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CASE REPORT

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Gorham's disease of the spine

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Abstract Massive osteolysis is a rare condition and is very uncommon in the spine. The MRI appearance of Gorham's disease of the spine has not previously been reported. We present here a case of this condition with imaging details.

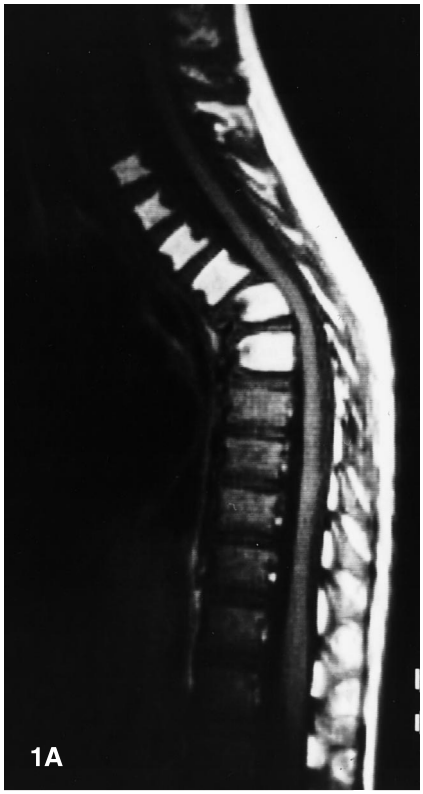
Key words Gorham's disease · Spine · Magnetic resonance imaging

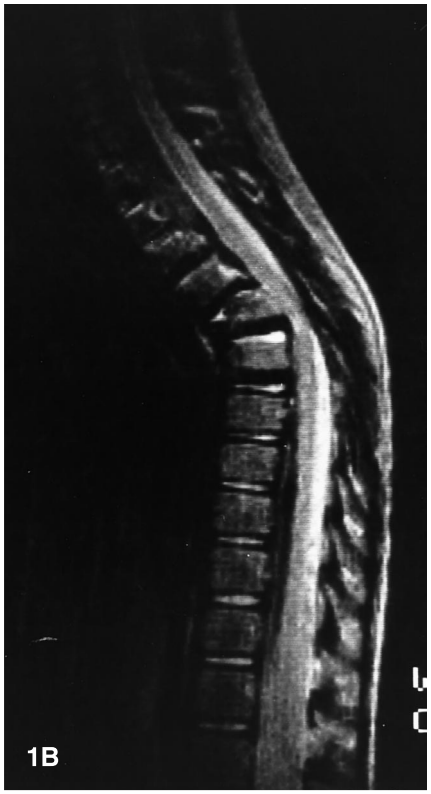
Case report

A 5 ½-year-old male child of Asian origin presented to his local hospital with back pain following a minor fall. On examination there was some spinal tenderness but there were no neurological abnormalities in the limbs. A radiograph of the thoracic spine demonstrated a mid-thoracic kyphosis with a bone defect. All haematological investigations were within normal limits, and a bone scan did not show any abnormal activity. Magnetic resonance imaging (MRI) at presentation demonstrated extensive replacement of the T2–T7 vertebral body bone marrow by tissue of high signal on T1-weighted spin echo (WSE) scans (Fig. 1A). There was no appreciable signal change on T2-WSE scans (Fig. 1B). A biopsy of the spine was performed. Successive scans over a 3-year period demonstrated progressive kyphosis and cord compression with disappearance of T6 and T7 and extension of the high signal into T8, T9 and T10 (Figs. 2, 3). At no stage was a soft tissue mass demonstrated.

Macroscopically, the biopsy showed a lymphangiomatous process eroding the vertebral body and the neck of the ribs. Microscopically, reactive bone and dense fibrovascular tissue were found; there was no evidence of tumour (Fig. 4). Cultures of the biopsy material were negative. On clinical and radiological grounds a diagnosis of Gorham's disease was made.

The patient rapidly developed a kyphus and signs of cord compression. He underwent anterior spinal vertebrectomy and strut grafting 6 weeks after presentation. Although placed in a brace he was lost to follow-up and returned 4 months later without having used his brace. As a result he had developed an angular kyphus and a recurrence of the signs of neurological compression. An uninstrumented posterior fusion was performed; he was placed in a halo jacket. The disease progressed, reabsorbing the graft and adjacent vertebrae. The halo jacket was retained, and the patient was treated experimentally with pamidronate 5 mg intravenously once a day for 8 days followed by 10 mg intravenously once a month for 12 months. He also received calcitonin at a dose of 25 U/day for 8 days followed by 25 U three times a week for a year. A biphosphonate, calcium carbonate (Calcichew), was also given at a dose of one tablet per day for a year. Although accurate evaluation was impossible, the disease did not obviously progress during this time. He has now remained in his halo jacket for 30 months. The progression of the disease has slowed, but there is no evidence of healing. A further biopsy has been taken to confirm the diagnosis. The neurological symptoms have remained stable. It is intended in the future to carry out a further vertebrectomy.





