

Ocular manifestation of granulomatosis with polyangiitis

Mary Grace, Jacob K.J., Shameer V.K., Anitha Sebastian

Department of Medicine, Government Medical College, Thrissur, Kerala, India

Correspondence to Mary Grace, MD, Department of Medicine, Government Medical College, Thrissur, Kerala, India
Tel: 9446460337;
e-mail: nc.grace@yahoo.in

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Ocular manifestation of granulomatosis with polyangiitis. Proptosis is the most common ocular manifestation of granulomatosis with polyangiitis. It is due to inflammation of the extraocular muscles or vasculitis of the vas nervosum. It is a poor prognostic sign.

Keywords:

granulomatosis with polyangiitis, ocular manifestations, proptosis

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Introduction

Granulomatosis with polyangiitis is a rare disease that typically occurs in patients in their fourth to sixth decades. It can involve any organ system, but most commonly affects the respiratory and renal systems and the head and neck, including the eye. The diagnosis of the limited form of this disease is at times difficult, as imaging and tissue biopsies are often negative or nonspecific.

Case report

A 47-year-old woman presented with swelling and painful restriction of both eye movements over 3 days' duration, associated with occasional episodes of fever. No definite history of arthritis was obtained. On examination she was conscious and oriented. Her blood pressure was 150/100 mmHg, pulse rate was 102/min, respiratory rate was 14/min, and she was afebrile. Both eyes showed bilateral upper lid edema, chemosis, and conjunctival congestion (Figs. 1 and 2).

The cornea was clear, pupils were round and reacting to light, lens were clear, and no papilledema was observed. External ocular movements were restricted in all directions due to pain. Other systems were within normal limits. She gave a history of endoscopic sinus surgery for a mass in the right nasal cavity about 1 month ago.

Investigations Hemoglobin 11.2 g%, total count 9400/mm³ polymorphs 78%, lymphocytes 21% eosinophils 1% erythrocyte sedimentation rate 135, C-reactive protein 9.6 (normal <0.6 mg/dl), blood sugar 180 mg%. Urine routine examination RBC 2-3 no dysmorphic RBC pH 7.5 total bilirubin 1.1 mg%, serum glutamate oxaloacetate transaminase 22, serum glutamate pyruvate transaminase 19, alkaline phosphatase 143 total

protein 5.8 albumin 3.9 Blood urea 22 mg% serum creatinine 6 mg%.

Other investigations revealed the following: antinuclear antibody, negative; rheumatoid factor, negative; serum sodium, 142 mmol/l; serum potassium, 3.8 mmol/l; serum bicarbonate, 21; serum calcium, 6 mg%; serum phosphorus, 9 mg%; 24 h urine protein, 06 g%; urine potassium, 32 mEq/l; 24 h urine calcium, 173 mg/day (normal 100–240); serum C3, 127 (normal 90–180); Venereal Disease Research Laboratory, nonreactive; thyroid function tests, normal; HIV, HbsAg, and anti-HCV, negative; Mantoux test, negative; serum angiotensin converting enzyme level, 28 U/l (6–52); cytoplasmic antineutrophil cytoplasmic antibody (ANCA), 72 U/ml (<5); perinuclear ANCA, 1.3(normal <5); and radiography of the chest, normal.

Computed tomography (CT) of the orbit revealed bilateral, peripherally enhancing bulky heterogeneous lacrimal gland with mass effect on globe and extraocular muscles. CT of paranasal sinuses revealed pan sinusitis, soft tissue filling the right frontal and ethmoid region and maxillary sinus extending to and occluding the right nasal cavity suggestive of polyposis, no bony erosion or displacement, and right inferior turbinate hypertrophy. Biopsy of the right nasal mass revealed a suppurative granulomatous lesion with areas of necrosis that stained negatively for microorganisms; there was no evidence of malignancy. Lacrimal gland biopsy showed nonspecific inflammatory reaction.

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Figure 1



A photograph showing bilateral eyelid swelling.

CT findings of the chest were consistent with allergic granulomatosis with polyangitis.

In view of the investigation results, a diagnosis of granulomatosis with polyangitis was made. She was started on pulse methylprednisolone and cyclophosphamide. She underwent pulse steroid therapy with an injection of methylprednisolone 1 g daily for 3 days. Pulse cyclophosphamide was given once in 2 weeks for three doses, followed by once in 3 weeks for three doses. However, she showed only transient improvement. Her proptosis worsened and she developed loss of vision in one eye despite decompressive surgery on both eyes. About 2 months later she succumbed to her illness.

Differential diagnosis

Differential diagnosis of orbital inflammatory disorders includes thyroid eye disease, sarcoidosis, Churg–Strauss syndrome, Tolosa–Hunt syndrome, orbital cellulitis, tumor metastasis, lymphoma, and rhabdomyosarcoma. Normal thyroid function tests, negative ANCA, normal angiotensin converting enzyme level, and absence of malignancy on histopathology helped in ruling out these differential diagnosis.

Discussion

Granulomatosis with polyangitis is a rare disease that typically occurs in patients in their fourth to sixth decades, although it has been reported in patients with age ranging from 7 to 75 years. Upper airway disease is the most common presenting feature of granulomatosis with polyangitis, occurring in more than 70% of patients at onset and ultimately developing in more than 90% of cases. Symptoms and signs of nasal involvement include mucosal swelling with nasal

Figure 2



A photograph showing lacrimal gland swelling.

obstruction, crusted nasal ulcers and septal perforation, serosanguinous discharge, epistaxis, and external saddle nose deformity.

Pulmonary involvement is one of the cardinal features of granulomatosis with polyangitis. It occurs in 45% of cases at initial presentation and in 87% during the course of the disease. Cough, hemoptysis, and pleurisy are the most common pulmonary symptoms. Upto one-third of cases with radiographically demonstrable pulmonary lesions may not have lower airway symptoms. The most common radiological findings include pulmonary infiltrates (67%) and nodules (58%). The pulmonary infiltrates in granulomatosis with polyangitis may be quite fleeting. Persistent diffuse interstitial infiltrates are rare (<1%). Pulmonary nodules are usually multiple, bilateral, and often cavitate (50%). CT of the chest reveals infiltrates and nodules that were undetected by means of conventional radiographs in 43–63% of cases. Renal disease is estimated to occur in 11–18% of cases at presentation and in 75–80% during the entire course of the disease. Ocular manifestations have been reported to occur in 28–58% of patients and may be part of initial presentation in 8–16% of cases [1]. Any compartment of the eye may be affected. Keratitis, conjunctivitis, scleritis, episcleritis, nasolacrimal duct obstruction, uveitis, retro-orbital pseudotumor with proptosis, retinal vessel occlusion, and optic neuritis have been described. Proptosis is the most common ocular manifestation of granulomatosis with polyangitis [2]. Proptosis is due to inflammation of the extraocular muscles or vasculitis of the vas nervosum [3]. The most common symptoms are diplopia and pain on ocular movement. On examination, there is restriction of ocular motility. It is a poor prognostic sign. About one-half of patients with proptosis in one series lost vision due to optic nerve ischemia. Dacryoadenitis and lacrimal gland enlargement are recognized features of adenax involvement and may cause ocular sicca syndrome. Classical histological features of

granulomatosis with polyangitis are not always present in the biopsy material [4].

Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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