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REPORT OF A RARE CASE OF INTRACONAL ORBIT SCHWANNOMA

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Abstract: We described a case of orbital schwannoma in a patient undergoing oncological follow-up for 5 years for infiltrating ductal carcinoma of the breast. Patient treated with complete surgical resection of the tumor without the need for any adjuvant treatment.
Keywords: Cells dand Schwann,Orbital tumor, schwannoma.

INTRODUCTION

Schwannoma is a tumor of Schwann cells, found in the myelin sheath, which lines peripheral nerves. In general, it presents rapid growth, and may affect mainly the vestibular nerve, followed by the trigeminal nerve and terminal branches of the oculomotor nerves. This condition constitutes only 10% of intracranial tumors and is also rare in orbit, representing about 1% of all orbital tumors. Therefore, the objective of this study is to report a case of a patient diagnosed with malignant peripheral nerve tumor (schwannoma) in the orbit, being the second primary tumor, in a patient with a history of treated breast cancer. In addition, review pertinent information about this rare neural injury.

METHODOLOGY

This is a primary, descriptive study involving a review of the medical record of a patient diagnosed with Schwannoma, who underwent surgical treatment by the general surgery team of the General Hospital of Cuiabá-MT. A literature review and photographic record of the surgical procedure were performed. Data collection and follow-up of the case were performed by the authors of the research, being selected data such as age, previous pathological history, clinical presentation, results of imaging exams, anatomopathological reports and evolution of the case. The case report was carried out with the consent of the family member responsible for the patient.

CASE DESCRIPTION

Patient MHR 66 years old, born in Barra do Garças-MT, with cognitive and motor deficits secondary to poliomyelitis in childhoodwith aphasia of comprehension and expression, dysarthria, not properly contacting the environment, impaired verbal communication with dysfunctional speech emitting only incomprehensible sounds, in the musculoskeletal system there is spastic tetraparesis with difficulty in locomotion and reduced movements. The responsible/caregiver and main source of information is the sister. In May 2017 she was diagnosed with infiltrating breast ductal carcinoma, HER2 3+/3+, ER positive 10%, PR negative, FISH 3+, Ki67 20%. In time, she underwent surgical treatment with modified radical mastectomy on the right with axillary lymphadenectomy associated with adjuvant therapy.

She underwent adjuvant treatment with herceptin and anastrozole, remained under oncological follow-up for 5 years, with no evidence of active neoplastic disease. In May 2022, 5 years after the diagnosis of breast cancer, she was brought to the medical consultation by her sister, reporting discomfort in the left eye that started about 3 months ago, associated with intense and intermittent headache.

Physical examination of the orbit revealed significant hyperemia and proptosis of the left eye, with banded corneal ulcer and uveitis. At this first moment duethe pain, the patient was uncooperative, making it impossible to perform the eye fundus examination.

Figure 1

Cranial MRI shows a bilateral microphthalmia, brain volumetric reduction incompatible for age, expansive nodular lesion with a fusiform configuration, intraconal in the left orbit, heterogeneous with a predominance of hyposignal on T1 and hypersignal on T2, measuring approximately 2.8 x 2 .2 x 2.3 cm determining ocular proptosis, findings that

favor the diagnosis of Schwannoma-type neural sheath tumor.

Given the hypothesis of orbital Schwannoma by radiology, surgical treatment was proposed for the case. Enucleation of the left orbit was then performed, which was uneventful. On physical examination on the first postoperative day, the patient was in good general condition, with stable clinical parameters. He had a good clinical evolution and was discharged from the hospital on the second postoperative day for continuity of outpatient follow-up. She is in 2 months of follow-up with no new evidence of oncological disease or clinical complications related to the surgery.

Figure 2 / Figure 3

The anatomopathological study reports: left eyeball associated with a solid tumor, weighing together approximately 28 g and measuring 6.0 x 3.0 x 2.5 cm. The eyeball measures approximately 2.0 x 1.9 x 1.5 cm; the cuts show petrified content in the eyeball. On the posterior surface of the eyeball, a solid mass measuring 5.0 x 3.3 x 1.4 cm characterized by encapsulated proliferation of fusiform cells, with wavy nuclei and undefined cytoplasm, with more cellular areas (Antoni A type) and less cellular areas (Antoni B type) with a hyalinized appearance, described as Schwannoma.

Figure 4

DISCUSSION

Schwannoma is within a group of tumors generically called peripheral nerve tumors, electron microscopy was able to determine that Schwann cells are the main proliferative unit of the tumor, thus determining its name.Its incidence corresponds to about 1% of orbital tumors with a similar distribution between men and women. These are radioresistant tumors and surgical excision is the oncological basis of treatment.

Clinically it manifests with hyperemia, pain, proptosis and changes in eye movement. In the case reported, due to the patient having cognitive deficit, important motor sequel and being difficult to communicate, due to ocular proptosis and entropion, the condition evolved with significant uveitis and corneal ulceration, and there was a high probability of functional deficit in the affected eye.

