Granulomatous vasculitis of the penis with glomerulonephritis

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Summary

Wegener's granulomatosis is characterized by a necrotizing granulomatous vasculitis of the upper and lower respiratory tract together with glomerulonephritis. Presentation is usually with symptoms referable to the upper respiratory tract, the glomerulonephritis manifesting itself weeks or months later. A case is reported of a man presenting with a necrotizing granulomatous vasculitis of his penis who subsequently developed glomerulonephritis 18 months later.

Case report

A 67-year-old Caucasian presented with a long history of ischaemic heart disease and was receiving maintenance digoxin and diuretics. In July 1978 he presented with a 4-month history of painless swelling of the glans penis associated with purulent discharge and dysuria. Circumcision and biopsy was undertaken, histology showing marked acute and chronic inflammation with vasculitis and several foci of necrosis. Antibiotic therapy was commenced. ESR was 20 mm/hr (Westergren) and renal function normal. The symptoms failed to resolve and partial amputation of the penis was performed in September 1978. Histology showed extensive granulomatous inflammation with vasculitis and areas of necrosis (Figs 1 and 2). In September 1979 he was admitted with a myocardial infarct. Dysuria was again present and, in addition, nocturia, anorexia, nausea and weight loss of recent onset. There were no respiratory symptoms. ESR was 85 mm/hr and plasma creatinine 136 $\mu\text{mol/l}$. Disopyramide and spironolactone were added to his medication. While in hospital he developed haematuria, pains in the knees and ankles, and a papular purpuric rash appeared on his buttocks, ankles, feet, elbows and backs of hands. There was erythema over the joints of the hands and elbows. At this stage all drugs were discontinued. The ESR rose to 140 mm/hr and renal function steadily deteriorated, the plasma creatinine rising to 549 $\mu\text{mol/l}$. Blood cultures were negative, platelets

and clotting tests normal. Fifteen days following admission he had a cardiac arrest and died.

Post-mortem examination demonstrated a recent anterior myocardial infarction with thrombotic occlusion of the anterior descending branch of the left coronary artery. There was a cardiac aneurysm involving the posterior wall of the left ventricle. The remainder of the left ventricle was severely hypertrophied, and the pericardial sac was obliterated by an extensive fibrinous pericarditis. The valves were normal and showed no vegetations. No aneurysms of coronary, renal or other arteries were found. The upper respiratory tract was normal showing no evidence of a necrotizing process. The lungs were moderately oedematous but contained no discrete lesions. The bone marrow appeared normal. No other significant abnormality was found elsewhere in the body. Histologically the kidneys showed a focal necrotizing glomerulonephritis (Fig. 3).

Discussion

A granulomatous vasculitis of either the upper respiratory tract or lungs is a cardinal feature of both limited and classical Wegener's granulomatosis (WG) (Fauci and Wolff, 1973). However, it has been pointed out that in certain circumstances the diagnosis of WG should be considered in anticipation of the appearance of respiratory involvement with a view to starting cytotoxic immunosuppressive therapy at an early stage (Baker and Robinson, 1978).

In the case reported here, lesions histologically indistinguishable from those occurring in the respiratory tract in WG were present on the penis while the respiratory tract was uninvolved. Later the patient was found to have a focal necrotizing glomerulonephritis which is one of the typical renal lesions of WG. The delay between initial presentation and subsequent detection of renal involvement is also reminiscent of WG.

Reed *et al.* (1963) found focal ulceration of the skin as an initial feature in 5% of cases of WG. In

FIG. 1. Histological section from penis showing granulomatous inflammation (HE, $\times 350$).

FIG. 2. Histological section from penis showing vasculitis (HE, $\times 350$).

FIG. 3. Histological section from kidney showing focal necrotizing glomerulonephritis (martius, scarlet, blue, $\times 350$).

the present case, ulceration of the skin of the penis was the first abnormality noted. These authors also described cutaneous lesions in association with terminal renal involvement that closely resemble those present in this patient. Arthralgia or true arthritis of varying severity has also been frequently reported in WG (Fauci and Wolff, 1973). It seems likely therefore that this case represents an unusual presentation of WG, the initial lesion being situated on the penis instead of the classical site of the upper respiratory tract.

The significance of the penis in this respect may be that it contains a mucocutaneous junction. Lesions of WG in the mouth, another mucocutaneous junction, are well recognized as initial manifestations of the disease (Scott and Finch, 1972). The early occurrence of mucocutaneous lesions in WG has led to the suggestion that if, as seems likely, the disease is caused by a hypersensitivity mechanism, primary sensitization may be occurring at sero-mucosal surfaces (Edwards and Buckerfield, 1978).

The penis may therefore be an alternative route of sensitization to the mouth.

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