Abstract

Wegener’s granulomatosis is a systemic vasculitis of unknown aetiology. Although it classically involves the upper respiratory tract, lungs and kidneys, virtually any organ may be affected. We report a rare case of a 45 year old female who presented with bilateral dacroadenitis, otitis media, parotid enlargement and left sided lower motor neuron type of facial palsy as the initial manifestations of Wegener’s granulomatosis.

Introduction

Wegener’s granulomatosis (WG) is a clinicopathological syndrome characterised by extravascular granulomatous inflammation, granulomatous vasculitis of small-sized vessels, necrosis of the upper and lower respiratory tracts, and pauci-immune glomerulonephritis. WG and other antinuclear cytoplasmic antibodies related vasculitis are difficult to diagnose because of low prevalence and variable presentations, clinical features, mimicking common illnesses like infections, malignancies, thromboembolism and connective tissue disorders. Prompt recognition of the more unusual presentations of the disease is necessary to ensure early diagnosis and treatment. The early diagnosis and the timely immuno-suppressive therapy lead to high rates of remission of an otherwise lethal disease.

Case Report

A 45 year female presented with chief complaints of bilateral: ear discharge - two months, swelling in front of ears - one month, swelling over both upper eyelids - fifteen days and deviation of angle of mouth to right side - seven days.

Otorhinolaryngologist diagnosed her to be having acute otitis media two months back and put her on antibiotics, but there was no improvement. Over the next one month, the amount of ear discharge increased, which was purulent admixed with blood and she also developed bilateral hearing loss. One month later she developed bilateral painless swelling in front of both ears, which gradually increased in size, unassociated with dryness of mouth. Fifteen days later she developed bilateral swelling over both upper eyelids, which gradually increased in

Fig. 1: Parotid enlargement depicted in the arrow
A 20-year-old female patient presented to the hospital complaining of a persistent cough and fever of 10 days duration. Her temperature was 37.8°C. She reported a dry cough and dyspnoea on exertion. She also complained of dysphagia and dysphonia. She was a nonsmoker and was not taking any medication. She had no history of chronic respiratory disease, asthma, or allergy. She had no history of exposure to pets, recent travel, or recent contact with ill individuals.

On examination, her vital signs were in normal range. Bilateral symmetrical firm, non-tender swelling of parotids measuring 8 x 6 cms was noted (Figure 1). No pallor, icterus, cyanosis, oedema, lymphadenopathy, arthritis, nasal deformity, rashes, thyroid swelling, joint deformity, hair loss, oral or genital ulcers or photosensitive reactions were seen. In Respiratory system, left infrascapular fine crepitations were heard. Cardiovascular system and abdomen were normal. Ophthalmic consultation revealed bilateral dacryoadenitis (Figure 2), xerophthalmia and bilateral filamentary keratopathy (Figure 3). Nervous system examination revealed isolated left sided lower motor neuron (LMN) type of facial palsy (Figure 4). ENT consultation revealed bilateral otitis media, right ear central perforation with mucopurulent discharge, left ear dry perforation and bilateral sensorineural deafness.

Laboratory investigations revealed: Haemoglobin 11.2 g/dL, haematocrit 34.2%, leucocyte count 18,900/μL with neutrophilia, platelet count of 843,000/μL, and erythrocyte sedimentation rate of 136 mm/1st hour. Renal and liver function tests were normal. Tests for malaria, dengue, leptospira, tuberculosis, human immunodeficiency virus, hepatitis B and C were negative. Blood culture was sterile. Urine examination showed proteins 3+, WBCs 10-12 and

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**Fig. 2**: Dacroadenitis depicted in the arrow

**Fig. 3**: Bilateral xerophthalmia and keratitis depicted in this picture

**Fig. 4**: Left sided lower motor neuron type of facial palsy

**Fig. 5**: CT Chest showing interstitial infiltration and cavitation depicted in the arrow

**Fig. 6**: CT chest showing interstitial infiltration & cavitation depicted by the arrow
RBC casts. Rheumatoid arthritis (RA) factor: 96 IU/ml and C-reactive protein (CRP): 24 mg/dl. C-antineutrophil cytoplasmic antibody (c-ANCA) was positive in 1:10 dilution by immunofluorescence. Serum immunoglobulin (Ig) E levels were elevated (563.2 IU/ml). p-ANCA, anti nuclear antibody (ANA) test and ANA profile were negative. Serum angiotensin converting enzyme (ACE) and serum Ig A, Ig G, Ig M levels were normal. Chest X-ray revealed right side para-cardiac consolidation. CT chest revealed multiple nodules, interstitial infiltration and cavitations in bilateral lung fields (Figures 5 and 6). Biopsy of lacrimal and parotid gland revealed predominant lymphocytic infiltrate with areas of fibrosis.

