Rare disease

Osseous spinal sarcoidosis: an unusual but important entity to remember

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Summary

Sarcoidosis is a multi-system disorder of unknown aetiology characterised by non-caseating granulomatous inflammation with varying presentation and prognosis. Bone involvement is uncommon and vertebral involvement is rare. We report a case of vertebral osseous sarcoidosis which presented with pulmonary symptoms mimicking tuberculosis and later developed vertebral involvement despite administration of oral corticosteroids.

BACKGROUND

Sarcoidosis of the vertebral bones is rare, with nearly all publications being case reports. Most patients with vertebral body involvement have a known diagnosis of intrathoracic sarcoidosis. It is a difficult diagnosis to establish without biopsy as other diseases such as malignancy and granulomatous infections such as tuberculosis must be excluded before the diagnosis of vertebral body sarcoidosis can be made.

CASE PRESENTATION

A 41-year-old Pakistani male presented to a general physician with 9-month history of weight loss along with fatigue and a 2-month history of low-grade fever. He was found to have an elevated erythrocyte sedimentation rate (ESR) of 114 mm and an abnormal chest x-ray. Empirical treatment with four first-line anti-tuberculous drugs in combination had been commenced in the community 2 months previously. After taking the medication for 2 months, with some initial improvement, his condition started to deteriorate and he presented to the emergency room at our hospital.

On further inquiry he gave a 2-month history of dry cough, low-grade fever, malaise and a 7 kg weight loss. Physical examination revealed a young cachectic looking man with no respiratory distress. He had grade III digital clubbing and his chest was clear on auscultation.

INVESTIGATIONS

Initial laboratory investigations showed an ESR of 96 mm (normal: 0–20), alkaline phosphatase of 147 IU/l (normal: 28–124) and a γ -glutamyl transferase of 162 IU/l (normal: 3–50). The serum angiotensin converting enzyme level was normal. The tuberculin skin test was <5 mm. Chest radiograph showed bilateral hilar and right paratracheal adenopathy and right upper lung zone fibrocavitary infiltrate. CT of the chest confirmed the adenopathy and alveolar pulmonary infiltration. A transbronchial lung biopsy revealed chronic non-caseating granulomatous inflammation (figure 1). Special stains for fungi and acid fast bacilli (AFB)

were negative. Baseline spirometry revealed moderate restriction (forced vital capacity 2.15 litres, 53.6% predicted). A diagnosis of pulmonary sarcoidosis was made and the patient was commenced on oral prednisolone 1 mg/kg/day. Anti-tuberculous therapy was discontinued after AFB cultures were negative at 6 weeks.

On a follow-up visit at 8 weeks, the patient reported progressive resolution of symptoms along with radiological and spirometric improvement. Oral steroids were gradually reduced to 30 mg/day over an 8-week period. Soon thereafter, he started to complain of numbness in both lower limbs. This was initially thought to be isoniazid-induced neuropathy. However, neurological examination revealed atrophy of the muscles of the lower extremities with more proximal than distal weakness. Proximal strength was graded as 3/5 compared to distal which was 4/5 according to the Medical Research Council scale. Upper extremities did not show any atrophy or weakness. The deep tendon reflexes were symmetrically decreased in all extremities. The sensory examination was unremarkable. Electro-diagnostic studies revealed bilateral denervation of muscles supplied by L₂, L₃ and L₄ roots without any involvement of sensory nerves, consistent with the diagnosis of a polyradiculopathy. There was no evidence of neuropathy or myopathy. MRI of the spine showed disseminated bony involvement with multiple contrast enhancing areas of abnormal signal intensity involving the whole lumbar vertebral column, that is, L_{1–5}, along with evidence of minimal paravertebral and epidural collection (figures 2 A–C). Sarcoidosis involving the spine was suspected, and a biopsy of L₃ vertebra showed non-caseating chronic granulomatous inflammation (figure 3).

DIFFERENTIAL DIAGNOSIS

Metastatic disease, osteomyelitis, Pott's disease.

TREATMENT

Corticosteroids are the cornerstone of conservative management and have been shown to favourably effect the clinical, radiological and histological features of sarcoidosis.

Figure 1 Transbronchial lung biopsy revealing chronic non-caseating granulomatous inflammation.

Figure 2 (A) Sagittal T1 weighted image of the lumbar spine showing abnormal hypo-intense signals in L₁, L₂, L₃ and L₅. (B) Sagittal T2 weighted image of the lumbar spine showing abnormal heterogeneous high signals in L₁, L₂, L₃ and L₅. (C) Postcontrast sagittal T1 weighted image of the lumbar spine showing enhancement in L₁, L₂, L₃ and L₅.

