Case Report

Erosive rheumatoid arthritis co-existing with systemic lupus erythematosus. A report of a case, also showing atlanto-axial subluxation


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SUMMARY A 28-year old female is described with a 12-year history of seropositive, erosive rheumatoid arthritis. Subsequently, she developed systemic lupus erythematosus diagnosed both serologically and on renal biopsy. As has previously been emphasised, these two diseases occur rarely in the same patient. HLA typing revealed the patient was HLA - B8 positive and that she had inherited this genetic marker from her mother who is Irish, rather than from her West Indian father. The patient has developed marked atlanto-axial subluxation, a feature not noted in six patients previously described as having both diseases.

Key Words: Rheumatoid Arthritis, SLE, Atlanto-axial subluxation.

INTRODUCTION

The co-existance of erosive rheumatoid arthritis (RA) and systemic lupus erythematosus (SLE) with biopsy-proven renal disease within the same patient is rare. A recent report, reviewing the world literature cited a total of six cases (1). Little clinical information on the families of these cases was recorded and no HLA typing was described. We describe a patient who developed both RA and SLE, whose family was clinically examined and HLA typed.

CASE HISTORY

The patient was born in 1954. She is the eldest of three children, her mother being Irish and her father West Indian. She was well until 1970 when she presented with a one-month history of pain and stiffness in both knees. Her erythrocyte sedimentation rate (ESR), Westergren method, was 73 mm/1 hour and her latex test positive, titre 1 : 128. She soon developed morning stiffness and
was treated with soluble aspirin, four tablets (300 mg) daily.

In July 1970 she was admitted as an emergency case with a four-day history of neck stiffness and right-sided weakness. A presumptive diagnosis of viral meningio-encephalitis was made (the cerebrospinal fluid contained 194 w.b.c./c.mm mostly lymphocytes) and she made a rapid, complete recovery. It was noted at this time that her antinuclear antibody (ANA) test was negative and no LE cells could be found. By the end of 1970, her polyarthritis had affected her knees, hands and feet. Rheumatoid arthritis was diagnosed and gold therapy instituted.

However, in February 1971 she developed a rash on both cheeks. Although it was thought that the gold might be responsible and therapy was stopped for a short period, the possibility of this being a lupus rash was considered.

In June 1971 her ANA became positive (titre 1: 40) and SLE was thus considered likely. However, between 1971 and 1975 despite several courses of gold injections she developed gross erosive changes notably in the wrists (figure 1). She had bilateral excision of the ulnar styloids with wrist synovectomies and synovectomy of the flexor tendons of the index, middle and ring fingers. Histology of the synovial tissues from the wrists showed proliferation of the synovial cells with the formation of villous patterns. A fibrinous exudate was present within the synovial cavity and a chronic inflammatory cell infiltrate was found in the subsynovial tissue composed of lymphocytes, plasma cells and some eosinophils. The appearances were considered to be compatible with those of rheumatoid arthritis.

Gold therapy was finally stopped in 1976 and D-Penicillamine treatment prescribed from 1977 to 1979. It was then deemed to be responsible for the proteinuria she had developed and was stopped. Investigations in mid-1979 included a positive ANA (1:160) CH50,64% (determined by a minor modification of the Autoanalyser method of Nydegger et al. (2)), normal C3 and C4 estimations (by single radial immunodiffusion), immune complexes were detected in the serum (using a C1q binding technique) and a 24-hour urine collection showed Ig protein. In view of mild hepatic enzyme elevation, liver biopsy was performed but showed mild, non-specific inflammatory changes only. A renal biopsy done at this time showed generalised and diffuse deposition of small quantities of IgG on the glomerular capillary basement membranes and in the mesangium. No immune complexes were found however, and it was considered that the Penicillamine therapy was the most likely cause of the changes observed.

In April 1979 she developed a left pleural effusion which responded to Prednisolone, initially 40 mg/day reducing to 7.5 mg/day within two months.

In July 1980 she noted marked dyspnoea and increasing joint pain and stiffness. There was radiological evidence of shrinking lungs, and she was also found to be thrombocytopenic (45 - 75 x 10⁹/1). For three months she was also given azathioprine which was then changed to hydroxychloroquine. However, she developed a pneumococcal infection of the right wrist and a septicaemia; this responded to appropriate antibiotic therapy.

Fig. 1: X-ray of both wrists and hands, post-ulnar styloideectomy, showing severe erosive damage to the carpal bones.