Chronic relapsing inflammatory polyneuropathy complicating sicca syndrome

Sir: Sjögren’s syndrome is a multi-system autoimmune disease. The cardinal features of dry eyes and mouth are caused by a destructive mononuclear cell infiltration of the lachrymal and salivary glands. By themselves, these features are thought of as the sicca syndrome whereas involvement of other organ systems, in a connective tissue disease, comprises Sjögren’s syndrome. Lesions of the peripheral and central nervous systems are described in both syndromes but are rare.1-3 In this report a case of the sicca syndrome is described associated with chronic relapsing inflammatory polyneuropathy.

A 64 year old caucasian woman was diagnosed in 1977 as having sicca syndrome. She had the characteristic dry eyes and dry mouth. A Schirmer tear test showed a marked reduction of tears in both eyes. She was also found to have pernicious anaemia with the demonstration of gastric parietal cell antibodies positive at 1:10 and positive intrinsic factor antibodies. Thyroid, thyroglobulin and microsomal antibodies were also positive. Antinuclear antibodies were negative. In November 1981 she developed generalised weakness in her legs. There was a mild improvement initially, followed by a marked deterioration during February 1982. This relapse improved spontaneously a few weeks after its onset. She continued to improve until October 1982. She then suffered an acute deterioration in leg power. During November 1982 her arms became involved and she was unable to brush her hair. Over the next 2 to 3 months she became unable to walk. Examination at this time showed mild bilateral facial weakness, weakness of the trapezius muscles bilaterally with weakness of neck flexion. She had a generalised reduction in power, especially proximally with asymmetric sluggish reflexes and flexor plantar responses. The CSF protein was elevated at 0·8 g/l without cells and there was no oligoclonal pattern.

Nerve conduction studies showed a median distal motor latency of 6·5 m/s with a motor conduction velocity from the elbow to the wrist of 24 m/s. Using a surface electrode over the right extensor digitorum brevis muscle, there was a distal motor latency of 7·0 m/s and a common peroneal nerve motor conduction velocity from knee to ankle of 29 m/s. A sural nerve biopsy showed a moderate reduction in the myelinated nerve fibre population. There was no evidence of active nerve fibre breakdown, demyelination or remyelination but there were moderately numerous regenerative clusters. A muscle biopsy performed at the same time was consistent with chronic partial denervation.

She was treated with prednisolone 1 mg/kg and azathioprine 2 mg/kg daily. Over the next 3 months she improved, but even on this therapy, she then relapsed becoming rapidly weaker. During the first week of March 1983, she became tetraplegic and suffered respiratory embarrassment with a vital capacity between 800 and 1300 ml. At this time, whilst deteriorating, she received a course of five plasma exchanges. Each exchange removed three litres of plasma. After the first exchange she was stronger and in particular, there was improved movement in the hands and fingers. With each subsequent exchange there was further recovery in ventilatory function and limb power. She was continued on prednisolone 1 mg/kg and azathioprine 2·5 mg/kg on which she was maintained over the next 18 months, when with full recovery the therapy was reduced and subsequently stopped. Up to now there has been no further relapse.

The patient reported had a relapsing polyneuropathy demonstrated by the reduced motor conduction velocities to be demyelinating in type. Peripheral neuropathies have been described in Sjögren’s and the sicca syndrome,1 but these cases were mainly sensory distal axonopathies. Vincent et al2 reported the case of a 53 year old woman with six episodes of recurrent cranial nerve palsies, the diagnosis of Sjögren’s syndrome being made 7 years after the first episode. Others have reported on cranial neuropathies, in particular sensory trigeminal neuropathy. Mixed and asymmetric neuropathies together with monoclonal multiplex can also occur but these are uncommon.4-8 Marbini et al9 have reported on a case with chronic progressive hypertrophic neuropathy. They comment on the histological similarities between their case and some reported cases with chronic relapsing polyneuropathy.10 Both chronic relapsing polyneuropathy and Sjögren’s syndrome have an HLA association and it is surprising that no previous cases have been reported. Cases of chronic relapsing inflammatory polyneuropathies are associated with HLA A1, B8, DRW3 haplotype11 and in Sjögren’s syndrome there is an association with the HLA B8, DW2 and DW3 haplotype12,13 or associated with rheumatoid arthritis, HLA D4.14 A recent report13 included five out of eight patients with chronic demyelinating inflammatory polyneuropathy who had associated autoimmune disease. This lends support to the notion that the disease may have an autoimmune basis though other series have not had such a clear association.16

The mononeuropathy may be the result of vasculitic damage to the vasa nervorum through direct infiltration of nerve sheaths with mononuclear lymphocytes may also produce a neuropathy.2 The sural nerve and muscle biopsies in this case failed to reveal either vasculitis or lymphocytic infiltration. The patient responded well to the combined therapy of immunosuppression and plasma exchange. This was in keeping with a recent study, including this case, where immunosuppressive therapy with prednisolone (1 mg/kg), azathioprine (2-0-2·5 mg/kg) and plasma exchange was found to be more effective than immunosuppressive therapy alone.16 A retrospective analysis of cases2 has shown that corticosteroid treatment was of little benefit in the CNS complications of Sjögren’s syndrome. Steroids may however be of benefit where the peripheral nervous system involvement is more marked and clearly vasculitic, but this was not the case in two out of three patients recently reported.1

Previous publications have commented on the similarity of Sjögren’s syndrome to SLE in the involvement of the nervous system. This patient could not meet the minimum clinical criteria for having SLE, though the presence of chronic relapsing inflammatory polyneuropathy would also be uncommon in this disease.17,18

I am grateful to Professor PK Thomas for permission to report this case.