Reasons for treatment failure with steroids include irreversible fibrotic disease, non-compliance, inadequate dosage and/or intrinsic corticosteroid resistance. Medication alternatives to corticosteroids are considered when a patient

requires systemic treatment for sarcoidosis but has¹ intolerable side effects to corticosteroids,² disease progression despite an adequate trial of steroids (equivalent to prednisone 15 mg/day for at least 3 months) or³ disease

Figure 3 Biopsy of L₃ vertebra showing non-caseating chronic granulomatous inflammation.

persistence requiring long-term, high-dose corticosteroid therapy (prednisone ≥ 15 mg/day).^{4 5} Corticosteroid therapy improves neurological outlook in most patients, although a small minority require surgical intervention for progressive vertebral destruction and accompanying spinal instability in an effort to halt the neurological deterioration. A spinal reconstructive approach is usually adopted in accordance with the extent of the disease process and the accompanying clinical compromise.⁶

Clinical course is highly variable in sarcoidosis. It can range from spontaneous remission to chronic progressive disease in some patients. Fatalities occur in 1% to 4% of cases. Owing to the high rate of progression and recurrence following treatment, imaging follow-up is recommended in all patients.⁷

OUTCOME AND FOLLOW-UP

The maintenance dose of prednisolone was increased to 60 mg/daily and oral azathioprine of 50 mg/day was added. This resulted in rapid resolution of symptoms and a significant clinical improvement. At present, the patient is on oral combination therapy of prednisolone 10 mg/day and azathioprine 150 mg/day, with no signs of relapse.

DISCUSSION

Our case demonstrates the systemic nature of sarcoidosis with involvement of the nerve roots and osseous system. The use of imaging modalities and histological investigation of tissue contributed towards a successful diagnosis. In the absence of any spinal instability, neurological symptoms associated with vertebral sarcoidosis respond satisfactorily to non-surgical treatment with steroids and other

immunosuppressives.⁸ We believe that early identification and treatment of the disease in our patient avoided the need for surgical intervention. This highlights the importance of medical vigilance and recognition of typical radiological features during evaluation of chronic granulomatous diseases to facilitate the diagnosis and halt systemic spread and avoid serious sequelae in case of neurological and osseous involvement.

Less than 10 similar cases have been reported. The development of vertebral sarcoid despite oral corticosteroids was an unusual presentation in our patient.

Learning points

- ▶ Vertebral sarcoidosis is a rare condition that can present with lytic or blastic osseous lesions which on bone scan or MRI are indistinguishable from metastatic cancer.
- ▶ It usually occurs at the time of initial diagnosis of sarcoidosis but very rarely can appear many years after presumed resolution of thoracic sarcoidosis.
- ▶ Biopsy is needed for confirmation.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Lynch JP 3rd**, Sharma OP, Baughman RP. Extrapulmonary sarcoidosis. *Semin Respir Infect* 1998;**13**:229–54.
2. **Tzagarakis GP**, Papagelopoulos PJ, Sapkas GS, *et al*. Surgical management for instability and paraplegia caused by spinal sarcoidosis. A case report. *Spine* 1998;**23**:1711–4.

3. **Valencia MP**, Deaver PM, Mammarrappallil MC. Sarcoidosis of the thoracic and lumbar vertebrae, mimicking metastasis or multifocal osteomyelitis by MRI: case report. *Clin Imaging* 2009;**33**:478–81.
4. **Morgan SS**, Aslam MB, Mukkanna KS, *et al*. A rare presentation of sarcoidosis, back pain and spondylolisthesis. *J Bone Joint Surg Br* 2008;**90**:240–2.
5. **Barnard J**, Newman LS. Sarcoidosis: immunology, rheumatic involvement, and therapeutics. *Curr Opin Rheumatol* 2001;**13**:84–91.
6. **Brembilla C**, Signorelli A, Lamartina C, *et al*. Surgical management in spinal sarcoidosis. *Spine* 2009;**34**:E258–61.
7. **Lury KM**, Smith JK, Matheus MG, *et al*. Neurosarcoidosis review of imaging findings. *Semin Roentgenol* 2004;**39**:495–504.
8. **Ito M**, Motomiya M, Abumi K, *et al*. Vertebral osteonecrosis associated with sarcoidosis. Case report. *J Neuro Surg Spine* 2005;**2**:222–5.

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