Wegener's granuloma – a case report

Swapan Kr. Ghosh · Saumyajit Datta · Alok Banka

Abstract A 75-year-old man presented with nasal obstruction, purulent nasal discharge, crusting and a reddish ulcerated swelling over nose and upper lip. There was external nasal deformity. Histology of the lesions showed necrotic granulomatous vasculitis. There was no other systemic involvement. ANCA test was negative. The patient responded well to prednisolone and cyclophosphamide.

Keyword Wegener's granuloma.

Introduction

In 1931, Heinz Klinger of University of Berlin first reported patients who had died with prolonged sepsis with inflammation of the blood vessels throughout the body. About five years later Dr. Fredericke Wegener found a distinct syndrome in three patients. In 1954 a definite criteria was described for the diagnosis of a specific disease attributed with the name, Wegener's Granulomatosis (WG). WG is characterized by granulomatous inflammation of the respiratory tract, necrotizing vasculitis affecting small to

medium sized arteries and necrotizing glomerulonephritis. Although in its classical form WG is a multisystem disease, there can be a spectrum of clinical manifestations and the disease may present with limited organ involvement [1]. Detection of cytoplasmic antineutrophilic cytoplasmic antibody (c-ANCA) is a valuable adjunct to the diagnosis of WG, but it may be negative in localized forms of the disease [2]. Patients without renal involvement are said to have limited Wegener's. There is no known cause for W. G. although it is known not to be contagious or hereditary.

Case report

A 75-year-old male patient from rural West Bengal presented with foul smelling purulent discharge from the nose with nasal obstruction and crusting and occasional mild pain. He also complained of weakness and anorexia with gradual loss of weight. There was a reddish swelling over external nose and upper lip. The swelling was ulcerated and the surface of the swelling was granular. The external nose was deformed. The patient gave a history of trauma over nose and upper lip 8 years back. Granulation tissue formed over the site which increased in size gradually and became ulcerated. He was treated by some private doctors with antibiotics but the disease was not controlled. Then he got treatment in a cancer hospital where biopsy was done. The histopathology report stated that there was no malignancy but could not specify it as WG. Dorsum of nose became deformed 4 years back. There was destruction of cartilaginous portion of septum with collapse of nasal bridge. The patient gave history of pulmonary tuberculosis. He completed the antitubercular course. No family history of a similar or related illness was reported. On clinical examination, the regional lymph nodes were not enlarged. Blood examination revealed leucocytosis with elevated ESR (100 mm/1st hour) and microcytic hypochromic anaemia (Hb-9gm%). Serum glucose, urea and creatinine estimation revealed

S. K. Ghosh¹ · S. Datta² · A. Banka³

¹Associate Professor
ENT Department, Medical College and Hospital,
Kolkata, India

²MO

³PGT

ENT Department,
NRS Medical College,
Kolkata, India

S. K. Ghosh (✉)
Flat 101B, Girikunj,
390, S.N.Roy Road,
Kolkata - 700 038
India

no abnormality. X ray PNS revealed bilateral maxillary sinusitis with mucosal thickening and X ray chest revealed scarry lesion in right upper lobe suggestive of healed tuberculosis. 3 consecutive sputum examinations were negative

Fig. 1 Wegener's Granuloma involving external nose and upper lip

Fig. 2 CT scan showing involvement of right nasal cavity and right maxillary sinus

for AFB. A CT scan of the paranasal sinuses showed a soft tissue shadow in the right nasal cavity extending laterally into the right maxillary antrum. USG of the abdomen failed to reveal any discernable abnormality. Histopathological examination revealed necrotic granulomatous vasculitis. An ANCA test was negative. The patient was treated by a combined regiment of prednisolone 1mg/kg body weight and cyclophosphamide 2 mg/kg body weight orally. There was gradual improvement of the symptoms with reduction of the swelling over external nose and upper lip within a period of 6 weeks.

