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Case Report

Granulomatosis with polyangiitis presenting with unilateral exophthalmos: A case report ☆

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ABSTRACT

We report a case of a patient with no medical history, admitted for right exophthalmos. For whom imaging showed orbital masses without inflammatory signs pointing to a granulomatous origin. However, the histological and immunological workup revealed the diagnosis of Granulomatosis with polyangiitis.

Introduction

Granulomatosis with polyangiitis formerly "Wegener's granulomatosis" is a multisystem necrotizing granulomatous vasculitis which affects preferentially the respiratory tract and kidneys. The ocular involvement is less frequent, and is rarely the initial symptom.

Case Report

A 60-year-old male patient with no medical history, presented with progressive right exophthalmos. Ophthalmic examination found a painful right exophthalmos, right eye visual disturbance, and no oculomotor palsy. The fundus examination found no abnormality. The patient was afebrile, and examination of his ears and nose was normal. An orbito-cerebral CT scan was performed showing a soft tissue mass occupying the right intraconal and extraconal compartments, slightly enhanced after contrast injection, associated with a lacrymal gland enlargement (Figs. 1 and 2), in addition to a nodular mucosal thickening of the right maxillary sinus associated with bone thickening of the sinus walls without bone lysis (Fig. 3).

MRI was not performed due to lack of financial means. Blood count and C-reactive protein showed normal values.

The differential diagnoses included malignant orbital tumor, autoimmune and inflammatory pseudotumor.

Furthermore, the immunological workup, showed positive antineutrophil cytoplasmic antibodies (ANCA) of the c-ANCA type and of PR3 specificity. A biopsy of the orbital mass

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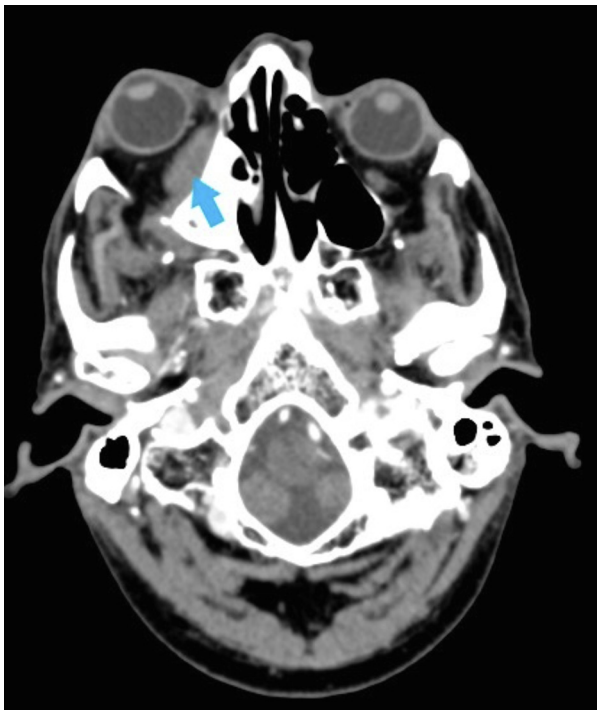

Fig. 1 – A contrast enhanced axial CT scan image showing right proptosis due to a homogeneous soft tissue mass (arrow),extending along the medial wall of the orbit, occupying the extraconal and intraconal compartments.