Benign Schwannoma in general is a tumor with well-defined contours and eccentric to the nerve of origin, and may present different distribution patterns of Schwann cells in addition to lymphocytes, macrophages and calcifications. The histological diagnosis of malignancy is based on the presence of cells irregular in size and shape, atypical mitoses, increased nucleus size, hyperchromatism and many tumor cells.

For tumors restricted to the orbit, surgery is the treatment of choice, sometimes there is fusion of the tumor capsule with the capsular barrier of the optic nerve, making the surgery more delicate.

The optic nerve must be preserved whenever possible, in this case, due to the patient's evolution and due to the functional deficit of the eyeball, enucleation of the left orbit was chosen.

CONCLUSION

The diagnosis of intraorbital Schwannoma presents difficulties, due to its rarity, but the hypothesis must always be considered in patients who present unilateral proptosis of rapid onset. It is necessary to carry out complementary imaging methods to formulate the diagnostic hypothesis, in the case described, magnetic resonance imaging was used to define the probable diagnosis and subsequent pre-surgical planning. The patient was treated with complete surgical resection of the tumor, defined by the literature as the treatment of choice, not requiring any adjuvant treatment.

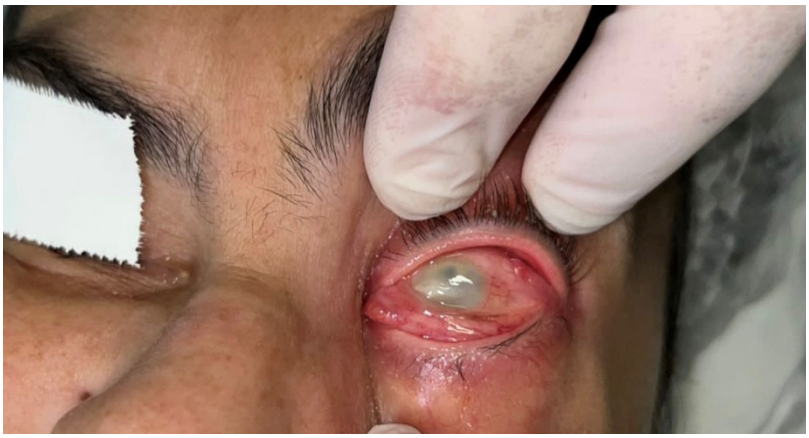

Figure 1: Physical examination showing significant uveitis with proptosis of the left eye.

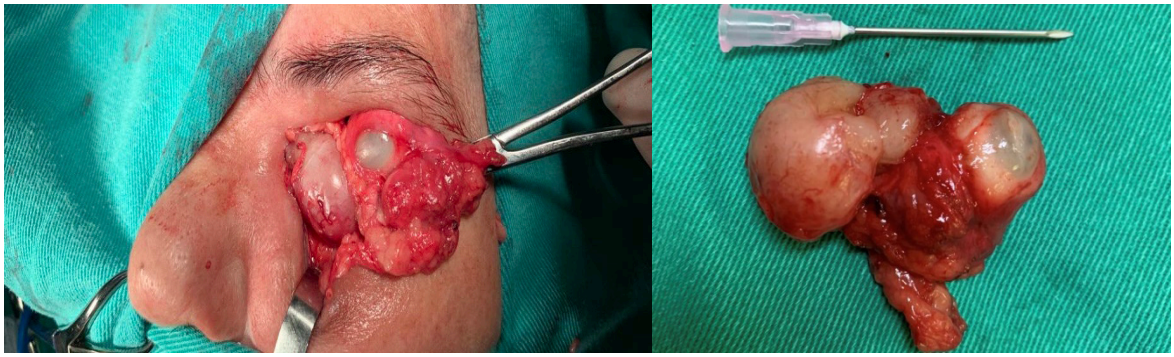

Figure 2: Exposure of the tumor posterior to the eyeball.

Figure 3: Surgical specimen, left eyeball together with the Schwannoma.

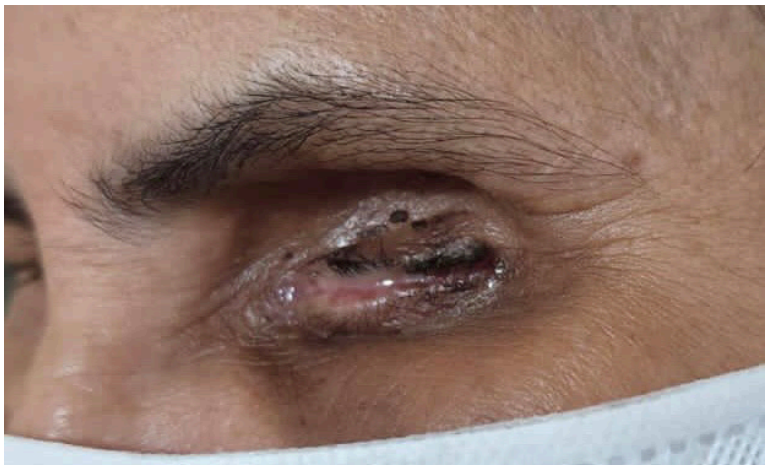

Figure 4: First outpatient return on the seventh postoperative day.

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