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Unusual and aggressive presentation of intraorbital melanoma

Apresentação incomum e agressiva de melanoma intraorbitário

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ABSTRACT

The purpose of this study is to report a intraorbital melanoma case with atypical and aggressive presentation, forming a large painful mass with erythematosus and inflammatory aspect protruding from the left orbit with eyeball damaged at its peak. Piece analysis identified malignant melanoma compound of epithelioid, spindle and anaplastic cells.

Keywords: *Melanoma; Eye neoplasms; Uveal neoplasms; Orbital diseases; Orbital neoplasms; Case reports*

RESUMO

O objetivo dos autores é relatar um caso de melanoma intraorbitário de apresentação atípica e agressiva, formando grande massa dolorosa de aspecto eritematoso e inflamatório projetando-se da órbita esquerda com o globo ocular danificado em seu ápice. A análise da peça identificou melanoma maligno com componentes celulares epitelióide, fusocelular e anaplásico.

Descritores: Melanoma; Neoplasias oculares; Neoplasias uveais; Doenças orbitárias; Neoplasias orbitárias; Relatos de casos

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Introduction

Uveal melanoma is the most common primary intraocular tumor in adults.1 Depending on the location and size, it can be asymptomatic or cause a wide variety of symptoms, such as worsening of visual acuity, floaters, loss of part of the visual field, or eye pain. Histologi-cally, melanoma is presented in different cell types, with epithelioid and anaplastic being the worst prognosis.2 There were advances in diagnostic methods, but mortality as a result of the tumor has not changed much in recent years.3

Our purpose was to make a brief review of current issues related to the subject and report an intraorbital melanoma case of atypical presentation with rapid growth, poor prognosis and recurrence after surgery.

Case Report

Patient JVS, 57 years old, male, came for urgent care at the emergency of the ophthal-mology service at Hospital Governador Celso Ramos - Florianópolis on March 2015 complaining of severe pain and mass in the left orbit. During the first appointment, he men-tioned trauma in left eye with foreign body 6 months before, and mass growth followed by pain since then. The examination showed erythematous, fixed mass, protruding approxi-mately 3 cm from the left orbit with exposed cornea and damage, upper eyelid free, edem-atous and unable occlude the region exposed. (Figure 1) There was no pain on palpation, any ocular motility, no light perception and lack of secretion. Former morbid story without any particularity. He denied use of eyedrops or previous surgery in the eye. He reported having normal vision until 6 months prior to the appointment. The right eye was normal to the exam, with visual acuity 20/20 and without any other particularity.

CT scan of the skull was performed showing large ovoid intraorbital mass with erosion of cortical bone and invasion of extraocular muscles.(Figure 2) The material collected for bi-opsy showed inconclusive results. The immunohistochemical analysis conducted then showed positive AE1/AE3 cytokeratin in epithelial cells, negative epithelial membrane an-tigen, positive S100 protein in the stromal cells, and positive CD31 in the endothelium. Af-ter the results, the patient returned to service with increased mass and persistence of pain, nintratumoral necrose, vascular and nervous invasion with staging pT4a. (Figure 3) The patient was then referred to the oncology service.

Approximately thirty days after, he returned with large mass relapse, now more darkened, crumbly without adhesion to the upper eyelid margin. At this point, we decided not to inter-vene surgically, analgesia remained, and he was referred to the oncology service. (Figure 4)

Discussion

The uveal melanoma, an entity that may affect the iris, ciliary body and choroid due to the presence of melanocytes in their tissues, is the most common primary intraocular tumor in adults.[1] Melanomas of the ciliary body can be initially asymptomatic and

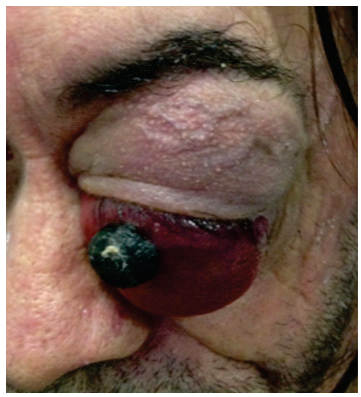

Figure 1: Patient with large, fixed erythematous mass, projecting from the left orbital cavity

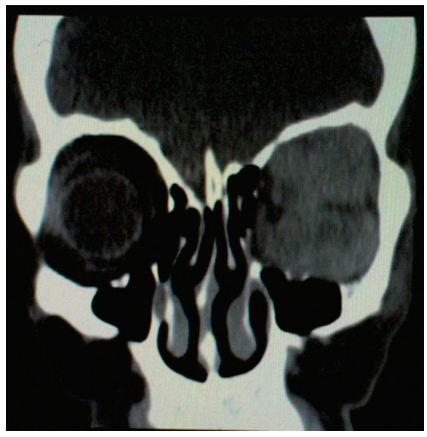

Figure 2: Computed tomography of orbit, revealing ovoid infraorbital mass with erosion of cortical bone and invasion of extraocular muscles.