MICHAEL GROSS

References


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**Letters**

Streptococcus bovis meningitis

Sirs: *Streptococcus bovis*, a Group D non-enterococcal organism, is a common cause of endocarditis but an uncommon cause of adult meningitis. Differentiation between enterococcus and *S bovis* is important because *S bovis* is sensitive to penicillin alone and is positively associated with underlying gastrointestinal disease such as colonic cancer.1,2 We describe a case of adult *S bovis* meningitis and review eight other cases from the literature in the English language.

A 59 year old man had a past medical history of adult-onset diabetes mellitus, excessive alcohol use, cirrhosis, and iron deficiency anaemia. In 1978, barium enema and colonoscopy had suggested a right colonic mass, but exploratory surgery revealed no neoplasm. In September 1980, ascites and personality changes had developed, both of which improved when spironolactone, hydrochlorothiazide, and neomycin were administered. In November 1980, polymyalgia rheumatica had developed; the patient responded well to prednisone therapy, 5 mg per day. Except for a change from neomycin to lactulose in 1981, medications had been the same, and his condition had remained stable. He had recently been drinking two to three ounces (60–80 g) of alcohol per day.

On 6 March 1984, the afternoon before elective cataract surgery, the patient was admitted to Kaiser Permanente Medical Center, Oakland, California. That evening, sudden, severe frontal headache developed, followed by vomiting and shaking chills. When examined, he was slightly confused without asterixis or papilloedema. His temperature was 40°C (104°F), pulse rate 112/min and regular, blood pressure 150/70 mm Hg, and respiration rate 18 per min (unlaboured). His neck was supple. Results of a cardiovascular examination were normal. The abdomen was soft and not tender; the liver was firm 4 cm below the costal margin. There were no focal neurological deficits. Initial chest radiographs and urinalysis results were normal. A leukocyte count was 2,500/cu mm with 67 polymorphonuclear leukocytes (PMNs) and 18 band forms. A second physical examination nine hours later showed no change except for a stiff neck with pain on flexion. Lumbar puncture revealed an opening pressure of 27 cm H2O and grossly cloudy cerebrospinal fluid (CSF). The CSF had a leukocyte count of 8,600/cu mm (98 PMNs, 2 lymphocytes), glucose of 110 mg/dl, protein of 610 mg/dl. Gram stain of the CSF was negative. Two of two blood cultures at 24 hr and CSF culture at 48 hr all grew Group D *Streptococcus bovis*. The minimal inhibitory concentration (MIC) and minimal bactericidal concentration (MBC) for penicillin were both less than or equal to 0·06 μg/ml.

The patient received a 14 day course of penicillin G, 18 million units intravenously per day. He had not clinically improved by the third day, and streptomycin 1 g intra- muscularily per day was added. The MBC results were not available this time. The temperature returned to normal on the fifth day. Mental status returned to normal on the seventh day.

Repeated abdominal examinations showed no tenderness or ascites. Bone marrow aspiration, computed tomography (CT) scan of the head, two-dimensional echocardiogram, and barium enema results were all within normal limits. A dental examination showed diffuse periodontal disease, one loose left central incisor, and no evidence of abscess. At follow-up examination three weeks later, the patient was well, working, neurologically intact, and abstaining from alcohol.

Nonnepocococcal streptococcal species account for about 10% of meningitis. Group D streptococci are an uncommon cause of meningitis; few data exist on enterococcal meningitis.3 Information on nonenterococcal Group D meningitis is limited to isolated case reports.

Both enterococcal and nonenterococcal Group D streptococci cause endocarditis; *S bovis* appears to cause endocarditis as frequently as enterococcus.4,5 Both infections are similar in clinical presentation but differ in two major aspects. First, *S bovis* septicemia occurs in the setting of underlying gastrointestinal disease, especially gastrointestinal neoplasms, and with oral disease. Second, *S bovis* is usually susceptible to concentrations of penicillin easily achieved in the blood, whereas enterococcal endocarditis requires combined penicillin-aminoglycoside therapy for cure. Mortality is greater in enterococcal disease.6

Differentiating between enterococcal and *S bovis* meningitis has similar implications. A review of cases in the English language literature yielded eight reported cases of adult *S bovis* meningitis, to which we have added our case. Three were associated with colonic disease, three with endocarditis, two with steroid use, and two with dental disease. Almost all occurred simultaneously with septicemia. The clinical presentation included negative Gram stain of the CSF in six of seven cases and a CSF leukocyte count of less than 2,000/cu mm in five of eight specimens.

Gram stain was also negative in two adults cases of enterococcal meningitis reported by
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*J Neurol Neurosurg Psychiatry* 1987 50: 939-940
doi: 10.1136/jnnp.50.7.939

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