Letters

Clinica Neurologica,
2a Facoltà,
University of Naples,
Italy.

Address for correspondence:
Dr L Bilo, Trav. M Pietzavalle 54, 80131 Napoli, Italy.

References


Accepted 22 September 1987

Nocardia asteroides infection of the cauda equina

Sir: Nocardiosis is most often localised to the lungs and skin, but frequently disseminates to involve the central nervous system. Primary extrapulmonary nocardiosis is extremely rare1–5 and nocardiosis of the nervous system most commonly presents as a cerebral abscess metastasising from a primary pulmonary focus.6–8 Purulent meningitis following rupture of a cerebral abscess into a ventricle is well recognised,9 but primary nocardial meningitis has only been reported twice to our knowledge.10 11 We describe a patient who initially presented with a rapidly progressive glomerulonephritis in the context of systemic vasculitis. Following successful treatment with prednisolone, cyclophosphamide and plasma exchange, she developed signs initially suggesting a multifocal peripheral neuropathy but then rapidly deteriorated with development of an obvious cauda equina syndrome. Nocardia asteroides was recovered from purulent cerebrospinal fluid and a diagnosis of primary nocardial meningitis was made. No extrameningeal focus could be identified and there was no evidence of previous pulmonary involvement. In vitro susceptibility studies demonstrated resistance to sulphonamides, but subsequent treatment with fusidin, amikacin, rifampicin and imipenem also proved ineffective, despite in vitro sensitivity to these agents. The site of infection, absence of an extrameningeal focus, myelographic appearances and antimicrobial sensitivities make this a unique case.

A 53 year old woman with previous good health was admitted to hospital in March 1985 with a short history of malaise, ankle swelling, oral ulceration and polyarthralgia. Shortly before admission she also noted a nasal discharge, sore red eyes, an erythematous rash over her ankles, and decreased urine volume. On examination she was apyrexial and pale. A resolving episcleritis was noted and vasculitic skin lesions were present over the lower legs and feet. The pulse was 80 beats per minute, blood pressure 130/70 mmHg and a soft systolic murmur was present. The chest was clear and abdominal examination unremarkable. An area of decreased sensation to pinprick and light touch was present over the dorsum of the left foot. Initial laboratory findings revealed a haemoglobin of 10.1 g/dl, white blood cell count of 14,500/mm³ with 88% neutrophils, and platelet count of 999,000/mm³. The serum urea was 28.2 mmol/l and creatinine 980 µmol/l. A skin biopsy revealed leucocytoclastic vasculitis and renal biopsy showed focal and segmental glomerulonephritis with fresh crescents in four of the 13 glomeruli present. A diagnosis of rapidly progressive glomerulonephritis, secondary to a systemic vasculitis within the spectrum of polyarteritis, was made. Peritoneal dialysis was commenced and the patient was treated with prednisolone 60 mg daily, cyclophosphamide 2 mg/kg daily and plasma exchange (five 4 litre exchanges for plasma protein fraction). Her clinical condition rapidly improved and renal function returned to normal. She was discharged on cyclophosphamide (later converted to azathioprine) and reducing doses of steroids, and remained well until August 1985.

She was readmitted with severe lancinating pain in both legs and an increased sensory deficit over the left lower leg. She had weakness of ankle and toe dorsiflexion and plantarflexion on the right and absent ankle jerks bilaterally. Cutaneous sensation was decreased over the dorsum of the left foot, the lateral side of the left lower leg, and the dorsum of the right halluc. Vibration sense was absent below the knees, and proprioception defective at the toes. Sensory nerve conduction studies in the right median and ulnar nerves were normal. The right median motor nerve conduction velocity (MNCV) was slightly reduced at 46 m/s, with a distal motor latency of 2.2 ms and evoked muscle action potentials (MAPs) of only 2.0 and 1.5 mV stimulating at the wrist and elbow. The left sural sensory nerve action potential was absent. Evoked MAPs stimulating the peroneal nerves were reduced in amplitude at 35 (right) and 350 (left) µV, with a MNCV on the left of 35 m/s. A diagnosis of a multifocal peripheral neuropathy secondary to vasculitis was made, and she was treated with increased doses of steroids and reintroduction of cyclophosphamide.

Three days later her condition deteriorated; she developed a moderately severe paraparesis and there was loss of all sensory modalities below T8. The tendon reflexes were absent in the lower limbs, and urinary incontinence and a lax anal sphincter were noted. A histamine flare test showed absent flares in affected dermatomes, suggesting that the site of the lesion was distal to the dorsal root ganglia. A myelogram was normal; the cerebrospinal fluid (CSF) white cell count was 104/mm³ and glucose concentration was 2.7 mmol/l with a blood glucose of 3.7 mmol/l. Plasma exchange was reintroduced to the treatment regimen without effect.

