Short communication

Orbital tumor due rhinosinusal extension in a patient with Wegener’s granulomatosis

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ABSTRACT

Case report: The case is presented of a 49-year-old woman with an orbital mass originating from the rhinosinus. She had a history of Wegener’s granulomatosis, refractory to both biological and immunosuppressive therapy. Clinical examination showed proptosis, diplopia, and restriction of ocular movements.

Discussion: Orbital mass resection was performed, due to its rapid growth, and lack of response to medical treatment.

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Tumoración orbitaria por extensión rininosusal en un paciente con granulomatosis de Wegener

RESUMEN

Caso clínico: Presentamos el caso de una mujer de 49 años, con una tumoración orbitaria por extensión rininosusal, por granulomatosis de Wegener, sin respuesta a tratamientos inmunosupresores ni a terapia biológica. La paciente mostraba una marcada proptosis con diplopia y limitación de los movimientos oculares.

Discusión: Se realizó extirpación tumoral mediante abordaje nasal y orbitario, con extirpación de la tumoración orbitaria, dada su rápida progresión y refractariedad al tratamiento médico.

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Introduction

Wegener’s granulomatosis (WG) is a systemic inflammatory disease characterized by vasculitis of small and medium-sized vessels, necrosis and formation of granulomas. It can affect any organ, mainly the upper airways, lung and kidneys. It appears between the 4th and 5th decade of life. Up to half of patients can exhibit ophthalmological alterations, with initial expression ranging between 8% and 16%. The most frequent ocular expressions are orbital and keratoconjunctivitis, whereas in the upper airways the most affected areas are the nasal cavity and paranasal sinuses.1,2

The case of a female with WG history refractory to medical treatment is presented, exhibiting rhino-sinusal tumor compromise with invasion of the orbit.

Clinic case report

Female, 49, with WG of 5 years evolution, who visited the practice due to progressive proptosis of the left eye (LE), and diplopia with 3 months evolution that did not remit despite treatment with corticoids, immunosuppressants and rituximab.

Visual acuity (VA) was of 1.0 in the right eye (RE) and 0.6 in the LE. Hertel exophthalmometry was 17 and 23 mm respectively. Intrinsic ocular motility was normal whereas the extrinsic exhibited adduction limitation in the LE, with diplopia in all diagnostic positions except the primary position. LE palpation detected 2 hard, fixed and nodular tumors in the internal region of the upper eyelid. Intraocular pressure was 12 and 16 mmHg. Anterior pole and ocular fundus were normal (Figs. 1 and 2).

Orbital computerized tomography with contrast revealed tumors in nasal cavity and ethmoidal sinuses that invaded the orbit through the medial wall with lateral LE displacement (Figs. 2 and 3).

Together with the ENT Dept., surgical approach was decided upon.

Through nasal endoscopic approach, the tumor that invaded the orbit was partially resected. By means of medial orbitotomy through the palpebral groove, the tumor resection was completed despite strong adherence to deep areas. It was removed in hard, irregular, brownish red-colored fragments. No medial rectus muscle involvement was found. The histopathological result was necrotizing granulomatosis with polyanangitis compatible with WG (Fig. 4).

One month after surgery the patient no longer exhibited diplopia, had VA of 1 in both eyes, and proptosis had diminished to 18.5 in exophthalmometry (Fig. 5).
Fig. 3 – CAT with axial section (A) showing tumor displacing the ocular globe, with corona section (B) showing the orbital tumor extension as from the rhinosinusal compromise.

Discussion

WG in paranasal sinuses causes mucosa thinning, ulceration and bone destruction.²

Orbital WG expresses as nasolacrimal obstruction, dacryocystitis, inflammatory pseudotumor orbital masses.³ Involvement is generally unilaterally, presenting with proptosis, occasional pain, epiphora, conjunctival injection and restrictions in ocular movement. Diplopia develops due to the mass effect or vasculitis of the vessels irrigating the extraocular muscles.⁴

The present patient exhibited a history of 5 years with the disease as well as multiple relapses, the last one associated

Fig. 4 – (A) Rhinoscopy nasal tumor image. (B) Tumor isolation image through medial orbitotomy. (C) Tumor extirpation. (D) Part of surgical instrument.
to orbital compromise. This complication generally occurs in later stages of the disease although sometimes it can be the first expression. The orbit can be compromised due to primary inflammation and extension of the tumor from the nasopharynx sinuses toward the extraconal space.\(^5,6\)

WG prognosis varies depending on it being multisystemic or compromising a single organ, on the stage at which treatment is established, response to treatment and length of the active disease. Worsened ocular inflammation in a patient with a well managed disease could indicate worsening of systemic expressions.\(^2\)

Orbital complications are a severe threat for VA. In these patients, final VA is below 0.1 in 20–50% of cases due to optical nerve compression or to complications such as keratopathy due to exposure and ocular perforation.\(^4\)

Medical treatment with corticoids and immunosuppressants has been the main approach for treating WG.\(^1,2\) For orbital expressions, good response to medical therapy has been described,\(^7\) but in severe orbital inflammation with proptosis and particularly if the optic nerve is compromised, orbital decompression must be considered.

Holle et al. observed that the orbital tumor expressions can be resistant to immunosuppressants treatment and associate to permanent visual alteration and blindness.\(^5\)

Some authors have described the efficacy of rituximab in patients with ocular disease refractory to cyclosporin.\(^9,10\) However, in the present case the patient did not respond to treatment and, due to the risk of optic nerve decompression, surgical intervention was decided upon. After surgery, the patient exhibited a good response with ocular motility recovery, disappearance of diplopia and proptosis, achieving a favorable evolution during 12 months follow-up.

The objective of this article is to raise attention to an infrequent WG ocular expression that frequently exhibits poor response to medical treatment and where surgery could be necessary for managing the local, recurring and destructive disease.

**Conflict of interests**

No conflict of interests was declared by the authors.

**REFERENCES**