Short Communication

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Systemic Lupus Erythematosus and Lymphoma

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Key Words

Lymphoma

Systemic lupus erythematosus

Abstract

2 cases of non-Hodgkin's lymphoma developing in young women with systemic lupus erythematosus (SLE) are described. The interval between the diagnosis of SLE and the development of lymphoma was 4 years and 4 months, respectively. In 1 patient the lymphoma was localised primarily in the lung. It is suggested that the development of lymphadenopathy in a patient with SLE should be an indication for early lymph node biopsy.

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Introduction

Green et al. [1978] described 4 female patients and cited 8 others who had SLE and developed lymphomas in the course of the disease. Since then 3 further cases have been described in the UK literature [Barrett et al., 1978; Gibbs and Seal, 1978]. We present 2 further cases which have recently occurred.

Case Reports

Case 1

A 15-year-old West Indian female born in the UK presented in 1974 with flitting arthritis and fever. She later developed a butterfly rash and alopecia. Investigations showed haemoglobin 10.3 g/dl, ESR (Westergren) 106 mm/h, ANF strongly positive, LE cells positive. A diagnosis of SLE was made and she responded well to prednisolone and remained well on a dose of 10 mg daily.

In November 1978, painless lymphadenopathy developed in her neck, axillae and inguinal regions. Gland biopsy showed a diffuse lymphoblastic lymphoma and she was treated with a combination of vincristine, adriamycin and prednisolone (VAP). The lymphadenopathy resolved and since March 1979 she has been in good health maintained on 6-mercaptopurine, methotrexate, cyclo-phosphamide and prednisolone.

Case 2

In October 1978 a 22-year-old female presented with pleurisy, fever and jaundice. The spleen was enlarged 6 cm. She was pancytopenic although the marrow showed active haemopoiesis. Further investigations showed alkaline phosphatase 160 IU, AST 150 IU, serum bilirubin 80 mmol/l, ANF positive (IgG 14 IU) and chest x-ray normal. Since she was extremely ill she was empirically treated with steroids.

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The clinical improvement was dramatic and the laboratory tests reverted to normal. When the steroids were tailed off in December 1978, she relapsed with fever, anaemia, pleurisy and nasal ulceration. Investigations showed haemoglobin 6.1 g/dl, white cells $1.8 \times 10^9/\text{litre}$, platelets $25 \times 10^9/\text{litre}$ ANF positive (IgG 14U), DNA antibody 33 U/ml (normal < 10), chest x-ray normal. Nasal mucosal biopsy revealed chronic inflammation. Prednisolone was re-started at 60 mg daily with clinical improvement and the blood count returned to normal.

In January 1979, she developed further malaise and intrapulmonary coin lesions were seen for the first time on the chest x-ray. She then suffered a spontaneous pneumothorax. At thoracotomy a nodule was removed which was found to consist of diffuse histiocytic lymphoma. After a stormy post-operative period the patient was eventually treated with VAP but died 3 days later following a further pneumothorax. At necropsy, the lungs were infiltrated by tumour as were the kidneys and adrenals. The lymph nodes, spleen and bone marrow were not involved. Histological interpretation was difficult but favoured extra-nodal diffuse histiocytic lymphoma.

Discussion

Since SLE is relatively common, it is curious that the association with lymphoma should be so uncommonly recognised. The reason for the association is obscure but it is possible that the abnormality of immune surveillance causing SLE may also predispose to the development of lymphoma. In case 2 the interval between the diagnosis of SLE and the development of lymphoma was short and it is possible that in this patient the 'SLE syndrome' resulted from the mechanism of tumour-derived antigenaemia, as suggested by Green et al. [1978]. It is interesting that this patient developed primary malignant lymphoma of the lung. This condition is extremely uncommon [Sakula, 1979] but has been reported in one other patient with SLE [Gibbs and Seal, 1979] who also had undifferentiated non-Hodg-kin's lymphoma.

In view of these cases, and despite the fact that benign lymphadenopathy frequently occurs in SLE [Cammarata et al., 1963], we would suggest that the development of lymphadenopathy in a patient with SLE should be an indication for early lymph node biopsy.

References

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