Intra orbital Schwanoma of the supraorbital nerve

Schwanoma do Nervo Supra-Orbitario

RESUMO

Introdução: A órbita pode ser o local de diversas lesões neoplásicas ou não neoplásicas, sendo muito raros os schwannomas do nervo supraorbitário.

Relato de caso: Os autores apresentam o caso de uma paciente feminina de 15 anos de idade com história de proptose unilateral progressiva, sem outros sintomas visuais. Com o diagnóstico de tumor intraorbitário, a paciente foi submetida a cirurgia com retirada total da lesão.

Discussão: Schwannomas intraorbitários são usualmente associados à neurofibromatose, podendo originar-se em diferentes estruturas (incluindo nervos periféricos ou cranianos). Os sinais e sintomas normalmente são crônicos. O tratamento cirúrgico é necessário, com diversas abordagens cirúrgicas descritas.

Conclusão: O décimo-primeiro caso de um schwannoma do nervo supraorbitário é descrito, ressaltando-se suas principais características.

Palavras-chave: tumor orbitário, schwannoma, nervo supraorbitário

ABSTRACT

Background: The orbital region can be occupied by various lesions including both neoplastic or non-neoplastic. Schwannomas of the supraorbital nerve are very rare tumors.

Case Report: A case of a 15 years-old female patient with a one-year history of progressive proptosis and no visual symptoms is presented with. This patient underwent a combined surgery with total removal of the tumor.

Discussion: Intraorbital schwannomas are usually associated with neurofibromatosis and different structures can originate these tumours, including peripheral and cranial nerves. The signs and symptoms usually follow a chronic process. Surgical treatment is necessary, with various reported surgical approaches.

Conclusion: We report, to the best of our knowledge, the eleventh case of schwannoma of the supraorbital nerve, describing the main features of this pathology.

Key words: Intraorbital tumours, Schwanoma, supraorbital nerve
INTRODUCTION

The orbital region can be occupied by various pathologies, including tumors, congenital, infectious and vascular lesions, all arising from the intracranial, facial, nasal, paranasal and skull base tissues.\(^2\)

Schwannomas arising from the intraorbital nerves are uncommon. These tumors can be single or multiple\(^2,10\) intra or periorbital, either benign or malignant.

Lesions of the orbital space present clinically with a chronic history of visual signs and symptoms, and a intraorbital mass with slow expanding rate.\(^2\)

In the literature there are only 10 reports of schwannomas arising from the supraorbital nerve (Table 1).\(^2,3,5,8,9,11\) In the present article, we report another similar case, in a 15 years-old patient.

Table 1. Reported schwannomas of the supraorbital nerves in the literature.

<table>
<thead>
<tr>
<th>Author</th>
<th>Cases</th>
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<tbody>
<tr>
<td>Schatz(^9)</td>
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<tr>
<td>Horie(^3)</td>
<td>2</td>
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<td>Shul'ga(^11)</td>
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<td>Rose(^8)</td>
<td>4</td>
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<td>Goel(^3)</td>
<td>1</td>
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<td>Barbagallo(^2)</td>
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<td>Present study</td>
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CASE REPORT

A 15 years-old patient without previous pathological history, complained of a one year history of progressive ocular protrusion. She had no complaints about visual deficit. The ophthalmological exam showed VA 20/40 in the right eye and 20/20 in the left eye. Confrontation visual fields were normal. Paralysis of the superior rectus muscle was observed with a non-painful proptosis of the right eye. Consensual and direct fotomotor reflexes were normal. Intraocular pressure was 15mmHg in the right and 10 in the left. Ocular fundoscopy was normal in both eyes.

CT of the orbits showed a intraorbital mass in the right eye extending to the orbital roof causing bony remodeling with enlargement of the superior orbital fissure [Fig. 1]. The tumor measured 4.7 x 2.6 x 2.1 in the maximum dimensions.

MRI disclosed a big extraconal lesion in the right orbit, superiorly located in the superior rectus muscle causing proptosis: the tumor caused inferior displacement of the superior rectus muscle, the lateral rectus muscle and the optic nerve; and the superior oblique muscle was displaced medially. This mass had a low signal on T1 images and a high signal on T2 with important contrast enhancement [Fig. 2].
The patient underwent a combined neurosurgical and ophthalmological surgery: the transcranial approach modified by Hayek was chosen. After a right frontal craniotomy, the frontal lobe was displaced and the orbital roof was opened, giving access to the tumor which was located above the superior rectus muscle. A complete removal of an encapsulated tumour, originating in the superior orbital nerve was obtained. An histological diagnosis of schwannoma was obtained [Fig. 3]. Postoperatively, the patient had a good recovery: three months later, in a follow-up consultation, no symptoms of visual deficit and a mild enophthalmy of the right eye were found.

**Figure 3.** Pathology report: neoplasia with fusiform cells justaposed in crossing bundles. Palisades formed by side-by-side nuclei. Poorly nucleated areas of a palisade (Verocay bodies) are present. (HE, 100).

**DISCUSSION**

Different lesions can compromise the orbital space, both neoplastic or non-neoplastic. These include mainly hemangiomas, hemangiopericitomas, histiocytomas and lacrimal gland tumors. Intraorbital schwannomas and neurofibromas are responsible for 1.7-6.8% of orbital tumors and they are often associated with neurofibromatosis. Various structures can originate a schwannoma of the intraorbital region, including different nerves: the III, IV and VI cranial nerves, and the branches of the ophthalamic nerve.

Various signs and symptoms have been reported: proptosis, pain, bone erosion and displacement of the eyeball. Papilloedema and optic atrophy were also described. These findings usually follow a chronic and slowly progressive process.

Surgical treatment with removal of the lesion usually results in a good outcome. Different surgical approaches have been reported: the size, location and involvement of orbital structures need be considered for the choice of the surgical strategy. There are in the literature 10 reported cases of schwannomas from the supraorbital nerve. With the exception of 2 cases, all other reported schwannomas have association with neurofibromatosis. In the present case, despite the fact that no genetic evaluation was done, there was no clinical evidence of cutaneous neurofibromatosis, as the clinical and neurological examination showed no evidence of the disease.

**CONCLUSION**

We report the eleventh case of a supraorbital nerve schwannoma, most of them involved with neurofibromatosis. To the best of our knowledge, this is the third case with no evidence of neurofibromatosis.

**REFERENCES**


**CORRESPONDING AUTHOR**

Apio Antunes*
Neurosurgical Unit, Hospital de Clínicas de Porto Alegre
Rua Ramiro Barcelos, 2350, segundo andar
Largo Eduardo Zaccaro Faraco
CEP 90.035-903 Porto Alegre, Brasil
Phone/Fax : +55 (51) 2101 - 8182