One week later the CSF white cell count had risen to 1675/mm³ and the glucose concentration was 1.1 mmol/l. Intrathecal infection was strongly suspected, but no organism was seen on gram stain or cultured. After a further 5 days, despite having commenced broad spectrum antibiotics, she became pyrexial, experienced rigors and developed neck stiffness. A myelogram was performed via cervical puncture which revealed poor filling of the cauda equina with multiple intradural extramedullary filling defects (fig. a). This appearance, together with the cerebrospinal fluid findings (lumbar: white cell count 3000/mm³, glucose undetectable; cervical: white cell count 204/mm³, glucose 3.7 mmol/l) suggested a focus of infection in the lumbar region. Under radiographic screening, pus was aspirated from the T12/L1 space. Gram stain revealed gram positive branching filamentous organisms, which were identified as N. aster-
Nocardiosis of the central nervous system is usually manifest by cerebral abscesses, which may eventually rupture into the ventricles or subarachnoid space. Meningitis commonly follows this event, but meningitis without abscess is rare. To our knowledge, only two cases of primary N. asteroides meningitis have been previously reported. In one instance direct inoculation from injuries sustained in a road accident was suspected. The portal of entry in our patient was not identified and no evidence for a primary pulmonary focus was detected during the patient’s illness, or at necropsy. At necropsy, the only relevant findings were of finely scarred kidneys and nocardiosis involving the spinal cord and meninges; no other focus of infection was apparent. In no previous reports have the myelographic appearances of spinal nocardiosis been presented. However, similar findings have occurred with aspergillumia complicating chronic lymphatic leukemia. Of further interest is the unusual finding of in vitro resistance to sulfonamides demonstrated in this case. Nocardiosis is not exclusive to immunocompromised individuals, although this is commonly a factor. Our case emphasizes the difficulty in distinguishing between underlying vasculitic disease activity and opportunistic infection in immunosuppressed individuals. The aggressive immunotherapy used in this case has been found to be necessary in patients with systemic vasculitis of the Wegener’s granulomatosis/microscopic polyarteritis group. This is particularly so in the context of oliguric rapidly progressive glomerulonephritis, but infective complications are to be expected. Diagnostic difficulties are compounded by the observation that infection may also increase the activity of the vasculitic disorder, so that the demonstration of an infection does not necessarily imply that it is responsible for any clinical deterioration. This case demonstrates an unusual presentation of nocardiosis, and illustrates the difficulties inherent in the diagnosis and management of this infection.

We thank Dr AE Harding for neurological assessments and the electrophysiological studies. We are grateful to Dr J Olney for the radiographs, Professor DJ Evans for the interpretation of the histology and Dr N Cohen for advice on anti-microbial therapy.

Stephen J Brecker
Charles D Pusey
Department of Medicine
Royal Postgraduate Medical School
Hammersmith Hospital
Ducane Road
London W12 0HS, UK

References

Motor entrapment neuropathies in the elderly

Sir: To our knowledge the frequency of entrapment neuropathies in the general population is unknown. From 1 June 1984 to 31 July 1985 we performed a survey on the neurological condition in the elderly in the Republic of San Marino, which is the smallest independent State in the world, located near the Adriatic Coast, within Italy. All people aged 67, 72, 77, 82 and 87 living in the Republic were invited to undergo a standardised neurological examination. Three hundred and ninety eight agreed to be examined out of a total sample of 498 but full examination was only possible in 396. People were not specifically asked about sensation disturbances or entrapment neuropathies. However, muscle wasting and loss of muscle strength were recorded during examination. Whenever entrapment neuropathy was suspected patients were further asked about their symptoms and a specific sensation examination was done. Diagnosis of entrapment neuropathy was based on neurological examination showing typical motor and sensory deficits. Whenever possible nerve conduction velocity (NCV) study was performed.

Among 396 people we found 7 (2%) with an entrapment neuropathy. Details of the patients are shown in the table.

In two patients NCV study was not done, but in these the typical signs and symptoms enabled us to establish a diagnosis of entrapment neuropathy. The design of our study was such that only subjects with motor signs of entrapment neuropathy (with or without sensory disturbances) were considered abnormal. Therefore our figure may be considered indicative of the frequency of motor entrapment neuropathies in a general population older than 65 years.

