Vertebral osteonecrosis associated with sarcoidosis

Case report

MANABU ITO, M.D., MAKOTO MOTOMIYA, M.D., KUNIYOSHI ABUMI, M.D., OSAMU SHIRADO, M.D., YOSHIHISA KOTANI, M.D., KEN KADOYA, M.D., EIHIRO MUROTA, M.D., AND AKIO MINAMI, M.D.

Department of Orthopaedic Surgery, Hokkaido University Graduate School of Medicine, Sapporo, Japan

Sarcoidosis is a systemic disease commonly affecting lung, skin, or eye. Sarcoidosis involved with osseous structures occurs in approximately 5% of patients, usually involving small bones. Spinal sarcoidosis is extremely rare. The authors report on a man in whom examination of a subclavicular lymph node biopsy specimen and its spinal involvement had established a diagnosis of sarcoidosis and who had undergone steroid therapy. Despite intensive conservative treatment, the authors observed progressive collapse of L-2 requiring spinal decompressive and reconstructive surgeries. Histological evaluation of the collapsed vertebra did not show the typical noncaseating granuloma; rather, the authors observed osteonecrosis of the entire L-2 structure without reactive cellular activities. Other potential diagnoses including infectious disease, metastatic spinal tumor, and osteoporotic vertebral collapse were excluded based on laboratory data, imaging studies, and pathological findings. Complete necrosis of the entire L-2 vertebra in this case can be considered as a rare clinical manifestation of spinal sarcoidosis. Because of osteopenia and systemic bone fragility, combined anterior–posterior spinal reconstructive surgery was performed to restabilize the severely damaged spine.

KEY WORDS • sarcoidosis • vertebra • osteonecrosis • spinal surgery • spinal instrumentation

Abbreviations used in this paper: CT = computerized tomography; MR = magnetic resonance; VB = vertebral body.
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Laboratory workup indicated a white blood cell count of 13,600/cm³ and erythrocyte sedimentation rates of 38 mm/1 hour and 75 mm/2 hours. The C-reactive protein level was 10.71 mg/dl and alkaline–phosphatase 605 IU. Tumor markers were normal. A skin test for tuberculosis was negative. All cultures from sputum, urine, and pharynx were negative. Spinal MR imaging revealed intravertebral cleft with low signal intensity on T1-weighted sequences, high intensity on T2-weighted sequences, and a soft-tissue mass around the L-2 VB (Fig. 2). The spinal canal was occluded by protrusion of the L-2 VB’s posterior wall. Axial CT scanning revealed destruction of the entire L-2 vertebra, including VB, pedicle, and facet joints (Fig. 3). Gallium scintigraphy demonstrated a cold spot at L-2. The bone mineral density was 0.724 g/cm² at the L-4 VB.

Operation. The patient underwent anterior decompression than involved an L-2 corpectomy and anterior L1–3 spinal fusion that involved placement of an autologous fibula graft and Kaneda-SR instrumentation (Depuy AcroMed, Johnson and Johnson Corp., Cleveland, OH). Two weeks after the first operation, plain radiography revealed sinking of the grafted fibula. In light of this, we performed posterior T8–L4 spinal fusion in which a bone graft and ISOLA hardware were placed (Depuy AcroMed, Johnson and Johnson Corp.). At the latest follow-up, 16 months after the second surgery, the patient’s neurological status had normalized and there was no evidence of instrumentation failure (Fig. 4).

Histological Findings. Histological and cytological examinations of the resected L-2 VB demonstrated massive bone necrosis, and the bone marrow was replaced by ghost cells (Fig. 5). There were no normal osteocytes, osteoblasts, or osteoclasts in the resected VB. Additionally no reactive new bone formation was demonstrated in the resected VB. No organisms could be detected in both smear and culture of the tissue. Polymerase chain reaction for tuberculosis was also negative.

Discussion

Spinal involvement of sarcoidosis is extremely rare. We found only 33 cases reported in the literature.1–6,8–10,13–14,18–20,22,24,26–30 Most of the patients with spinal involvement suffer from back pain and only a small number of them experience neurological deficits.1,4,18,28,30 To date, only seven reports have been published in the English-language literature. These patients underwent surgery for spinal lesions due to sarcoidosis.1,4,18,22,27,28,30 According to the authors, steroid therapy was effective in most cases involving spinal sarcoidosis;7,9,24,27 however, symptoms in our patient did not respond to steroid therapy, and severe collapse of the L-2 vertebra eventually occurred and the patient suffered neurological deficits.

We have questioned the diagnosis of the spinal lesion in this patient. Other possible diagnoses included the following: 1) infectious diseases; 2) metastatic spinal tumors, and
3) osteoporotic vertebral collapse. Pyogenic spondylitis or spinal tuberculosis was excluded based on the fact that any organisms could not have been demonstrated by the samples of the affected vertebra. Moreover, the fact that tumor markers had been negative and tumor cells had not been identified did not support a diagnosis of primary or metastatic spinal tumor. Osteoporotic vertebral collapse was excluded based on the following. 1) The shape of the cleft was much narrower than that of an ordinary osteoporotic intravertebral cleft. 2) Destructive changes in this patient were present in the VB and also the posterior elements, although osteoporotic collapse usually affects only the VB, not the posterior elements. 3) There was no evidence of reactive new bone formation in the entire collapsed VB, although osteoporotic fractures are accompanied by a fracture healing process including new bone formation, osteoblasts, and osteoclasts.

A typical pathological characteristic of sarcoidosis is noncaseating granuloma. It has been reported that definite diagnosis of a spinal lesion associated with sarcoidosis can only be affirmed by examination of biopsy samples of the lesions. In our search of the literature, however, we found several exceptional cases involving atypical histological findings. Sundaram, et al., reported the atypical case of a patient with spine-involving sarcoidosis. They found noncaseating granuloma accompanied by central necrosis. Additionally, Ozseker, et al., and Rohatgi and Schwab reported that pulmonary lesions of sarcoidosis were occasionally associated with cavities caused by central ischemic necrosis. Other authors have reported that liver or heart lesions due to sarcoidosis sometimes were not concordant with a typical noncaseating granuloma. Sarcoidosis does not always exhibit a simple clinical picture; commonly there are greatly varying clinical appearances. In addition, sarcoidosis exhibits various different patterns on radiographs, CT scans, or MR images. For example, radiography and CT scanning in cases of spinal sarcoidosis reveal lytic, sclerotic, or mixed findings. There are no definite findings of spinal sarcoidosis on MR imaging. Steroid therapy has been reported to improve the physical findings and suppress inflammation and granuloma formation in spinal lesions of sarcoidosis. In our case, however, steroid therapy was not effective and vertebral collapse became progressive. Surgical treatment is mandatory in the presence of progressive neurological deterioration and severe spinal destructive changes.

Steroid therapy has been reported to improve the physical findings and suppress inflammation and granuloma formation in spinal lesions of sarcoidosis. In our case, however, steroid therapy was not effective and vertebral collapse became progressive. Surgical treatment is mandatory in the presence of progressive neurological deterioration and severe spinal destructive changes. In most patients with sarcoidosis, osteopenia and systemic bone fragility are present. Because we observed severe destruction of both L-2 anterior and posterior elements as well as severe bone fragility, combined anterior–posterior spinal reconstruction was needed to restabilize the damaged spine.

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Address reprint requests to: Manabu Ito, M.D., Kita 15 Jo Ni-
shi 7 Chome, Kita-ku, Sapporo 060-8638 Japan. email: maito@med.
hokudai.ac.jp.