Short communication

Juvenile nasopharyngeal angiofibroma with orbital extension

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ABSTRACT

Clinical case: The case of a 21-year-old male with a history of left proptosis and diplopia of two weeks of onset is presented. The MRI showed an ethmoid-orbital vascular lesion with anterior skull base invasion and orbital extension. Biopsy of the ethmoid confirmed fibrovascular tissue, which supported the diagnosis of angiofibroma.

Discussion: It is a benign neoplasm with local characteristics of malignancy due to its ability to invade adjacent areas. In this case, the debut presented with manifestations of orbital extension. A broad and multidisciplinary approach is needed in order to improve prognosis.

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Angiofibroma nasofaríngeo juvenil con extensión orbitaria

RESUMEN

Caso clínico: Varón de 21 años con historia de exoftalmos izquierdo y diplopía de 2 semanas de evolución. La resonancia magnética mostró una lesión muy vascularizada etmoido-orbitaria con invasión de base del cráneo anterior y extensión orbitalia. La biopsia etmoidal confirmó un tejido fibrovascular compatible con angiofibroma.


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Introduction

Juvenile nasopharyngeal angiofibroma (JNA) is a locally invasive benign tumor which almost exclusively originates in the posterior area of the nasal cavity. It expresses in preferably male teenage patients and accounts for less than 0.5% of head and neck growths.\(^1\) It corresponds to a vascular lesion the growth of which erodes the bone structures in the base of the skull with the ability to extend to the nose, paranasal sinus and orbit. Intracranial invasion has also been described.\(^2\)

The case of a patient with orbit involvement and intracranial extension due to intracranial extension of nasopharyngeal angiofibroma with ethmoidal origin is presented.

Clinical case

A male, 21 years of age, without relevant history, presented with left exophthalmos and diplopia in extreme gaze positions with 2 weeks evolution, without clinic of epistaxis or nasal obstruction (Fig. 1). Visual acuity was 20/20 in both eyes and intraocular pressure (IOP) 24 mmHg in left eye (LE) without limitations in ocular movement. Ocular fundus did not reveal alterations. Cerebral-orbital magnetic resonance (MR) exhibited a highly vascularized left ethmoidal lesion with prominent vascular structures. Orbital extension was observed with intra- and extraconal components which displaced the internal and inferior rectus. Ethmoidal cells exhibited bone destruction and extension toward the anterior cranial fossa, associating contrast capturing in the adjacent dural meningeal zone (Fig. 2). Arteriography revealed a highly vascularized tumor at the ethmoidal level, discarding the presence of arteriovenous malformations (Fig. 3). The remainder of the extension study was negative.

Initially and in the light of the clinical behavior and radiological findings an anterosuperior biopsy was performed, with the result of abundant bleeding. For this reason it was decided to postpone the intervention in order to approach the lesion through the anterior and nasal endoscopic path.

Fig. 1 – Left ocular proptosis.

Fig. 2 – MR pondered in T1 with contrast, axial section showing highly vascularized lesion with ethmoid-orbit occupation.

Fig. 3 – Internal and external left carotid selective cerebral arteriography showing orbit vascular lesion with ethmoidal branch contribution from the ophthalmic artery and distal branches of the left internal maxillary artery.

Discusión: El angiofibroma nasofaríngeo juvenil (ANJ) es un tumor benigno con características locales de malignidad debido a su capacidad de invadir áreas adyacentes. En nuestro caso, el comienzo se presenta con manifestaciones de extensión orbitaria. Consideramos necesario un conocimiento amplio y un abordaje multidisciplinario con el fin de mejorar el pronóstico.

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The lesion biopsy exhibited the presence of capillary and venous blood vessels with irregular muscular walls which in addition were dilated and covered by prominent endothelium. Said vessels were surrounded by spindle cells with oval nuclei but without atypical mitosis. Associated inflammatory infiltrate was found which produced necrosis and destruction in the ethmoid bone tissue with extension toward the adjacent sinus mucosa and intracranially toward the dura mater (Fig. 4). The immunohistochemical study revealed angiomaticus and fibroblastic properties with positive staining against smooth muscle vimentin, CD34, bcl-2, CD99, actin and myosin.

The sample analysis enabled the JNA diagnostic. Surgical approach was decided by means of anterosuperior and endoscopic orbitotomy. When the region was accessed, significant bleeding was evidenced and the tumor was partially resected. One week post-surgery MR revealed an intra-orbit nodule suggesting tumor remains and persistence of contrast capture at the frontal dural level (Fig. 5). Subsequently, a left orbit ethmoidal endonasal approach was decided to achieve almost complete removal of the mass and medial wall reconstruction (LactoSorb® Plating System, Biomet Microfixation, Jacksonville, FL, USA). Postop. MR revealed a small left frontal post-surgery collection with small nodular capture at the meningeal level (Fig. 6). Proptosis exhibited significant improvement, and ocular parallelism was maintained with discreet exophthalmos without diplopia. IOP in the LE improved to 16 mmHg.

**Discussion**

Although the exact nature of JNA is not known, the first histological studies suggested it could originate in fibrocartilage, periosteum or embryonary fascia. Subsequently, Liang et al. reviewed a series of 25 tumors which concluded a vascular origin.³

JNA is a benign neoplasia with local malignity characteristics due to its ability to invade adjacent areas. In the present case, onset comprised orbit extension expression without nasal clinic, exhibiting diplopia in extreme positions due to compromised space. In this way, the tumor invaded the posterior portion of the orbits through the media wall from the ethmoidal lamina papyracea.

The diagnostic is based on clinical findings and imaging tests which also determine the tumor extension.⁴ A differential diagnostic must be performed with other types of nasopharyngeal tumors (choanal polyp, adenoid hyperplasia), syphilis and nasopharyngeal tuberculosis, craniopharyngioma or lymphoma, among others.

JNA is a therapeutic problem due to the risk of abundant bleeding and high recurrence rates. In general, surgical treatment is preferred with or without presurgery

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**Fig. 4** – Histological sections in hematoxylin–eosin of the ethmoid bone and paranasal sinus patient samples. (A) 10× increase samples showing a typical proliferation with prominent spindle cells arranged in fascicles. (B and C) 10× increase with PAS staining showing tumoration comprised by dilated blood vessels with irregular muscular walls covered by prominent endothelium.

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**Fig. 5** – MR pondered in T1, coronal section evidencing rough dural capture at the anterior frontal level as well as intra-orbit nodule suggesting tumor remains.
In the present case, the endonasal approach facilitated control of the intra-surgery bleeding and satisfactory tumor resection. Radiotherapy allowed control of the disease in highly advanced stages. At present, after a follow-up of one year, the patient has not exhibited relapses of the disease and has been referred to our practice every 6 months.

Conflict of interest

No conflict of interest has been declared by the authors.

References