrow)* adhered to the medial aspect of the right optic nerve sheath *(white arrow)*. **(B, C, D)** Brain MRI with contrast, in axial, sagittal, and coronal section respectively, in T1 sequence, where an expansive process *(blue arrow)* dependent on the medial aspect of the optic nerve sheath *(white arrow)* is evidenced. that avidly captures contrast. **(E)** Brain MRI, T2 sequence, in coronal section, showing a slightly hyperintense expansive process *(blue arrow)*. **(F)** MRI brain, diffusion sequence, in axial section, showing an expansive process that does not restrict *(blue arrow)*.

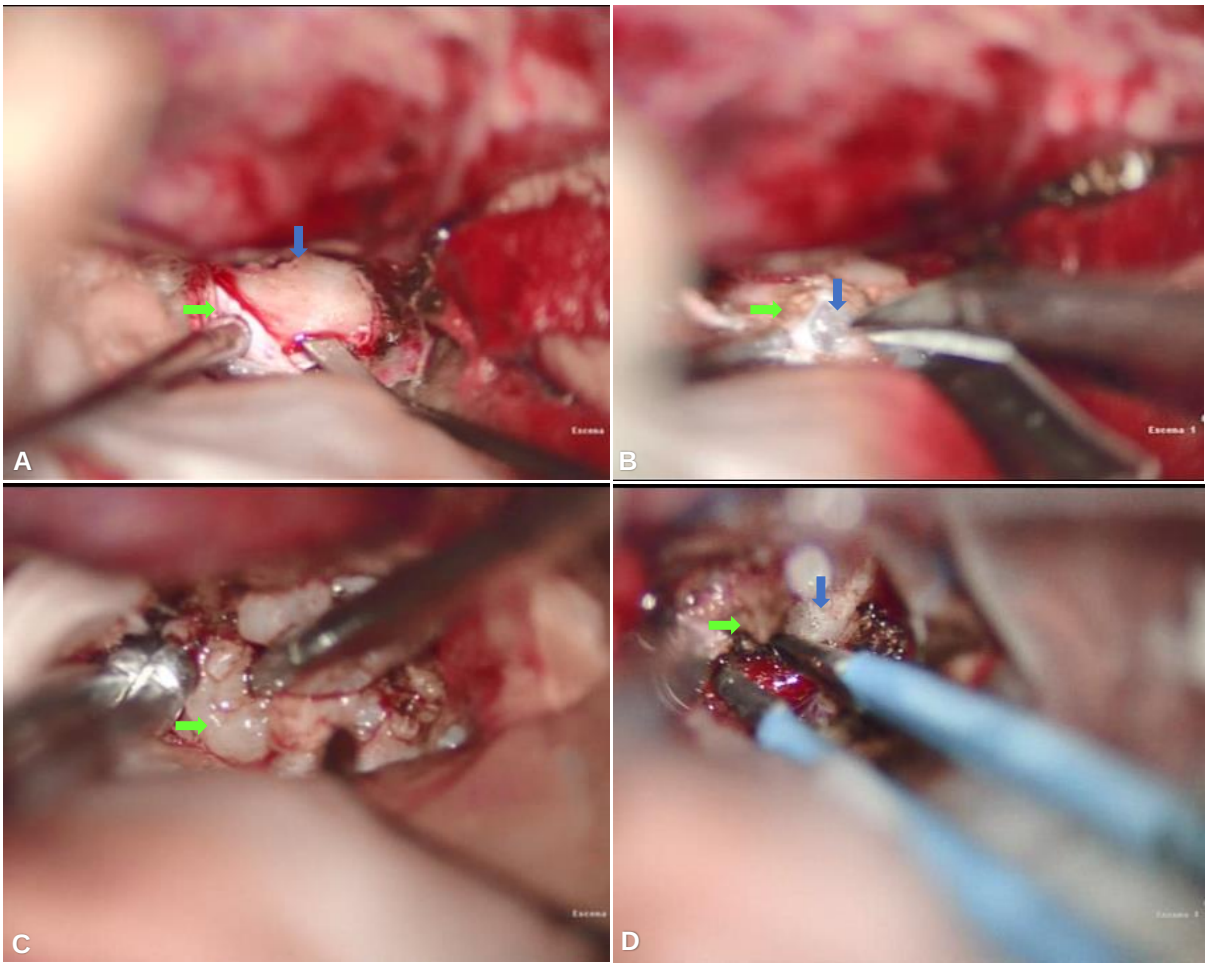

Fig 2. Intraoperative images under a surgical microscope. **(A)** The opening of the dura mater which is folded medially *(green arrow)* and the anterior clinoid process *(blue arrow)* are shown. **(B)** Opening of the tumor capsule *(green arrow)* with a No. 11 scalpels and the softer intracapsular tumor can be seen *(blue arrow).* **(C)** Intracapsular debulking of the white-nacreous tumor *(green arrow),* with little bleeding. **(D)** The capsule folding towards the lateral and anterior part *(green arrow)* is appreciated, which is coagulated with bipolar. The medial bony part of the optic canal is visualized *(blue arrow).*

One month after surgery, the patient slightly improved vision in the right eye (vision of light and shadows, without counting fingers). A brain MRI with contrast showed total resection of the lesion. *(Fig. 3)*

DISCUSSION

It is known that myelin is formed by oligodendrocytes in the central nervous system and by Schwann cells in the peripheral nervous system,[2] being schwannomas benign tumors of the latter, [1, 2, 3] which have been reported between 8- 10% of all intracranial tumors.[4,6] The most common location of a schwannoma is the vestibular region (VIII cranial nerve), followed by trigeminal schwannoma (V cranial nerve). They are associated with type 2 neurofibromatosis. [1,4]