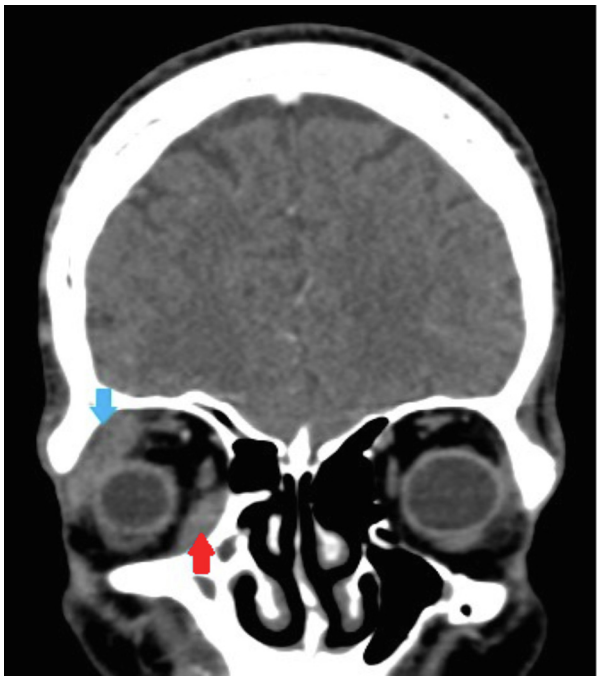

Fig. 2 – A contrast enhanced coronal reformat showing the soft tissue mass (Red arrow) and the lacrymal gland enlargement (Blue arrow).

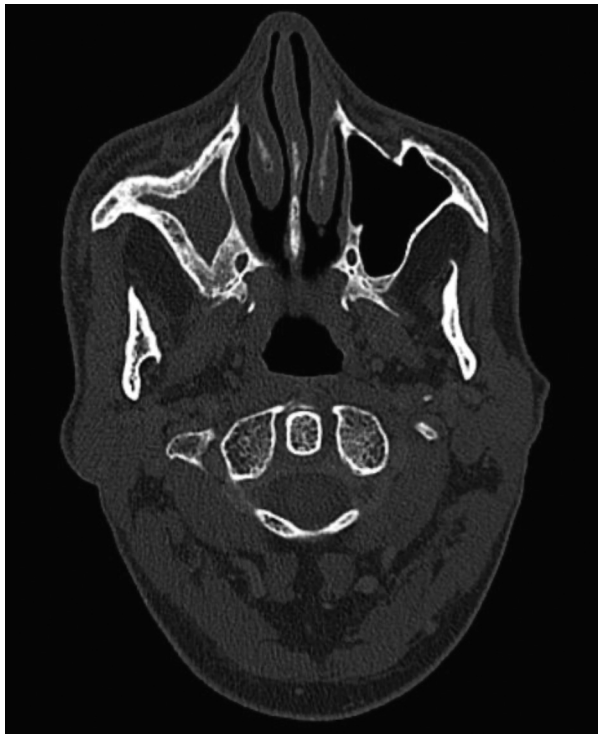

Fig. 3 – An axial CT image showing a mucosal thickening associated with wall thickening of the right maxillary sinus.

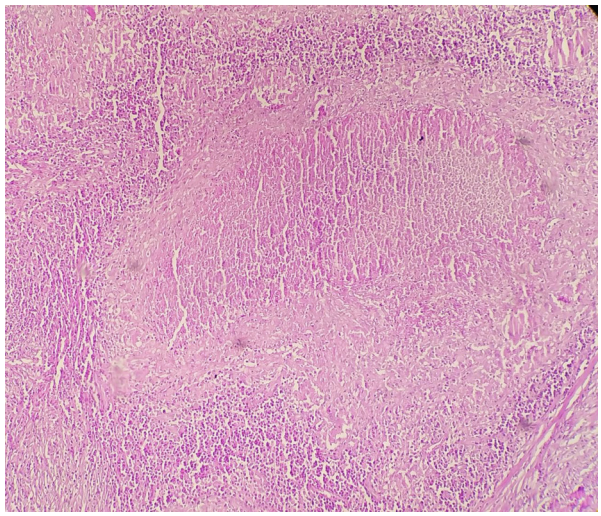

Fig. 4 – Granulomatous inflammation centered on coagulation necrosis (HEx100).

was performed, revealing Granulomatosis with polyangiitis (Figs. 4 and 5).

The patient received therapy with intravenous then oral corticosteroid, in addition to a monthly dose of 750 mg of cyclophosphamide which significantly reduced pain and restored the visual disturbance within 10 days.

