

patients tends to confirm (but not necessarily prove) that FER is a malignant disease. Because the first child showed evidence of disease recurrence after one year of chemotherapy, we treated the second for 24 months. Although both patients died, no definite tumour was seen in either at necropsy.

Chemotherapy may have predisposed both children to fatal infection. In the first child an opportunistic pathogen was responsible for a terminal illness mimicking Crohn's disease; chemotherapy possibly also contributed to the episode of necrotising enterocolitis.

We conclude that patients with FER may, as a result of appropriate combination systemic chemotherapy with intrathecal methotrexate, enjoy good quality, prolonged remissions of disease. Whether or not they can be cured in this way remains to be seen. Infants treated with 'CHOP', and probably other combination therapy regimens, must be observed closely for unusual complications.

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Sjögren syndrome and lupus erythematosus nephritis

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SUMMARY A 9 year old girl with symptoms of the Sjögren syndrome showed interstitial lymphocytic infiltrate on renal biopsy. Two years later she had clinical and biological evidence of systemic lupus erythematosus, as sociated with a typical glomerulonephritis.

Sjögren syndrome (keratoconjunctivitis sicca and xerostomia with or without another autoimmune disease) is uncommon in children.¹ We studied two successive renal biopsies in a child who progressed from Sjögren syndrome without autoimmune disease to systemic lupus erythematosus.

Case report

A 9 year old girl was admitted to this hospital for recurrent parotitis. The first parotid enlargement

had been noticed four years previously, when she was admitted to another hospital because of convulsions. At that time treatment with phenobarbitone had been prescribed and this had been continued for four years.

Physical examination showed bilateral conjunctivitis without corneal erosion, and cutaneous vasculitis with purpuric pattern around the nails and the mouth, associated with erythema of the palms. Laboratory investigations showed a high serum protein value (108 g/l), a raised IgG value, and circulating immune complexes detected by the Clq binding test. She had a positive antinuclear antibody test (1/1600 speckled pattern), but her anti-DNA test (on *Crithidia luciliae*) and extractable nuclear antigen test were negative. Her complement value was normal. Her blood creatinine concentration was normal and neither proteinuria nor haematuria were detected.

On sequential salivary scintigraphy, the uptake of

per technetate by the parotid glands was reduced and the oral activity delayed. Biopsy of three minor salivary glands showed a diffuse lobular lymphocytic infiltration with partial destruction of acinar tissue, corresponding to grade III on Tarpley's grading.² A skin biopsy, performed on a sun-exposed area, showed a mild lymphocytic perivascular infiltrate, with IgM and IgG deposits at the dermal-epidermal junction and around dermal capillaries on direct immunofluorescence. On renal biopsy (Fig. 1), a heavy lymphoplasmocytic infiltrate was observed in the interstitium but no immunofluorescent deposit could be found either in the glomeruli or the interstitium.

A diagnosis of Sjögren syndrome was established, but as associated systemic lupus erythematosus was not fully ascertained a non steroid anti-inflammatory treatment was prescribed. The child was followed regularly for two years, without incident.

In 1982, when the girl was aged 11 years, another relapse of parotitis occurred, proteinuria was found, and she was admitted to hospital. Clinical examination was normal except for the cutaneous vasculitis. Laboratory analyses showed proteinuria (2 g/day), a serum albumin value of 22 g/l and serum creatinine concentration of 80 $\mu\text{mol/l}$. Her IgG value was high and an antinuclear antibody test (1/10240, speckled pattern) was positive. Anti-DNA (1/160) and extractable nuclear antigen tests (1/320 for anti-SSB) were also positive. Her complement values were 26 mg/100 ml for C3, and 4 mg/100 ml for C4 (60% below mean for normal serum donors). The association of systemic lupus erythematosus with Sjögren syndrome was ascertained and a second renal biopsy (Fig. 2) was performed. There was still heavy lymphocytic infiltrate in the interstitium but mild diffuse mesangial proliferation of the glomeruli was also present. No glomerulus was available in the

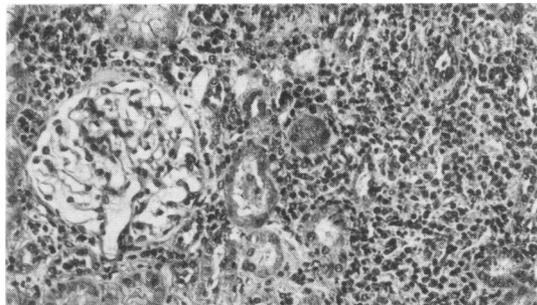


Fig. 1 *First renal biopsy: heavy lymphocytic infiltrate in the interstitium with normal glomerular structure. (HES \times 40).*

