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CASE REPORT

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Chronic recurrent multifocal osteomyelitis causing spinal cord compression

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Abstract Chronic recurrent multifocal osteomyelitis (CRMO) is a very rare condition of unknown etiology and most commonly occurs during childhood or adolescence. The purpose of this paper is to present a case of CRMO in a vertebral location with severe kyphosis, spinal cord compression, and neurological dysfunction requiring anterior decompression and fusion. After 12 weeks, the patient was physically able to return to school. At 2-year follow-up, neurological and functional outcomes are fair. Magnetic resonance imaging shows good restoration of the sagittal spine alignment despite residual mild kyphosis, and restoration of a normal sagittal diameter of the spinal canal.

Key words Chronic recurrent multifocal osteomyelitis · Thoracic vertebra · Spinal cord compression

Introduction

To the best of our knowledge, this is the first report of chronic recurrent multifocal osteomyelitis (CRMO) in a thoracic spine location, causing major spine instability with severe thoracic kyphosis associated with anterior cord compression and neurologic deficit.

CRMO is a very uncommon disease of childhood or adolescence, first described by Giedon et al. [10], and today its etiology remains unknown [1, 15]. The clinical course is characterized by an insidious onset of pain, redness, and swelling over the affected bones, with a chronic intermittent progression marked by exacerbations and remissions of the disorder, resembling osteomyelitis [2]. The lesions predominantly affect the clavicles and the metaphysis of the long bones, most often the distal tibia, followed by the distal femur and the proximal tibia [3]; bilateral symmetrical locations are reported to be typical of the disease [10, 11, 12, 19, 20]. In the literature involvement of the vertebral column has been described in only 10 cases [3, 5, 6, 21] (Table 1), and vertebral body lesions are predominantly localized to the thoracic spine, most often with sclerosis without wedging.

Characteristic radiographic features were first described by Probst [17, 18] and are usually consistent with those of osteomyelitis, such as lacunar, osteolytic, or polycystic lesions with a sclerotic margin, periosteal thickening and a localization in metaphyses close to the growth plates. In the experience of most authors, epiphyses are not affected [8, 11, 17]. Microbiological studies eliminate acute bacterial osteomyelitis (blood cultures and culture of biopsies are always sterile), and failure of attempts to isolate any organism is a criterion for establishing the diagnosis of CRMO [3]. To date immunological studies with serologic tests have mainly remained negative and an autoimune disorder is ruled out [3, 4, 13]. Histologically, biopsies reveal nonspecific inflammatory

Table 1 Published reports of CRMO in vertebral locations

Reference	Case no.	Location of vertebral lesions	Vertebra plana	Neurological involvement	Surgery
Bjorksten et al. [2]	1	L3-L5	–	–	–
	2	C6	–	–	
Brown & Wilkinson [5]	1	T4-T5-L1	T4	–	–
	2	T4-T5-T7	T5	–	–
	3	L3	–	–	–
Yu et al. [21]	1	T5-T6-T7	T5	–	–
	2	T4-L1	T4	–	–
	3	T8-L1-L2	L1	–	–
	4	T4-T5-T7	–	–	–
Carr et al. [6]	1	T5	Kyphosis	–	+

changes [3, 14]. In the differential diagnosis, infections (especially hematogenous osteomyelitis) and neoplastic conditions must be considered when evaluating a child who has multiple osseous lesions.

No specific treatment is required for CRMO; the osseous lesions resolve spontaneously with general suportive measures [9, 16]. Antibiotics have no obvious effect on the symptoms or on the clinical course, but intravenous antibiotic treatment at the beginning is appropriate in any case after the biopsy has been performed, until negative microbiological results have been obtained [13]. Nonsteroid anti-inflammatory drugs may be beneficial. Steroids may be of benefit for severe exacerbations [13].

Case report

A 14-year-old girl (who was known to have CRMO at the age of 12 years) was seen at our institution in February 1994 with CRMO in a vertebral localization. Between November 1992 and February 1994, the patient had developed a total of five lesions (proximal right femur, proximal right humerus, right scapula, sternum, left metatarsus). The diagnosis of CRMO, which is one of exclusion, was obtained after biopsy of the femoral location. The vertebral lesion was the sixth location, where disease had completely destroyed the second thoracic vertebra leading to a major kyphosis with anterior cord compression (Fig. 1) and a rapidly progressive neurological deficit, stage C in the Frankel score. At presentation the patient had a 4-week history of upper thoracic pain, and in the week before presentation she had noticed weakness in both lower limbs. She underwent emergency anterior surgical decompression using a sternum-splitting approach as described by Cauchoix and Binet [7], which provided wide and direct exposure to the vertebral

