Dermatomyositis/polymyositis and carcinoma of the ampulla of Vater

Sur., The association of dermatomyositis/polymyositis (DM/PM) with malignancy has been recorded in several reports and reviews. Although cases of carcinoma of the pancreas and dermatomyositis have been reported, we have found no report of carcinoma of the ampulla of Vater with DM/PM. We wish to report such an association.

A 62 year old woman was admitted to hospital because of fever and chills. Two weeks before admission she developed increasing fatigue, persistent sore throat with chills and fever reaching 39-4°C, night sweats, malaise, weight loss, pain in her left knee, and a morbilliform rash which in five days assumed an urticarial appearance. On admission, a painful tender left knee and oedematous dusky erythema on the periorbital region were noticed. Her temperature was 39°C, pulse 95 beats/min, and the blood pressure 125/80 mmHg. The rest of the systematic examination was unremarkable. A tentative clinical diagnosis of dermatomyositis was made.

Laboratory investigations showed erythrocyte sedimentation rate 100 mm/h, leucocytes 13.8×10⁹/l with a shift to the left (total granulocytes 90% and lymphocytes 10%), and packed cell volume 40%. Alkaline phosphatase was more than 200 SIU (normal<75 SIU). Serum aspartate transaminase 126 U/l (normal<27 U/l), serum alanine transaminase 117 U/l (normal<30 U/l), lactate dehydrogenase 290 U/l (normal<290 U/l), and γ-glutamyl transferase 224 U/l (normal<30 U/l). The following were normal or negative: renal function studies, bilirubin, hepatitis B surface antigen, heterophil agglutinins, creatine phosphokinase, aldolase, amylase, thyroid function tests, rheumatoid factor, antinuclear antibodies, antimitochondrial antibodies, smooth muscle antibodies, serum complement levels, cultures from throat, urine, and blood, tuberculin skin test, stool specimen, chest x rays, electrocardiogram, electromyogram (EMG), upper gastrointestinal study, intravenous pressure, ultrasonographic study, and the computed tomographic scan of the abdomen. A muscle biopsy showed typical changes of fragmented and degenerated muscle fibres in a background of fibrous tissue heavily infiltrated by leucocytes (Fig. 1).

Three weeks later pyrexia continued and the patient developed jaundice with pruritus and ascites. Her condition deteriorated, she had a massive haematemeses, and died. The postmortem examination showed an anaplastic adenocarcinoma of the ampulla of Vater (diameter 1.5 cm). Liver histology showed acute cholestasis. Pancreas and spleen were normal. No metastases or other primary tumours were found.

This case represents an example of DM/PM satisfying the proposed criteria. The patient developed the characteristic skin findings of dermatomyositis with mainly the cutaneous leucocytoclastic vasculitic lesions, a rare manifestation of DM/PM. Muscle enzymes and EMG were normal. Other authors have also reported cases without EMG or muscle enzyme changes, but with characteristic histological changes of polymyositis. In a recent review
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the development of jaundice prevented us from establishing the diagnosis and undertaking radical treatment.

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References


there is no mention of any case of DM/PM with pancreatic carcinoma, though such cases have been described.4 5 The presenting signs and symptoms in this case were typical of the onset of cancer of the ampulla of Vater.6 7 Unfortunately, the complicating massive haematemesis soon after

Fig 1. Myositis of the deltoid muscle (inflammation with mononuclear cells between muscle fibres and destruction of muscle). (Haematoxylin and eosin).
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