A diagnosis of Wegener’s granulomatosis was made based on Positive c-ANCA, cavitative lesions on chest CT, renal involvement in the form of haematuria and proteinuria and the presence of otitis media, dacroadonitis and parotitis. The patient was started on oral cyclophosphamide (2 mg/kg/day) and prednisolone (1 mg/kg/day). Inspite of immunosuppression and steroid therapy for one day, a minimal reduction in the size of parotid and lacrimal gland and in the amount of ear discharge was observed. One month later, she developed sudden onset of bilateral loss of vision and fundus fluorescein angiography revealed bilateral central retinal artery occlusion. Fifteen days following this, she developed gangrene of the toes of the right foot. She refused further treatment and died one week later.

**Discussion and Conclusions**

WG was first described by Freidrich Wegener in 1936 and 1939. In 1954, Godman and Churg further delineated the clinical and pathological features by describing the classical triad of necrotising granulomata affecting the upper and lower respiratory tracts, disseminated vasculitis and glomerulonephritis. The aetiology of WG remains unknown.

The most common anatomical site for presenting lesions of WG is the upper airway, which occurs in 95% of patients. Patients often present with severe upper respiratory tract findings such as paranasal sinus pain and drainage and purulent or bloody nasal discharge, with or without nasal mucosal ulceration. Pulmonary involvement may be manifested as asymptomatic infiltrates or may be clinically expressed as cough, haemoptysis, dyspnoea, and chest discomfort, in 85–90% of patients. Renal disease (77% of patients) generally dominates the clinical picture and, if left untreated, accounts directly or indirectly for most of the mortality in this disease.

Otolological involvement may occasionally be the first and only sign of WG. The prevalence of ear involvement varies from 19% to 45% of all cases. Otological involvement may be divided into: (1) serous otitis media, (2) chronic otitis media, (3) sensorineuronal hearing loss, (4) vertigo, and (5) facial nerve palsy. Facial nerve palsy is due to compression of the nerve in the middle ear, especially in the presence of a dehiscent fallopian canal or due to vasculitis of its microvasculature. Facial nerve decompression is not useful but may aggravate the problem.

Ocular involvement may be seen in 30-50% of all cases of WG. Eye involvement may range from a mild conjunctivitis to dacryocystitis, episcleritis, scleritis, granulomatous sclerouveitis, ciliary vessel vasculitis, and retroorbital mass lesions leading to proptosis. Dacroadenitis may be sole presentation of the disease with absence of systemic symptoms, or may be an early or first manifestation of classic WG followed by other extra-ocular features, and should warrant a search for other features of WG.

Salivary gland enlargement is a rare feature in WG, which is mostly confined to the parotid and submandibular glands but even sublingual salivary glands can be affected. Descriptions of salivary gland enlargement as an initial symptom of systemic manifestation of WG are rare but some cases have been published.

Accurate diagnosis of WG is based on clinical, histopathological, and immunological investigations as per the criteria of the American College of Rheumatology (ACR). However, there are cases in which the ACR criteria are not fulfilled. In particular, so-called localised WG, is not considered in the ACR criteria. In these cases, an early c-ANCA test may aid diagnosis and provide essential information for early therapy and thus prevent disease progression. A disadvantage is that the sensitivity depends largely on the extent of the disease. For systemic WG, the sensitivity approaches 96%, while in localised forms, sensitivities of about 67% or less are described.

Early diagnosis is important, expediting aggressive immunosuppressive therapy with glucocorticosteroids and cyclophosphamide, which can potentially limit a more severe systemic disease progression. WG, if untreated, has a poor prognosis with a mean survival time of five months. Prompt recognition of the more unusual presentations of the disease is necessary to ensure early treatment.

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References


