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Case Report Open Access

Wegner's Granulomatosis Revealed by Exophthalmos

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Abstract

Ophthalmic involvement is frequent, between 30% and 70% of patient's present ophthalmic symptoms during the course of Wegner's granulomatosis. Orbital disease may present initially before the onset of systemic manifestations in only 8 to 16% of patients and it could delay final diagnosis. We report a case illustrates the diagnosis of Wegner's granulomatosis presenting with proptosis (exophthalmos) of the orbit. Patient was treated with corticosteroid and intravenous cyclophosphamide (CYC) with good response. This case emphasizes early diagnosis and treatment to avoid further complications.

Keywords: Inflammation; Orbit; Wegner's granulomatosis; Exophthalmos

Introduction

Wegner's granulomatosis (WG) is a systemic vasculitis which in its classical form, is characterised by involvement of the upper and lower respiratory tracts together with glomerulonephritis and is strongly associated with antineutrophil cytoplasmic antibodies against proteinase 3 [1].

Ophthalmic involvement is frequent, between 30% and 70% of patients present ophthalmic symptoms during the course of the illness [1,2]. Orbital manifestation may range from a mild conjunctivitis to dacryocystitis, episcleritis, scleritis granulomatous sclerouveitis, ciliary vessel vasculitis and retro-orbital mass lesions and more rarely retinal vasculitis [1,2] . Orbital disease may present initially before the onset of systemic manifestations in only 8 to 16% of patients and it could delay final diagnosis. We report a case of Wegner's granulomatosis revealed by exophthalmos, who was treated with intravenous CYC and corticosteroid with good reponse. This case emphasizes early diagnosis and treatment to avoid further complications.

Observation

A 47-year-old man had suffered from painful left exophthalmos and blurred vision for 1 month without consulting. He last exhibited purulent nasal discharge, epistaxis, generalized malaise, weight loss and fever. On examination, we found left axial proptosis and conjunctivitis (Figure 1).

He was febrile (38°C) , with a heart rate 90 beats/mn, tachypneic, blood pressure was 150/90 mm Hg. His lungs were clear to auscultation bilaterally. He also presented with symmetrical synovitis of joint (elbow and knee) with restriction of extension. A nodule of the right elbow was noted. On urinary examination both microhematuria and proteinuria were present.

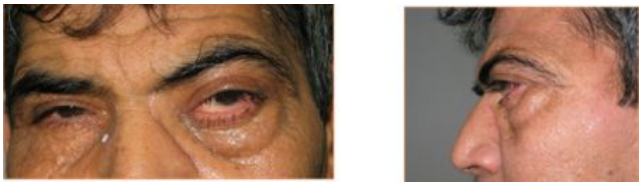

Figure 1: Showing left proptpsis amd conjunctivitis.

Laboratory studies included: WBC 12 10 3/µL, hemoglobin 11 g/dl, serum urea nitrogen 0.45 mg/dl, creatinine 121 µmol/l. The erythrocyte sedimentation rate (ESR) was 75 and the C-reactive protein (CRP) concentration was raised to 40 mg/l.

Eye examination, found ulcerative keratitis and orbital pseudotumor. Conjunctival biopsy showed necrotizing vasculitis in small vessels. A biopsy specimen of the left orbit demonstrated necrosis, granulomatous inflammation and vasculitis. No sign of vasculitis was found in fluorescein angiography. Chest X-rays and CT-scanning showed bilateral opacities (Figure 2).

Nasal biopsy showed granuloma. Electromyography was used for the diagnosis of polyneuropathy. Renal histopathologic study revealed vasculitis. Rheumatoid factor and serum antinuclear antibody were negative but c ANCA were positive and elevated. Our patient was subsequently diagnosed as having Wegener's granulomatosis. Because the patient had severe and rapidly progressive disease, he was treated with high doses of corticosteroids and oral CYC. His treatment ameliorated his glomerulonephritis and renal insufficiency. His ocular symptoms resolved and did not recur after treatment with corticosteroid in combination with cyclophosphamide.

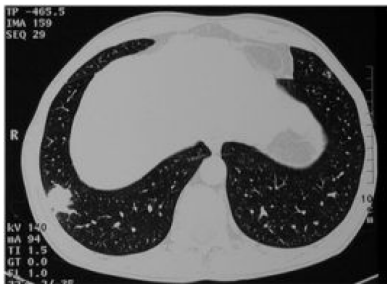

Figure 2: Scanning Pulmonary Nodule.

CT-scanning demonstrated extensive mucosal thickening in almost all sinuses including sphenoidal, frontal, ethmoidal and maxillary and a retro-orbital infiltrate without granuloma (Figure 3).

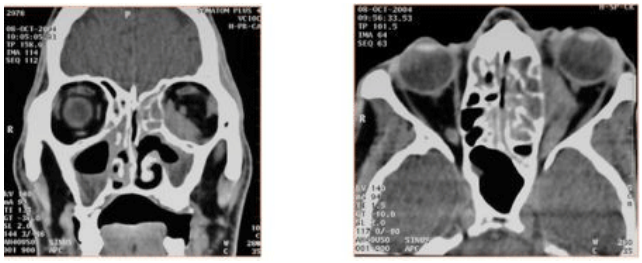

Figure 3: Extensive mucosal thickening maxillary and a retro-orbital infiltrate without granuloma.

Discussion

In 7% patients of generalized WG, proptosis is the initial presentation. Ocular/ orbital involvement is ultimately seen in 50% cases of generalized WG [2-5]. Orbital involvement occurs either due to contiguous spread from the sinuses or due to primary involvement secondary to small vessel vasculitis. The most common ocular manifestation reported are : proptosis, scleritis, peripheral ulcerative keratitis, dacryoadenitis, optic nerve vasculitis, retinal vein or artery occlusion, conjunctivitis and uveitis [6]. Orbital tumours required advanced differential diagnosis between sarcoidosis, tuberculosis and WG [7].

When clinical and serologic findings were inconclusive, biopsy remains indispensable. It demonstrates necrosis, granulomatous inflammation and vasculitis, but these histopathological triad is seen in less than 50% of orbital biopsies [4,5]. Positive ANCA may help establishing the diagnosis in cases in which typical pathological features are lacking, and it has a value in following disease activity. A combination of cyclophosphamide and corticosteroids is essential and critical not only for the ocular condition but also for the survival of the patient [6,7].

Conclusion

WG is a potentially lethal systemic vasculitis in which ocular involvement may be the first clinical presentation. Scleritis and peripheral ulcerative keratitis indicate systemic involvement, highlighting the importance of prompt diagnosis and treatment. When clinical and serologic findings were inconclusive, biopsy remained indispensable. A combination of cyclophosphamide and corticostroids is essential and critical not only for the ocular condition but also for the survival of the patient.

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