The review of the medical literature does not provide evidence that there is a schwannoma of the optic nerve sheath itself, which is why they should be considered as orbital schwannomas until evidence of a histopathological communication between the optic nerve sheath and the optic nerve is found. schwannoma, since in all the cases described, the only thing that is evidenced is an adherence to the optic nerve sheath. [2]

Furthermore, it is theoretically impossible for a schwannoma to originate from the optic nerve sheath since it does not have Schwann cells, although some exceptional cases (less than 15 cases) have been described in the literature. [1, 4]

For this reason, an explanation has been attempted with the following theories: Orbital Schwannomas can arise from the intraparenchymal ectopic location of Schwann cells; by terminal differentiation of the pial cells in the optic nerve sheath (known as "schwannosis") and/or of the sympathetic nerves that arise from the perioptic vasculature. [1, 2, 4]

These tumors have been reported in 1-6.5% of orbital tumors. The sensory nerves (from the ophthalmic division of the trigeminal nerve) and the sympathetic nerves (from the perioptic vasculature) are the 2 main origins of orbital schwannomas. [1, 2, 4, 6]

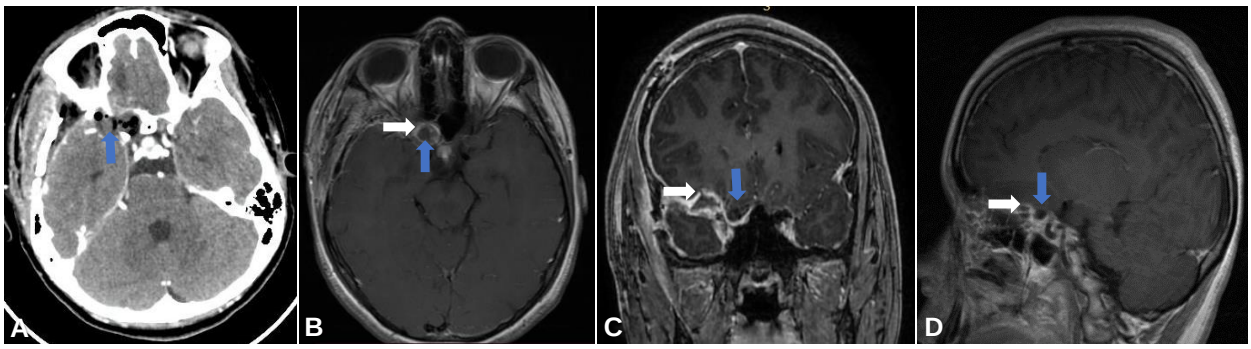

Fig 3. (A) Brain tomography with contrast, in axial view, where the absence of the lesion is evidenced *(blue arrow)* and there is no evidence of acute postoperative complications. **(B, C, and D)** Brain MRI with contrast a month after surgery contrasted T1 sequence, where the absence of the lesion *(blue arrow)* is evidenced with meningeal inflammatory reaction due to the surgical process *(white arrow)* at the temporal and medial level of the canal. optical. **B**. Axial view. **C**. Coronal view. **D**. Sagittal view.

Schwannomas partially located in the optic canal are rare, but those confined entirely within the optic canal are much rarer. Intraorbital schwannomas account for 1–2% of all orbital tumors. [5]

Histopathologically, schwannomas have slow growth, encapsulation, and mass effect on adjacent structures.[4] There is a risk of malignant transformation, especially in children, with pain being a sign of this. [1]

The symptoms of these patients are usually loss of visual acuity, retroocular pain, and proptosis. [4]

The visual prognosis is generally poor, but early surgery provides a better visual prognosis. The Gold standard within the diagnosis is the brain MRI with contrast, where they are characterized by being isointense lesions on T1, with intense contrast uptake, and hyperintense on T2. [2]

Surgery, by craniotomy or orbitotomy, is the primary treatment, with complete excision being curative and without risk of recurrence.[1,2,4,6] Radiation therapy has been described with potential treatment, but there are no studies to support its indication. [2, 4]

A modification of the classic pterional craniotomy, towards a slightly subfrontal one, is preferred, an approach that is simpler and faster.[4] The use of bipolar in hemostasis should be restricted to a minimum to avoid thermal injury to the second cranial nerve. Likewise, damage to the perineural vessels of the optic nerve sheath and nerve fibers should be avoided through careful dissection of tumor adhesions and neurovascular structures, as this achieves a better patient prognosis. [6]

Differential diagnoses include optic nerve glioma and optic nerve sheath meningioma since these tumors are much more frequent.[5] Our case had imaging characteristics to be considered an orbital schwannoma, but due to its low frequency it was not the first diagnostic possibility; its confirmation was only made with the pathological anatomy. As mentioned in the medical literature, surgery is the first option, a treatment that we carry out in our case, achieving the cure of the patient.

On the other hand, as mentioned in the reports, the time of illness is a primary factor in the prognosis of the patient.[2] In our case, even though the patient had a prolonged illness (9 years of illness) with severe vision impairment, after 1 month of surgery a slight recovery of vision was observed, with the possibility of further improvement in the following months.

CONCLUSION

Orbital schwannomas are a rare pathology, the diagnosis of which should be suspected based on its clinical-imaging characteristics and should be treated surgically as soon as possible to improve the visual prognosis of the patient. Morbidity and mortality rates are low if total resection is achieved.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Authors Contributions

Conception and design: All authors. *Drafting the article:* Vargas. Critically revising the article: Palacios. *Reviewed submitted version of manuscript:* Vargas. *Approved the final version of the manuscript on behalf of all authors:* Vargas.

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