Case reports

Case presented
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Polyarteritis nodosa presenting as meningoencephalitis

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Central nervous system involvement in polyarteritis nodosa (PAN) is much less frequent than peripheral neuropathy. It usually occurs late in the disease and presents as stroke or as diffuse encephalopathy with normal cerebrospinal fluid^{1,2}. In the case reported here, meningoencephalitis was the first evidence of organ involvement in a woman in whom polyarteritis nodosa was ultimately diagnosed.

Case report

A 49-year-old English woman was first seen in this hospital in May 1982. She had been well until two years previously, when she began to feel tired and unwell and lost two stone (12.6 kg) in weight. She was seen at another hospital with arthralgia, skin rash and anaemia. No diagnosis was established and the rash and arthralgia remitted.

Two days before admission to this hospital she developed a severe headache and over the next 24 hours became confused and drowsy. On admission she was semi-comatose. There was no neck stiffness, but bilateral papilloedema without haemorrhages or exudates was noted. There was a generalized macular erythematous rash, and her temperature was 38°C. She was normotensive. There were no other abnormalities.

Investigations included: Hb 10.1 g/dl, WBC $21 \times 10^9/l$ (96% neutrophils), ESR 48 mm/1st hour, urea 13.9 mmol/l, creatinine 183 $\mu\text{mol/l}$. There were no cells and no protein in the urine. A computed tomographic scan of the brain was normal. Lumbar puncture yielded clear cerebrospinal fluid (CSF) under normal pressure. CSF protein was slightly raised (600 mg/l). The glucose was normal. There were 88 WBC/mm³ and 1 RBC/mm³. Cytological examination of 400 cells showed that 90% were neutrophils.

There was no evidence of infection: cultures of blood and CSF for bacteria, mycobacteria and fungi were negative. Blood and CSF serology for syphilis, brucella and a range of viruses showed no evidence of recent infection, and tissue culture and electron microscopy of CSF were negative for viruses. The EEG was diffusely abnormal with increased slow activity. Tests for a range of autoantibodies, including antinuclear factor and rheumatoid factor, were negative and anti-DNA antibodies not found in the

serum. Complement levels were normal and tests for cryoglobulins and hepatitis B were negative. A skin biopsy revealed no evidence of vasculitis, but polyarteritis nodosa was felt to be a likely diagnosis.

The patient was treated with prednisolone 60 mg/day with rapid improvement in the level of consciousness, although she continued to complain of mild headache. The fever and rash settled and renal function returned to normal. She was discharged home on prednisolone 20 mg/day.

Over the next two years she had several episodes of mild headache, rash and arthralgia accompanied by raised ESR and leukocytosis, which settled with an increase in steroid dose. Papilloedema regressed slowly.

In 1984 she was readmitted with a severe exacerbation of headache and blurred vision, skin rash and fever. CSF examination on this occasion again showed a pleocytosis with 20 WBC/mm³, of which 62% were neutrophils. There was no evidence of infection. Fluorescein angiography suggested chronic ischaemic papillitis. Again symptoms settled rapidly with an increase in prednisolone to 60 mg/day.

Confirmation of the diagnosis of polyarteritis was sought. Biopsies of skin (on two occasions) and muscle showed no evidence of vasculitis, so renal angiography was performed. This showed irregularity and occlusion of small vessels with multiple small aneurysms and patchy areas of ischaemia, consistent with a diagnosis of polyarteritis nodosa.

Discussion

Patterns of neurological involvement differ considerably between the vasculitides¹. In PAN, peripheral nerve involvement is especially common (50–75%)¹ with mononeuritis multiplex being characteristic^{2,3}. The reported incidence of central nervous system involvement varies from 8%⁴ to 40%¹. In Rose's series⁵ it was noted that CNS involvement (which occurred in 37%) was often partly attributable to hypertension or atheroma and uraemia, and drug treatment may also be implicated.

Two patterns of CNS disease in PAN were described by Moore and Fauci². There may be a diffuse encephalopathy with normal CSF or focal lesions due to haemorrhage or infarction. Seizures may accompany diffuse encephalopathy. In their series and others^{2,4–8}, sterile meningoencephalitis with headache, impaired consciousness and CSF pleocytosis – as occurred in our patient – were not described, although Ford and Siekert⁷ described CSF abnormalities in 2 patients with cerebral infarcts.

The differential diagnosis of aseptic meningoencephalitis does include some forms of vasculitis and it is interesting that in Behçet's disease, where CNS involvement may occur in up to 28% of patients^{9,10}, aseptic meningitis is a common neurological presentation and peripheral neuropathy infrequent.

Several features of this patient's illness, especially the recurrence of fever, rash and arthralgia at the

same time as headache and CSF neutrophilia, and the dramatic response to steroids, suggest that an exacerbation of small vessel vasculitis was responsible. This pattern of CNS involvement in PAN is very unusual.

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Erythema chronicum migrans

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Lyme disease, a recently described zoonosis that affects man, is caused by *Borrelia burgdorferi* and is transmitted by the bite of a hard tick¹. A patient with erythema chronicum migrans (ECM), the cutaneous hallmark of Lyme disease, is now reported and the clinical features, diagnosis and incidence of Lyme disease in the United Kingdom are discussed.

Case report

A 68-year-old woman presented with an asymptomatic enlarging red ring on her right upper thigh. Four months previously, whilst on holiday on the family farm in County Mayo, Republic of Ireland, she developed an itchy red papule behind her right knee which she attributed to an insect bite. After 10 days a slowly enlarging patch of erythema appeared around the bite. The erythema then disappeared from the centre of the affected area, leaving a ring which slowly expanded. Over the next 4 months the upper part of the ring migrated cranially and the lower caudally, with eventual disappearance of the latter. She gave no history of systemic symptoms at the onset or during the evolution of the lesion.

Examination showed an indurated red 1 × 0.5 cm nodule in the right popliteal fossa consistent with a persistent insect bite reaction. Careful inspection of the right thigh showed a faint and slightly indurated 1 cm wide rim of erythema which encircled the thigh, compatible with ECM. General examination was otherwise normal.

Skin histology of the lesion behind the knee showed a heavy mixed cellular infiltrate in the upper and mid-dermis composed of lymphocytes, histiocytes and eosinophils. Although no mouth parts were

Figure 1. Histology of bite reaction. Diffuse dermal infiltrate of lymphocytes, histiocytes and eosinophils. No mouth parts seen. (× 70)

identified, the histology was compatible with an insect bite reaction (Figure 1). Histology from the erythematous rim showed a light perivascular lymphohistiocytic infiltrate in the upper dermis (Figure 2). Warthin-Starry stains were negative for spirochaetes. Skin from both lesions did not yield spirochaetes when cultured for 3 weeks in modified Barbour-Stoenner-Kelly medium. Antibody of the IgG class to *B. burgdorferi* was detected by indirect immunofluorescence at a titre of 512 (positive titre 64).

Oral penicillin V, 250 mg four times daily, produced complete disappearance of the encircling rim of erythema within 3 days. There have been no sequelae to date.

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