Familial systemic lupus erythematosus—a case report

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THE occurrence of systemic lupus erythematosus (SLE) in more than one member of a family has been increasingly reported. Brunjes and his colleagues (1961) reviewed world literature and found 20 reports, occurrence in sisters being the more frequently reported. Of the 20 reports, six involved two sisters, two involved three sisters, and in one instance four sisters had SLE. Mother and daughter featured in six reports and there was one separate report for brother/sister, mother/son, brother/brother, father/daughter, and aunt/niece. The age of onset varied between two and 50 years. Dubois (1972) gathered evidence of 66 cases in 30 families, and subsequently eight cases in four additional families. Evidence that genetic factors play a part in the aetiology of SLE has come from the reporting of SLE in nine pairs of identical twins (Spector *et al.*, 1973), and First (1973) has emphasized the importance of obtaining a truer incidence of familial SLE to evaluate the genetic factors.

Case report

Miss C. A. and Miss R. A. were the only two children of Scottish parents, now dead. The mother died at the age of 80, and the father at 75 years of age. Neither parent had any condition remotely resembling SLE.

Miss C. A.

Miss C. A. was born in 1910. In January 1953 she developed bronchopneumonia, treated at home with intramuscular penicillin. She could not work for six weeks and subsequently she developed pain and swelling of the proximal phalangeal joints of both hands. She was referred to hospital when her haemoglobin was 10 g/100 ml and ESR 102 mm/h. A diagnosis of rheumatoid arthritis was made and treatment began with gold and phenylbutazone. Six weeks later she was symptomatically improved and her ESR was 60 mm/h.

In January 1954 the joint pain in her fingers recurred, with pain in the wrists and shoulders, and the ESR was 74 mm/h. In March 1955 her ESR was 103 mm/h and her white cell count 3,800/cm with 72 per cent polymorphs. Calcium aspirin in high dosage was prescribed and by August 1956 her ESR was 76 mm/h. In January 1959 her ESR was 99 mm/h but her arthritis was not causing her much inconvenience. By May 1961 her ESR had fallen to 22 mm/h, but on 10 October 1961 she awoke with retrosternal pain, was short of breath, and examination revealed a tachycardia and bilateral pleural effusions. An ECG showed no great abnormality, but an x-ray showed the heart shadow to be greatly enlarged. A considerable number of lupus erythematosus cells were present. Hypergammaglobulinaemia was demonstrated and SLE diagnosed. She responded quickly to prednisolone and has remained reasonably well since, her main disabilities being joint pains and the development of skin haematoma and sloughing after slight trauma, or even without trauma.

Miss R. A.

Miss R. A. was born in 1907. In 1958 she consulted frequently compaining of shortness of breath on exertion, transient facial rashes, and on one occasion an enlarged cervical lymph node which subsided without treatment. In December 1958, following a cold, she developed fever, aches and pains in the limbs, and sweating. Clinically she had bronchopneumonia but hospital admission revealed the presence of hypergammaglobulinaemia, an ESR of 55 mm/h, Hb 9·8 g/100 ml, a leucopenia of 3,000 WBCs with normal differential, and bilateral pleural effusions. No LE cells were demonstrated but she recovered quite quickly on prednisolone.

In 1959 she developed some nodules on her legs, one of which was excised. The histology showed hyperkeratosis and keratotic plugging consistent with the diagnosis of SLE. In 1961 ANF tests became strongly positive at 1/1,600, and tissue antibodies for SLE were also strongly positive. In 1972 the tests were repeated and remained strongly positive, and she developed further joint pains. Prednisolone was again prescribed. In 1973 she suffered a myocardial infarction, and

deteriorated in health, suffering a terminal cerebrovascular infarction in 1975.

Discussion

The occurrence of similar cases of disease in a family could be due to chance, but the number of cases of familial SLE now reported in the world literature, and the number of cases in one family—a total of four sisters was reported by Brunjes and his colleagues (1961), and three by Leonhardt (1957)—may seem more than chance.

Leonhardt (1964) and Larsson and Leonhardt (1959) have shown increased frequency of hypergammaglobulinaemia in families where there is a case of SLE. They have also shown, in these cases, that hypergammaglobulinaemia preceded the development of SLE. Other studies (Larsen, 1972) have confirmed the increased frequency of hypergammaglobulinaemia, antinuclear factor, rheumatoid factor, and rheumatic diseases in relatives of patients with SLE compared with controls.

Larsen considers that three or more of the following criteria are crucial for the diagnosis of SLE: intermittent fever, polyserositis, myocarditis, nephritis, pulmonary lesions, lupus skin lesions, skin haemorrhages, numerous LE cells, thrombocytopenia, leucopenia, and hypergammaglobulinaemia.

References

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Clinical judgement and antibiotic use in general practice

Illustrated booklets containing standard physical but variable social and psychological information relating to 16 patients with sore throats were posted to 1,000 general practitioners. They were asked whether they would prescribe antibiotics in each case. Replies were received from 634 doctors, and it was found that the variations in social and psychological history provided had significantly affected the doctors' responses.

Reference

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