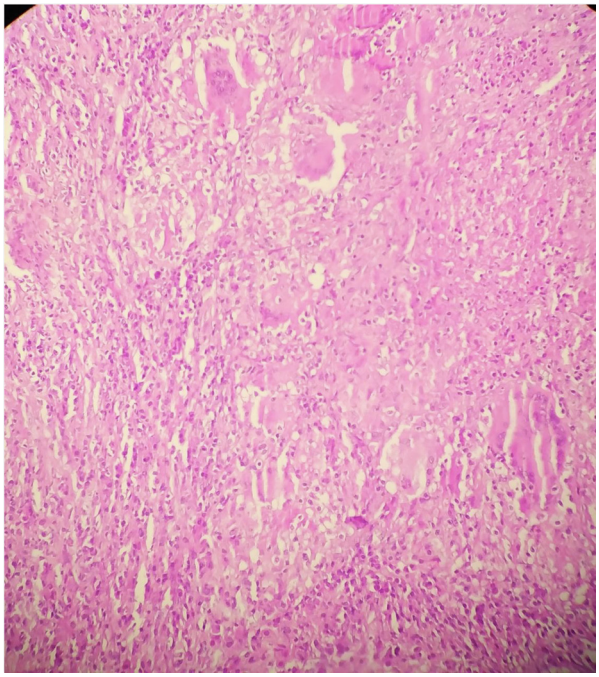

Fig. 5 – Epitheloid granuloma with many giant cells (HEx400).

Discussion

Granulomatosis with polyangiitis is a systemic disease of undetermined origin characterized by its triple tropism: Ear nose and throat (ENT), pulmonary and renal. Ocular manifestations represent 28%-52% of cases [1] but they are revealing in only 7%-41% of cases [2]. It can be manifested by exophthalmos, ptosis, palpebral edema, partial or total limitation of eye movements or diplopia [3,4]. Orbital involvement may be related to local vasculitis or to the extension of a contiguous granulomatous ENT infiltration (naso-sinusopharyngeal) explaining the concomitance of ENT symptoms [4,5]. The diagnosis is based on clinical, biological, and histological arguments.

From the radiological point of view, CT and MRI make it possible to explore the orbital masses by specifying their size and topography, to assess the extent of the exophthalmos, to analyze the lacrymal glands and ducts, the vessels, and the peripheral muscles. They also make it possible to assess the diffusion of granulomatous lesions and sinus involvement and to look for bone lysis or compression of the optic nerve. On CT, the intra-orbital masses appear homogeneous, contiguous, with nasosinus involvement. They appear isodense to the orbital muscles and are weakly enhanced afterwards; a few signs should give rise to the suspicion of granulomatosis with polyangiitis, a thickening of the nodular nasosinus mucosa with irregular tissue surfaces, punctiform bone erosions or the presence of an inter-nasosinus septum [6,7]. On MRI, the orbital masses present a clear hypo signal on the T2-weighted sequences; the T1-weighted sequences, before injection and without saturation of the fat signal, obtain a better contrast between normal structures and lesions with a slight enhancement after injection [7,8]. Despite advances in imaging, no aspect is specific to the disease, therefore, imaging findings should be correlated with biological and histological data; the presence of anti-neutrophil cytoplasmic antibodies (ANCA) subtype c-ANCA against PR3, is a major diagnostic argument and makes it possible to follow the phases of remission and exacerbation [9]. Histologically, it associates three characteristic lesions: ischemic necrosis, giant cell granulomatosis and vasculitis which affects small and medium vessels [1]. The main differential diagnoses are neoplastic pathologies such as orbital lymphoma or metastases and infectious or inflammatory pathologies (sarcoidosis or idiopathic inflammatory pseudotumors). Treatment is based on a combination of corticosteroids and immunosuppressants [10]. Early diagnosis and treatment are important to control the progression of the disease and to improve the life quality of patients.

Conclusion

Granulomatosis with polyangiitis can present as an orbital pseudotumor, with unilateral exophtalmia as the initial symptom of the disease, hence the importance of including it the differential diagnosis of orbital masses. Immunological and histological assessment is necessary to confirm the diagnosis. The evolution under treatment is very favorable.

Patient Consent

Informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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