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 periobital 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 transference 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, aspartate aminotransferase, alanine aminotransferase, alkaline phosphatase, γ-glutamyl transpeptidase, and lactate dehydrogenase.

Weeks later pyrexia continued and the patient developed jaundice with pruritus and ascites. Her condition deteriorated, she had a massive haematemesis, 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.1 The patient developed the characteristic skin findings of dermatomyositis with mainly the cutaneous leucocytoclastic vasculitic lesions, a rare manifestation of DM/PM.2 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.3 In a recent review

References

the development of jaundice prevented us from establishing the diagnosis and undertaking radical treatment.

1st Department of Internal Medicine,
Medical School of Athens,
Laikon General Hospital,
Goudi, Athens 115 27,
Greece

G VAYOPOULOS
C CONSTANTOPOULOS
C FOTIOU
PH KAKLAMANIS
PH FESSAS

References