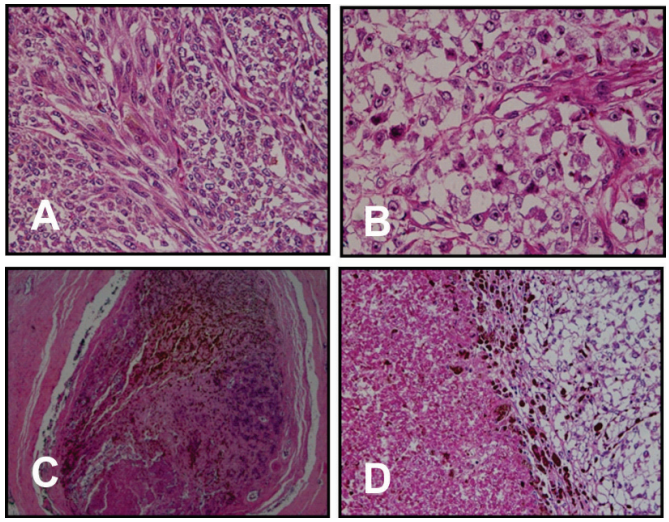

Figure3: Histopathology slide.
(A) Neoplastic melanocytes with cell pattern in spindle fascicles and frequent aberrant mitoses (HE 400x).
(B) Neoplasic melanocytes with epithelioid cell pattern (HE 400x).
(C) Infiltration of the optic nerve by atypical melanocytes (HE 40x).
(D) Pigmented neoplastic cells with aberrant mitosis and extensive areas of necrosis (HE 100x).

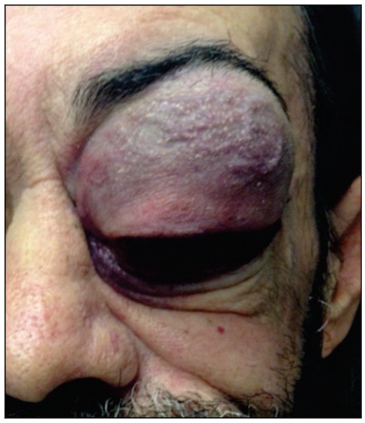

Figure 4: Patient photograph showing the appearance of the tumor relapse in the left orbital caviy.

difficult to see due to its location behind the iris, and the first symptoms may be nonspecific, such as poor visual acuity, floaters, loss of visual field or ocular pain due to secondary glaucoma.[2] The choroidal melanomas typically assume the dome conformation, pigmented and elevated below the retina, with color ranging from amelanotic to dark brown, and bringing more symptoms related to visual acuity.[4] Our case shows a patient in advanced stage of the disease with the content and orbital continent compromised.

Epidemiologically, the patient lies within the age group with the highest incidence of the disease, and a recent study shows that there may be some relation between the patient's age and the type of melanoma cell, with fusiform being the most prevalent in patients with average of 60 years, epitelioide with 65 years old, and mixed with 64 years old, but such correlation did not correspond to the case.[5]

Despite the better accuracy in diagnosis, mortality as a result of these tumors has not changed significantly in recent years.[3] The incidence of metastasis is high, and survival in metastatic cases is on average of 12 months, with the liver being the most affected organ.[1]

Enucleation is the appropriate treatment for medium (T2), large (T3) and very large (T4) tumors, and in cases of pain. Small and medium-sized tumors, without documented growth and with less than 1 mm thick can be treated more conservatively with brachytherapy, which offers good rates to control the disease, but little is known about the survival of these patients who were treated conservatively, although a study confirms the reduction of mortality in the following years.[4-6] There is a consensus that the conventional external radiotherapy is not effective as a single mode of treatment of melanoma, and cases where the enucleation was required even after the radiation therapy are associated to the worst prognosis.[4-7] The exenteration, traditionally indicated for posterior uveal melanomas with extra-escleral expansion, is today little indicated due to enucleation and radiation therapy show similar survival outcomes.[4-8] We chose exenteration in this case due to the pain symptoms and the large volume and invasion of the tumor mass. A study that evaluated patients who suffered enucleation showed that the recurrence can occur in 3% of cases of melanoma restricted to the intraocular compartment, and in 18% of cases in which there is evidence of extra-escleal extension.[9] The degree of the lesion and rapid recurrence give this case a reserved prognosis.

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