Discussion

There are three main forms of WG: types 1, 2 and 3. Type 1 is the limited form of WG. Typically, the patients present with symptoms of an upper respiratory tract infection persisting for several weeks, which is unresponsive to antibiotics and associated with serosanguinous nasal discharge, pain and crusting in nasal cavities. In type 2 there is nasal involvement similar to the patient with type 1 disease, but other organs are involved. Type 3 is widespread disease with involvement of multiple organs including airway, pulmonary, renal, and sometimes cutaneous lesions [3].

WG can appear in patients of any age, but it typically manifests during middle age. It is rarely seen in black population. It affects both sexes equally, although a slight predilection toward men has been reported. It is a potentially life-threatening disease, but complete remission

Fig. 3 Microphotograph showing granulomatous vasculitis (H & E X 100)

can occur if it is diagnosed early and treated properly with immunosuppressants like cyclophosphamide and corticosteroids in particular. Otherwise median survival is only 5 months. The most common early clinical manifestation of WG is ear, nose and throat involvement (92% of patients). The turbinates, nasal bones and the nasal septum are most commonly affected. The common signs and symptoms are persistent nasal obstruction, nasal discharge, sinus pain, epistaxis, hoarseness of voice, otorrhoea and hearing loss. Systemic symptoms like fever, malaise, night sweats, weight loss and migratory arthralgias may occur. In severe and advanced cases erosion of the orbital wall and other facial bones has been seen [4]. In the present case there was involvement of upper lip and nose. There was destruction of cartilaginous septum with collapse of nasal bridge but there was no involvement of orbital wall.

The typical history and characteristic findings on examination and the presence of mild to moderate anaemia associated with an elevated ESR are all suggestive of the clinical diagnosis of WG [3]. The diagnosis is confirmed histopathologically. A positive c-ANCA test is very important in confirming the diagnosis of WG, but these autoantibodies may be absent in a proportion of cases, particularly in the limited variety of the disease. The sensitivity of this test was found to be 67% by immunofluorescence and 60% by ELISA for patients with active local or regional symptomatology [5]. So a negative c-ANCA test does not exclude the diagnosis of WG. The sensitivity of the test depends on the extent and activity at the time of study [3]. In the present case the c-ANCA test was negative.

In the present case the patient was treated by cyclophosphamide and steroid. A comparative study of the classical and localized forms of the disease found that the groups shared many features, particularly their requirement for immunosuppressive therapy, since WG causes much tissue destruction regardless of whether it is a localized or a widespread process. The localized WG group may change the pattern of their disease to involve the kidney [6]. So long-term follow-up of such patients is necessary.

References

1. Gravallesse EM (1993) Systemic vasculitis. In: Fitzpatrick TB, Eisen AZ, Wolff K et al, editors. *Dermatology in general medicine*, 4th ed. New York: McGraw-Hill, pp 2169–2171
2. Nolle B, Specks U, Ludemann J, Rohrbach MS, DeRemee RA, Gross WL (1989) Anticytoplasmic autoantibodies: their immunodiagnostic value in Wegener granulomatosis. *Ann Intern Med* 111: 28–40
3. Thomas J McDonald (1998) Manifestations of Systemic Diseases of the Nose, Ed. Charles W. Cummings et al, *Otolaryngology Head & Neck Surgery*, (Mosby), 3rd Edn., 844–846
4. Hoffman GS, Kerr GS, Leavitt RY et al (1992) Wegener granulomatosis: an analysis of 158 patients. *Ann Intern Med* 116:488–498
5. Ghosh A, Bandyopadhyay D, Basu S et al (2004) ANCA-negative limited Wegener's granulomatosis. *Indian J Dermatol Venereol Leprol* 70:102–104
6. Luqmani RA, Bacon PA, Beaman M, Scott DG, Emery P, Lee SJ et al (1994) Classical versus non-renal Wegener's granulomatosis. *QJ Med* 87: 161–167