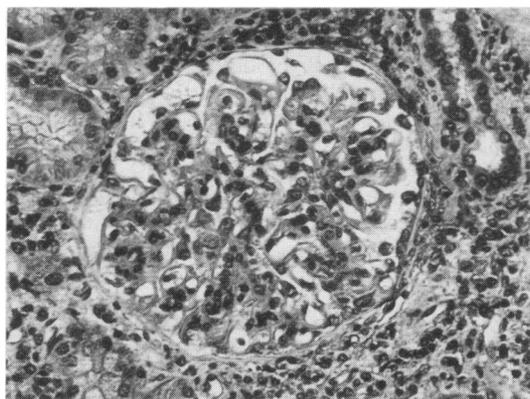


Fig. 2 *Second renal biopsy: mild diffuse mesangial proliferation and interstitial lymphocytic infiltrate. (HES \times 40).*

tissue sample studied by direct immunofluorescence.

Treatment with prednisone (2 mg/kg/day) and plasmaphereses (on 8 occasions) successfully reduced proteinuria and anti-DNA antibodies, and raised the serum complement value. The child has been followed up regularly for the past year. She has been taking prednisone (10 mg/day) and has remained well.

Discussion

There are two major points of interest associated with this patient—whether or not Sjögren syndrome and systemic lupus erythematosus were initially associated, and the interpretation of the sequential renal biopsies.

Association between Sjögren syndrome and systemic lupus erythematosus. Recurrent bilateral parotitis, bilateral conjunctivitis, and alterations of the sequential salivary scintigram with lymphocytic infiltration of minor salivary glands were characteristic of Sjögren syndrome. The first time we examined the child two features only could be attributed to systemic lupus erythematosus—her antinuclear antibody test was positive (but her anti-DNA and extractable nuclear antigen tests were negative and only later became positive), and there was a cutaneous vasculitis with IgG and IgM deposits around dermal capillaries and at the dermal-epidermal junction. Thus, except for the skin biopsy findings, clinical and biological features of Sjögren syndrome preceded those of systemic lupus erythe-

matusus. A similar case of Sjögren syndrome and subsequent development of systemic lupus erythematosus has been reported in a 12 year old boy.³ In our patient, two points remain to be clarified, whether the seizures were the first symptom of systemic lupus erythematosus central nervous system involvement and whether any relation can be found between the phenobarbitone treatment and the onset of systemic lupus erythematosus.

Interpretation of sequential renal biopsies. The first renal biopsy showed notable interstitial lymphoplasmocytic infiltrate without glomerular alteration. Similar features have been reported in Sjögren syndrome,⁴ but in some cases of systemic lupus erythematosus, where interstitial lymphocytic infiltrate has been found on renal biopsy,⁵ the initial nephropathy worsened and rapidly progressed to renal failure.⁶ On the second renal biopsy a mild diffuse proliferative nephritis typical of systemic lupus erythematosus was found, but the interstitial lymphocytic infiltrate persisted. Thus, the sequential renal changes in our patient may have been those of systemic lupus erythematosus, with lym-

phocytic interstitial infiltrate at the beginning, progressing to the typical glomerular lesions.

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Systemic carnitine deficiency exacerbated by a strict vegetarian diet

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SUMMARY A 12 year old boy suffered episodes of vomiting, lethargy, and hypoglycaemia from the age of 1 year. Adhering to a vegetarian diet caused an increase in frequency and severity of the attacks. It was found that he was suffering from systemic carnitine deficiency that responded promptly to treatment with L-carnitine.

Carnitine deficiency, first described in 1973,¹ can manifest itself in two forms—myopathic and systemic.² In myopathic carnitine deficiency, carnitine concentrations are reduced only in muscle, but in the systemic form, carnitine concentrations are reduced in several tissues, and plasma concentrations are also reduced. The patients often present in infancy with features resembling Reye's syndrome, (encephalopathy, fatty liver, raised hepatic en-

zymes, and hypoglycaemia). Recently, dietary dependent carnitine deficiency has been described.³ We report a child with systemic carnitine deficiency that was aggravated by a strict vegetarian diet. treatment with L-carnitine resulted in a dramatic improvement.

Case report

The patient, who is now 12 years old, was first evaluated at the age of 3 years. He was born after a normal pregnancy and delivery to healthy parents. His birthweight was 3.2 kg. At the age of 1 year he suffered his first episode of vomiting and lethargy, and low serum glucose values were noted. He was treated with intravenous glucose and was discharged after three days. Since then similar episodes occurred every few months. When first evaluated at our hospital a blood glucose of 20 mg/dl (1.1 mmol/l)