Five patients had not previously sought neurological consultation. Their symptoms had been generically attributed to "arthritis". This shows that entrapment neuropathies although not very frequent in the elderly, may be easily overlooked.

R D'ALESSANDRO
G BENASSI
P MARTINELLI
Clinica Neurologica, University of Bologna, Via U Foscolo 7 40123 Bologna, Italy.

Accepted 22nd September 1987.

Table Features of motor entrapment neuropathies found among 396 people aged over 65 years

<table>
<thead>
<tr>
<th>Age (yr)</th>
<th>Sex</th>
<th>Nerve involved</th>
<th>Site of entrapment</th>
<th>Sensation deficit</th>
<th>NCV</th>
<th>Underlying factor</th>
</tr>
</thead>
<tbody>
<tr>
<td>67</td>
<td>♂</td>
<td>Right ulnar</td>
<td>Elbow</td>
<td>Yes</td>
<td>No</td>
<td>Elbow fracture aged 45 years</td>
</tr>
<tr>
<td>67</td>
<td>♂</td>
<td>Right ulnar</td>
<td>Wrist</td>
<td>Doubt</td>
<td>Yes</td>
<td>Articular blockade</td>
</tr>
<tr>
<td>72</td>
<td>♀</td>
<td>Left median</td>
<td>Wrist</td>
<td>Yes</td>
<td>No</td>
<td>Mild arthritis</td>
</tr>
<tr>
<td>77</td>
<td>♀</td>
<td>Peroneal</td>
<td>Caputulum fibulae</td>
<td>Yes</td>
<td>Yes</td>
<td>Moderate arthrosis</td>
</tr>
<tr>
<td>77</td>
<td>♂</td>
<td>Bilateral median</td>
<td>Wrist</td>
<td>Yes</td>
<td>Yes</td>
<td>Mild arthrosis</td>
</tr>
<tr>
<td>77</td>
<td>♂</td>
<td>Right median</td>
<td>Wrist</td>
<td>Yes</td>
<td>Yes</td>
<td>Moderate arthrosis</td>
</tr>
<tr>
<td>77</td>
<td>♂</td>
<td>Right ulnar</td>
<td>Elbow</td>
<td>Yes</td>
<td>Yes</td>
<td>Moderate arthrosis</td>
</tr>
</tbody>
</table>

NCV, nerve conduction velocity.

Relapsing dermatomyositis associated with sarcoidosis

Sir: Two cases of dermatomyositis in association with sarcoidosis have been described, both in Japanese patients. We report a further case in an Anglo-Japanese man, his illness running a relapsing course over a period of 6 years.

In October 1979 a 56 year old Anglo-Japanese accountant presented with a 6 month history of pain, stiffness and paraesthesiae in both hands, the symptoms tending to be worse at night but also aggravated by heavy work. He also had swelling of the knuckles of both hands, and an irritating skin rash. He had lost 8 kg in weight. There was a past history of duodenal ulcer.

On examination he was a thin man (weight 57.5 kg). He had an erythematous scaling rash of his lower legs, arms, forehead and back, periorbital oedema, and bilateral axillary lymphadenopathy. Examination of his chest, heart and abdomen was normal. There was no muscle weakness or wasting, though he did have some muscle tenderness. Reflexes were normal and sensation was intact. Full blood count, bone marrow aspirate and trephine biopsy, barium enema, barium meal and intravenous pyelogram were normal. RA latex and ANA were negative. Erythrocyte sedimentation rate was 17 mm in 1 hour. Creatine kinase was elevated at 384 IU/l (normal less than 240 IU/l). A chest radiograph showed bilateral apical pleural thickening with a localised calcification suggestive of previous pulmonary tuberculosis. There was also some interstitial shadowing at both bases, though this resolved after 1 month. Pulmonary function testing indicated a minor restrictive impairment and reduced carbon monoxide diffusion. Lymph node biopsy showed benign reactive changes with follicular hyperplasia, sinus histiocytes and numerous aggregations of histiocytes in the interfollicular cortex and medulla.

A diagnosis of dermatomyositis was made. His symptoms fluctuated over the next 5 years. The CK was persistently mildly elevated until the most recent relapse. From 1980–1982 he had only mild rash and muscle stiffness. In early 1982 he was given a 3 week course of steroids with resolution of his symptoms. Six months later he had a recurrence of skin rash and developed paraesthesiae which resolved without treatment in 2 months.

In November 1983 he complained of a 2 month history of general malaise and muscle tenderness and was found to have mild proximal weakness (MRC grade 4–5). This resolved without treatment over the next 3 years.