Short communication

Undifferentiated high-grade pleomorphic sarcoma of the orbit: A case report

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ABSTRACT

Case report: A 27-year-old female presented to us with a complaint of proptosis of her right eye. Imaging tests showed an extracanal mass in the medial orbital wall, which affected the medial rectus. The tumor was removed and adjuvant radiotherapy was required. The histological examination showed an undifferentiated high-grade pleomorphic sarcoma of the orbit. There has not been recurrence after 6 years of follow-up.

Discussion: Undifferentiated high-grade pleomorphic sarcoma is an uncommon cause of malignant orbital mass. It grows fast and can metastasize, so extension studies are required. Surgical excision is the treatment of choice.

Sarcoma pleomorfo indiferenciado de alto grado orbitario: a propósito de un caso

RESUMEN

Caso clínico: Mujer de 27 años con proptosis del ojo derecho. Las pruebas de imagen mostraron una lesión extracanal en el tercio anterior medial de la órbita que afectaba al músculo recto interno. Se realizó exéresis del tumor con radioterapia adyuvante. El diagnóstico fue de sarcoma pleomorfo indiferenciado de alto grado. Tras 6 años de seguimiento no se ha observado recidiva.

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Introduction

Undifferentiated high-grade pleomorphic sarcoma, a term accepted in 2002 by the World Health Organization and otherwise known as fibrous malignant histiocytoma, is a malignant neoplasia of mesenchymal origin with an incidence of 1–2 for every 100,000 inhabitants.\(^1\) Orbit location is rare and accounts for only 1\(^\text{st}\) of the orbit tumors.\(^2\) Most frequently it affects Caucasian males between the sixth and eighth decade of life. It exhibits rapid local growth and is potentially metastasizing.\(^1\)

Clinic case

A 27-year-old female visited our service with upper right eyelid proptosis and edema. The rest of the ophthalmological exploration was within normal ranges. The patient did not have personal and familial antecedents of interests. Basic analytics were performed with thyroid hormones with normal results.

Computerized tomography (CT) and magnetic resonance of the orbit revealed the existence of a contrast-capturing extracorneal lesion having a size of 1.1 × 0.7 × 3 cm in the medial anterior third of the orbit involving the internal rectus muscle without bone involvement (Fig. 1). The tumor was excised by means of transcanaicular orbitotomy with inferior transconjunctival expansion without requiring muscle deinsertion. In the post-surgery period the patient exhibited internal rectus paresis.

Anatomopathological analysis revealed a clearly circumscribed and pseudo-encapsulated 1.5 × 1 cm nodule, peripheral inflammatory and hemorrhagic areas. The nodule comprised a proliferation of numerous fusiform cells in a vascularized collagen extracellular matrix. The said fusiform cells were arranged forming a storiform or “Malta cross” pattern (Fig. 2). Immunohistochemistry tests were positive for vimentin, actin 1A4 and actin HHF35 (Fig. 3). Staining with marker Ki-67/MIB-1, a prognostic aggressiveness marker, was 20%. Therefore, the final diagnostic was undifferentiated high-grade pleomorphic sarcoma.

The remote extension study (chest, abdomen and pelvis CT) was negative. Post-surgery orbit imaging tests demonstrated the persistence of irregular thickening in the medial orbital area with homogeneous contrast capture. Accordingly it was decided to establish radiotherapy treatment on the tumor, with a total dose of 54 Gy. After 6 years follow-up with imaging tests for screening tumor relapse and metastasis, no signs of the disease have been identified. At present, the patient exhibits a very slight limitation of right eye supradduction in adduction without clinical significance.

Discussion

Undifferentiated high-grade pleomorphic sarcoma is a malignant tumor of mesenchymal origin with predisposing factors such as traumatic or surgical history, ionizing radiation, fractures, osteonecrosis, Paget disease, non-ossifying fibroma and fibrous dysplasia.\(^3\) From the anatomopathological viewpoint, the said tumor does not exhibit specific morphology and the diagnostic is based on exclusion. Locoregional and systemic extension study must always be carried out. In radiological imaging studies these tests are usually circumscribed and contrast capturing.\(^2\) Infiltration of the extraocular muscles has been described and on some occasions it can also produce bone erosion.\(^4\) For this reason, the diagnostic possibility of undifferentiated high-grade pleomorphic sarcoma must be taken into account with the thickening of a single extraocular muscle in any patient who does not have thyroid disease.\(^5\) The differential diagnostic also includes other causes of differentiated orbital mass such as cavernous hemangioma, tumors derived from nerve sheaths and other vascular neoplasia such as hemangiopericytoma.\(^6\) The patient of the present case did not have thyroid disease and the image tests revealed a tumor invading the middle rectus muscle.

Fig. 1 – CT image with intravenous contrast showing a contrast-capturing extracorneal lesion in the medial wall of the right orbit involving the internal rectus muscle without producing bone erosion. (A) Coronal plane. (B) Transversal plane.
Fig. 2 – (A) Histological section of the surgical piece with hematoxylin–eosin staining and increase of 1.25 x showing a well-defined pseudo-encapsulated nodule with inflammatory peripheral patches and hemorrhagic areas. (B) The same piece with larger increase (20 x) showing fusiform cells arranged in storiform or “Malta cross” pattern.

Fig. 3 – Two histological sections for immunohistochemical study of the surgical piece. (A) With positive staining for vimentin. (B) Positive staining for actin HHF35.

without causing bone erosion, which was a positive prognosis factor.

The treatment of choice is surgical resection with safety margins although in this location complete excision is impossible. In the present case a broad excision was decided and, as the tumor did not produce bone erosion and the MIB-1 2 tumor aggressiveness prognostic marker was low, subsequent surgery was not performed due to the morbidity it could produce and to the functional and cosmetic cost in a young person. In the series by Delaney et al., 85% of the patients treated with partial resection and subsequent radiotherapy were controlled after 5 years. Neoadjuvant chemotherapy could reduce tumor size prior to surgery.

Local recurrence is frequent but locoregional remote metastasis is rare. In case of remote extension, the treatment of choice is surgery with adjuvant chemotherapy and required subsequent radiotherapy. Regular follow-up of patients is essential for controlling possible local relapses or remote metastasis. The patient of this case did not exhibit signs of tumor relapse or metastasis after a follow-up of 6 years.

Conflict of interests

No conflict of interests has been declared by the authors.

REFERENCES