Unusual presentation of more common disease/injury

Pins and needles and unilateral foot drop: a presentation of sarcoidosis

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Summary

We describe a case of sarcoidosis in a 57-year-old man who presented with neurological symptoms of pins and needles in both of his hands, left leg weakness and left foot drop. Neurophysiological examination revealed asymmetric motor and sensory polyneuropathy. Common peroneal nerve involvement accounted for the left foot drop. Thoracic CT scan revealed bilateral hilar and mediastinal lymphadenopathy. He had hypercalcaemia and raised serum ACE level. Histological examination of a mediastinal lymph node showed non-caseating epithelioid cell granulomas consistent with the diagnosis of sarcoidosis. There was no evidence of acid-fast bacilli or fungi on special stains. This case highlights the importance of considering sarcoidosis as a potential diagnosis in patients presenting with peripheral neuropathy. Although response to corticosteroids and immunosuppressive therapy may be seen, in our case the patient's neurological deficit remained persistent despite treatment.

BACKGROUND

Sarcoidosis is a multisystem inflammatory disorder characterised by granuloma formation. Its prevalence is 10–20/100 000 and exact aetiology is yet to be established despite extensive research efforts.^{1 2} It presents between 10 and 40 years of age in the majority of cases and lungs are the most frequently affected organs accounting for 90% of cases.³ Neurological involvement is rare and is reported to be less than 5% in a study of 736 patients.³

CASE PRESENTATION

A 57-year-old man presented with lethargy and a 3 stones weight loss over a period of 7 weeks. He had noticed over a period of few months to have pins and needles and pain affecting digits 5, 4 and 3 and contiguous area of the palm of both hands and difficulty in holding objects such as cutlery. He also noted weakness of his left leg. On examination, the patient had wasting of interossei (figure 1) and the left foot drop. There were no abdominal or cardiac symptoms. He denied any joint pains or skin rash. He had no exposure to tuberculosis. He was an ex-smoker with a 2-pack-year history.

Thoracic CT scan showed multiple mediastinal nodes as well as bilateral hilar nodes (figure 2). An echocardiogram and 24 h ECG were normal. His initial biochemical profile revealed hypercalcaemia with serum calcium 3.5 mmol/l, which was treated with rehydration and intravenous bisphosphonate. Serum ACE level was raised at 109 (normal range 8–52) U/l. He underwent a mediastinoscopy and lymph node biopsy. Histology showed non-caseating epithelioid cell granulomata consistent with sarcoidosis. Special stains showed no evidence of acid-fast bacilli or fungal elements. Prednisolone 40 mg daily was started. The patient developed uveitis so topical corticosteroid eye drops were introduced with a significant

improvement in his vision. MRI brain and spine showed no evidence of sarcoid involvement. Neurophysiological examination showed a motor and sensory polyneuropathy, asymmetric in nature, mixed demyelinating and axonal degeneration, with severe left ulnar mononeuropathy and milder right ulnar mononeuropathy. The diagnosis of neurosarcoidosis (NS) was made affecting peripheral nervous system, a polyneuropathy and a mononeuropathy multiplex. The patient was treated with gabapentin, azathioprine and prednisolone as well as regular physiotherapy. These measures resulted in stabilisation but no significant improvement of his neurological deficit.

DISCUSSION

NS may present with multiple neurological manifestations in patients known to have sarcoidosis or presenting de novo at the time of initial diagnosis (approximately 50% of patients). The estimated prevalence of NS is 1/100 000 in a UK study.⁴ It may involve any part of central or peripheral nervous system (figure 3). It may pose a diagnostic challenge, particularly when presented as first episode of neurological dysfunction without prior diagnosis of sarcoidosis. If sarcoidosis is suspected as a cause of neurological symptoms, extraneural tissues such as lungs, skin or lymph nodes should be evaluated for histological confirmation, given the difficulties obtaining nervous tissue for biopsy. Neuroimaging in the form of MRI may help elucidate the extent of disease and exclude other common conditions, but it is not able to confirm the diagnosis with absolute certainty and there are no pathognomonic features. Cerebrospinal fluid examination may reveal reduced glucose, mononuclear pleocytosis, oligoclonal bands and occasional elevation of ACE concentration. Nerve conduction studies are useful in the evaluation of

Figure 1 Wasting of interossei at presentation. (A) Dorsal aspect, (B) ventral aspect.

peripheral nerve dysfunction and help characterise the neuropathy more precisely. It is important to recognise that no neurological investigation is diagnostic of NS and the diagnosis is predominantly clinical and

Figure 2. Thoracic CT scan demonstrating bilateral hilar lymphadenopathy.

supported by appropriate investigations after other causes of nervous system dysfunction have been excluded.

There is a lack of high-quality randomised clinical trials of treatment in NS and corticosteroids are the mainstay of management strategy in this condition. A number of immunosuppressive agents, including azathioprine, cyclosporine, mycophenolate, methotrexate, cyclophosphamide and infliximab have been used in NS with significant benefit.^{5–9} Our patient failed to respond to a combination of prednisolone and azathioprine. In case of NS refractory to corticosteroids and immunosuppressive therapy, cranial or spinal irradiation may be considered, particularly in acute life-threatening situations.¹⁰ A ventriculo-peritoneal shunt may be indicated in acute hydrocephalus with continuation of immunosuppressive therapy.

The prognosis of NS is variable. The patients with visual disturbance secondary to optic neuropathy generally have a poor prognosis as demonstrated by a study of 54 patients with NS.¹¹ Approximately 50% of patients lost vision in at least one eye in this series. Meningeal and parenchymal brain lesions tend to have a prolonged course. Furthermore, as described in the case presented, peripheral neuropathy and myopathy runs a chronic course with occasional remissions.

Learning points

- ▶ Peripheral neuropathy may be a presenting feature of sarcoidosis.
- ▶ Sarcoidosis is a multisystem inflammatory disorder and clinicians should be aware of a wide range of clinical presentations.
- ▶ There are a number of management options for neurosarcoidosis (NS) and there is a variable response to corticosteroids and immunosuppression.

Figure 3 Classification of neurosarcoidosis (NS) according to central and peripheral nervous system involvement and associated clinical presentations.

Competing interests None.

Patient consent Obtained.

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Fahim A, Kastelik JA, Morjaria JB, Hart SP. Pins and needles and unilateral foot drop: a presentation of sarcoidosis. *BMJ Case Reports* 2012;10.1136/bcr-2012-007395, Published XXX

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