Wegener's Granulomatosis in the Elderly

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ABSTRACT

Wegener's granulomatosis classically involves the upper respiratory tract, lungs and kidneys. Rarely, it also affects the skin and heart. Cardiac involvement is uncommon in Wegener's granulomatosis and myocardial infarction is seldom highlighted. It can be a difficult diagnosis to make in the elderly who often have multiple co-existing illnesses. We present a case of a 75-year-old Chinese woman with interesting cardiac and dermatological manifestations of Wegener's granulomatosis.

Keywords: pustules, aged, cutaneous, vasculitis

INTRODUCTION

Wegener's granulomatosis is a granulomatous disease characterised by necrotising granulomata and vasculitic lesions which typically involve the upper respiratory tract, lung and kidneys. The frequency of upper respiratory tract, lung and kidneys lesions at presentation in a study by the British Thoracic Society Research Committee has been reported to be 75%, 63% and 60% respectively[1]. However, Wegener's granulomatosis is a systemic disease and can rarely present at non-classical sites. This can be misleading in the diagnosis of elderly patients where multiple illnesses often can co-exist. We present an unusual case of an elderly patient with Wegener's granulomatosis.

CASE REPORT

The patient was a 75-year-old Chinese woman with a history of dementia and essential hypertension. She was a non-smoker and did not have a previous history of ischaemic heart disease or diabetes mellitus. The patient was admitted in September 1996 with a generalised rash and purulent right nasal discharge for about one week. She had crops of pustules, papules and ulcers over her face, trunk and limbs (Fig 1), and the dermatologist's opinion was that of either septic emboli or primary pyoderma.

Echocardiography did not show the presence of vegetations and the left ventricular ejection fraction at that time was 62%. Blood cultures done repeatedly did not grow any microorganisms. Computerised tomographic scan of her nasal sinuses revealed presence of pansinusitis and an endoscopic drainage of the frontal, ethmoidal and sphenoidal sinuses was done. A biopsy of the right ethmoidal sinus mucosa showed only the presence of chronic granulomatous inflammation which was negative on the Ziehl-Nielsen stain. Her electrocardiogram and chest X-ray were unremarkable. Serum creatinine was 74 emol/L. She was treated with cloxacillin and amoxycillin/sulbactam for the skin lesions which resolved subsequently. Following the drainage of her sinuses, fluticasone propionate nasal spray was prescribed for the sinusitis.

Four months later, she was readmitted for acute deterioration of her mental state, anorexia and drowsiness. Serum creatinine was mildly elevated at 182 emol/L. Electrocardiography showed atrial fibrillation and poor R wave progression. New murmurs and a pericardial rub were noted and subsequent echocardiography indicated the presence of a small pericardial effusion and polyvalvular incompetence. In addition, the left ventricular ejection fraction had decreased to 26% and regional wall motion abnormalities were present, consistent with a silent myocardial infarction which had occurred sometime over the past few weeks. The circulating antineutrophil cytoplasmic antibody level was assayed with a positive titre of 1/320 and a diagnosis of Wegener's granulomatosis was made. Intravenous hydrocortisone was started but her renal function deteriorated rapidly with her serum creatinine rising to 678 emol/L by the second week of admission. Haemodialysis was recommended but both the patient and her family members declined further treatment and the patient died shortly after of renal failure.

DISCUSSION

Wegener's granulomatosis is a rare disease typically seen in the middle-aged population and the mean age of diagnosis range from 41 to 50 years[1,2]. The diagnosis is often delayed with a mean time from the onset of symptoms to diagnosis of 15 months[2]. In our patient, the diagnosis was established 5 months after the presentation of her initial symptoms. Given the multi-systemic involvement of Wegener's granulomatosis and the presence of multiple co-existing diseases in the elderly, establishing the diagnosis of Wegener's granulomatosis in the elderly is particularly challenging and probably accounts for the delay in the diagnosis and treatment. In a study by Anderson et al, the prevalence of renal involvement at presentation in his series was 60%[1]. However, this significant feature may be easily overlooked in the elderly and a raised serum creatinine is often attributed to other causes. Similarly, the presence of myocardial ischaemia in the elderly may be wrongly ascribed to the commonly occurring atherosclerotic coronary arterial disease instead of Wegener's granulomatosis.

Cardiac involvement is uncommon in Wegener's granulomatosis with an incidence of 6% to 12% in two large series[2,3]. In this small subgroup of patients, pericarditis was the most common manifestation[2,3]. Myocardial infarction due to active Wegener's granulomatosis is even less well recognised and has only been described in a few case reports although a retrospective review by Walton in 1958 revealed that about 28% of the cases had focal necrotising arteriolitis in the heart[4-7]. An autopsy on a 29-year- old housewife with Wegener's granulomatosis was reported by Reed et al to show a diffuse gross pattern of left ventricular and septal myocardial infarction, and microscopic examination revealed massive necrotising arteriolitis of a coronary artery[6]. Papo et al described two middle-aged patients with no previous history of angina, who developed silent myocardial ischaemia

during a flare-up of Wegener's granulomatosis, which was diagnosed by electrocardiography and echocardiography[5]. Our patient resembles these patients and her myocardial infarction may have been secondary to Wegener's granulomatosis rather than being a coincidental event. The incidence of myocardial infarction in Wegener's granulomatosis may be underestimated as it can occur silently. Consequently, screening electrocardiograms may be useful for patients with Wegener's granulomatosis.

The skin is involved with an overall incidence of about 45% during the course of the illness[2,3,8]. However, it is less common as a presenting feature with a frequency of only 1% to 13%[2,8]. Recognised dermatological lesions include purpura, nodules, pustules, ulcers and necrotic papules[2,6,8]. Skin manifestations appear more often in patients with renal involvement[8]. In retrospect, the rash which our patient had during her initial hospital admission was probably a dermatological manifestation of Wegener's granulomatosis.

Before the 1950s, when treatment of Wegener's granulomatosis was only supportive, it has been reported that more than 80% of patients with Wegener's granulomatosis would be dead within a year of diagnosis from renal failure[7]. The mean survival period of untreated disease is 5 months and the delay in diagnosis may be particularly important in affecting mortality[7]. In 2 later series[1,2] which reviewed cases beyond 1975, renal failure, active pulmonary involvement and infections were amongst the most common factors leading to deaths in patients with Wegener's granulomatosis. Although the mortality is higher in the elderly despite treatment with immunosuppressive agents, the side effects of therapy are not significantly different to that experienced by patients of a younger age group[9]. The elderly are also more likely to remain in clinical remission compared to the young[9]. Hence, treatment with cyclophosphamide and glucocorticosteroids is justified in the elderly. Unfortunately, our patient chose not to receive further treatment.

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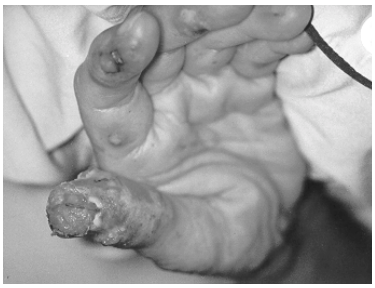

Fig 1 - Cutaneous ulcers and pustules on the patient's hand.