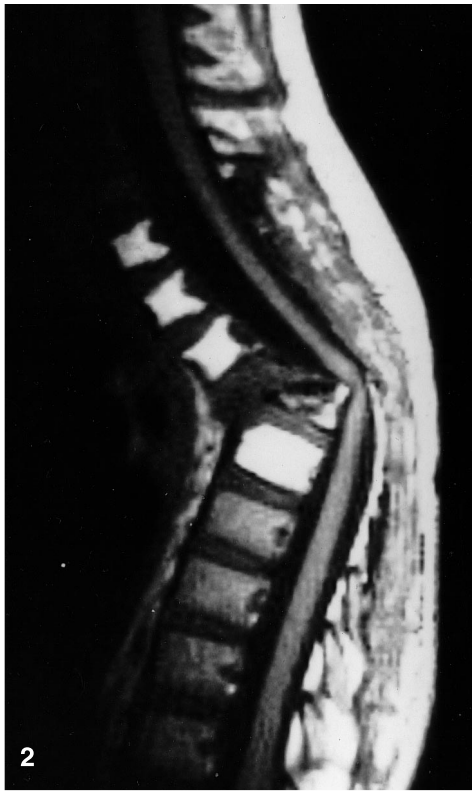
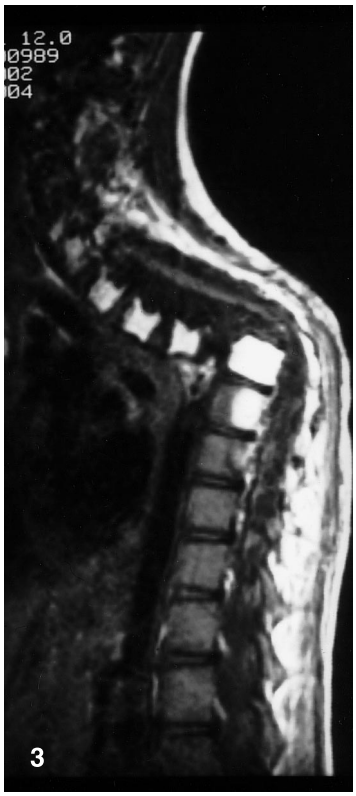


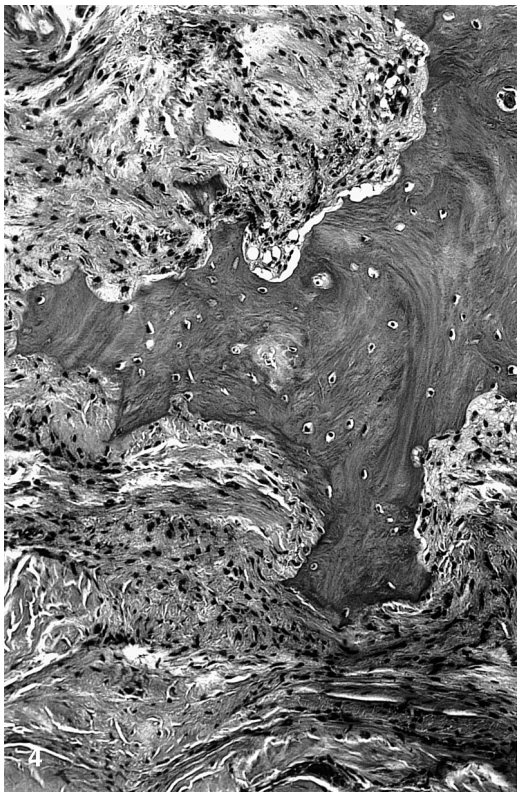
Fig. 1A, B MRI of the thoracic spine at presentation. **A** T1-WSE scan (TR500/TE15) showing replacement of marrow of T2–T7 by tissue of high signal intensity, and a kyphosis centred on T6. **B** T2-WSE scan (TR2500/TE90) showing no appreciable change in signal intensity. Note absence of adjacent soft tissue mass

Fig. 2 T1-WSE scan (TR460/TE15) of the thoracic spine 16 months after presentation. T7 is partially destroyed and there is now involvement of the T8 vertebral body

Fig. 3 T1-WSE scan (TR500/TE20) of the thoracic spine 33 months after presentation. The disease process has spread into T9 and T10 with involvement also of the pedicles and posterior arches

Fig. 4 The pathology specimen removed at operation shows a lymphangiomatous process eroding bone with associated collagen formation and reactive bone formation





Discussion

Massive osteolysis, phantom bone disease or Gorham's disease, was defined as a specific pathological entity by Gorham [1] in 1955. The disease can be subdivided into five syndromes [2] depending upon the sites affected and the inheritance pattern. This patient was type IV disease, the monocentric form with no associated nephropathy and no genetic associations. The mean age of presentation of Gorham's disease is 27 [3]; it is uncommon in young children. Patients usually present with pain following a trivial injury, or with a long history of non-specific pain. The appendicular skeleton is most commonly involved. Only three previous cases of spinal disease have been reported [1].

The radiological features of the disease are well described [1, 4]. There has been only one report of the MRI features of the disease [5]. The case described was a 36-year-old man with massive osteolysis of the scapula. Both T1- and T2-WSE scans showed the scapula to be replaced by tissue of high signal intensity, consistent with the haemangiomatous or lymphangiomatous process identified pathologically. Similar findings were seen in the present case with the vertebral bone marrow being replaced by tissue of very high signal intensity on T1-WSE scans. The T2 scan was not bright in this case; the marrow of children, who are more active, usually has low signal intensity on T2-WSE scan. MRI was capable of demonstrating the progressive nature of the disorder and the lack of an associated soft tissue mass which is characteristic of Gorham's disease.

Confirmation of the diagnosis requires biopsy to exclude tumour, infection and aseptic necrosis [6]. The pathological process is of a locally aggressive angiomatosis. Usually the disease runs a benign course with the osteolytic process stopping after a few years. Excision and bone grafting of the affected areas during active disease usually results in recurrence of the disorder [7]. Radiotherapy has been used to treat Gorham's disease, with variable results [7, 8]. The recommended treatment is excision and endoprosthetic replacement [7]; unfortunately, this is not possible in the spine.

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