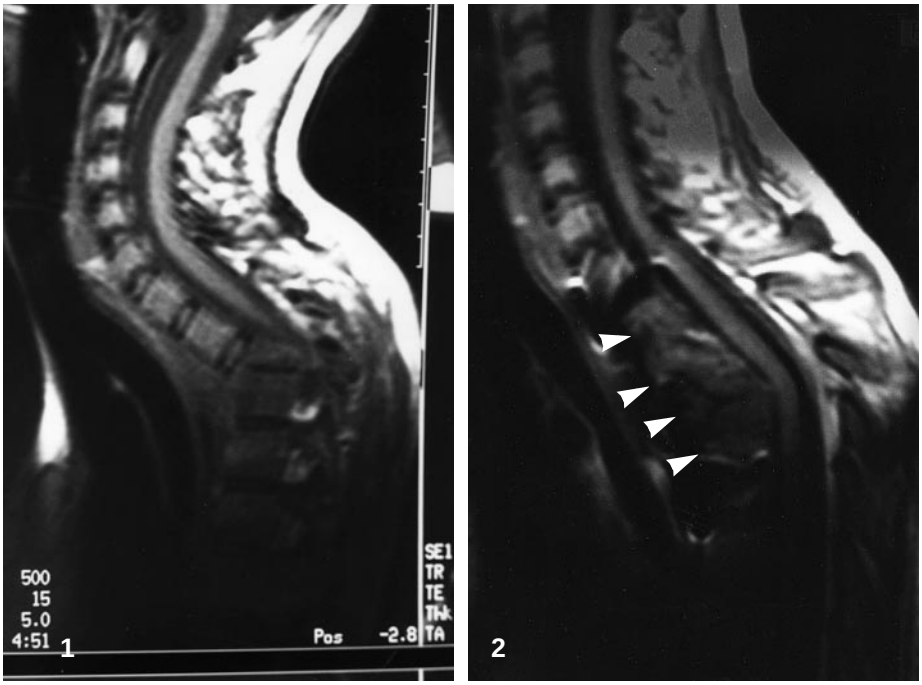

Fig. 1 Preoperative magnetic resonance image taken on admission, demonstrating complete destruction of the vertebral body of the second thoracic vertebra causing severe instability with a major kyphotic deformity. This lateral view shows that the spinal cord was stretched and compressed over the apex of the kyphosis

Fig. 2 Magnetic resonance imaging at 2-year follow-up shows spinal cord decompression and correction of kyphosis. The deformity was reduced by approximately 50°. *Arrows* outline the interbody graft interlocked into the troughs in T1 and T3 vertebral bodies. The anterior titanium device is not seen

bodies of the upper thoracic vertebrae. This exposure confirmed that the spinal cord was stretched at the apex of the kyphosis, and was adequate for complete anterior cord decompression, radical excision of the adjacent intervertebral discs, kyphosis correction, and stabilization. Placement of a tricortical and cancellous block harvested from the anterior iliac crest was performed to restore vertebral height and to obtain anterior interbody fusion. Mechanical stability of the graft segment was achieved by anterior plating fixation using a titanium device. At the time of surgery, laboratory investigations showed that the white blood-cell count was normal and the sedimentation rate was 33 mm/h. Bacterial and fungal cultures from blood were negative. Histologic examination of the removed bone lesions demonstrated a predominance of lymphocytes, neutrophylic polymorphonuclear leukocytes, and a few plasma cells. Granulation tissue was evident and around this tissue osteoblasts and signs of new bone formation were observed. Aerobic and anaerobic bacterial cultures including cultures for fungi and tubercle bacilli of the vertebral bone removed were negative. The patient improved neurologically and by the 12th postoperative day full neurological recovery was achieved. When the patient was ambulant, a Milwaukee jacket was added for 6 months to provide supplementary support to counteract the natural force of the kyphosis. After 12 weeks, the patient was physically able to return to school. At 2-year follow-up, neurological and functional outcomes are fair and magnetic resonance imaging shows good restoration of the sagittal spine alignment despite residual mild kyphosis, and restoration of a normal sagittal diameter of the spinal canal (Fig. 2).

Discussion

This is the first reported case of CRMO in a vertebral location associated with severe kyphosis, spinal cord compression, and neurological dysfunction which required prompt surgical anterior decompression, correction of the kyphosis, and stabilization. In this case, the diagnosis of CRMO was prior to its occurrence in the vertebral location. Histological findings from the vertebral bone confirmed the previous results of the femoral biopsy and were consistent with those of CRMO as described in the literature [2, 3, 8, 10]. CRMO in vertebral locations is rare, as is shown by the scarcity of reported cases in the literature [2, 5, 6, 21]. Involvement of the spine can be by either mixed lytic destruction and sclerosis or by partial or complete collapse of vertebral bodies presenting as vertebra plana. Vertebra plana caused by CRMO has been reported in five cases with mild kyphotic deformity without any instability or neurological complication [5, 21]. In the case of vertebra plana eosinophilic granuloma, by far the most common cause of vertebra plana, has to be considered in the differential diagnosis [21].

In all reported cases treatment was nonoperative with a short period of bed rest followed by protected mobilization, and outcome was fair in all cases [2, 5, 21]. Carr et al. [6] were the only group to describe a surgical procedure using posterior fixation and fusion, in a case of a 9-year-old girl who presented with CRMO at the fifth thoracic vertebra level with severe kyphosis but without neurological involvement; her outcome was fair. Physicians have to be aware of this major complication, and this case report strongly suggests that a very close survey of the spinal alignment and of the neurological status is advisable in cases of CRMO in a vertebral body location. In our case, the patient had a single lesion at T2 producing major instability with severe spinal cord compression causing neurological dysfunction. Prompt anterior surgical decompression and spinal stabilization with bone grafting and titanium plate fixation saved the spinal cord function with full neurological recovery.

Conclusion

This case report supports the conclusion that CRMO in spinal locations must be taken very seriously by physicians because severe kyphosis with spinal cord compression is possible, causing neurologic dysfunction. In our case, prompt anterior surgery allowed the patient full neurological recovery and at follow-up the vertebral reconstruction was confirmed to have avoided physical impairment.

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