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CASE REPORT

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Cystic angiomatosis of bone with massive osteolysis of the cervical spine

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Abstract The case of a 2½-year-old boy with diffuse cystic angiomatosis of bone is presented. No evidence of visceral involvement was recorded. The clinical, radiographic and histologic findings during the course of the disease process are discussed. At the age of 15 years the patient died of neurologic alterations developed due to massive osteolysis and collapse of the cervical spine, and severe dyspnoea, secondary to persistent bilateral pleural effusions.

Key words Angiomatosis · Bone · Osteolysis · Cervical spine

Introduction

Cystic angiomatosis of bone is a rare condition of obscure origin. It represents either a benign neoplasm or a congenital vascular malformation of multicentric origin. In most cases it is recognized by the incidental radiologic demonstration of multiple cystic lesions scattered throughout the skeleton. The chance of survival is limited especially when similar lesions arise in soft tissues, particularly the spleen, liver and lungs [9, 11].

The case of a male child with diffuse skeletal angiomatosis is presented. The gross facial abnormality caused by the oral manifestations of the disease has already been described [8]. The general features of this disease, and particularly the combination of massive osteolysis and collapse of the cervical spine as well as the pleural effusions that developed in this patient, are discussed.

Case report

A previously healthy 2½-year-old boy started limping without any history of injury. He had pain radiating from his hip, and radiographic examination showed cystic lesions involving the right proximal femoral metaphysis.

A year later he sustained a fracture of the shaft of the left femur in an automobile accident. At that time skeletal survey revealed cystic lesions involving the entire axial skeleton – skull, vertrebrae and pelvis – and the long bones – both humeri, femora and tibiae – (Fig. 1).

On physical examination the child's general condition was good. Laboratory findings were all within normal limits and a technetium-99m bone scan showed no uptake in the cystic areas.

Histologic examinations revealed predominantly cystic cavities lined by a single layer of endothelium and separated by collagen septa. Only a few of them contained a homogeneous pink-staining coagulum or hypervascular fibrous tissue. All cavities were surrounded by dense sclerotic bone with prominent osteoblastic rimming of the bone trabeculae. The diagnosis was cystic angiomatosis of bone.

At the age of 12½ years, signs of increased intracranial pressure developed and cord compression was evident on MRI scans (Fig. 2). The arch of C1 was removed and laminectomy of C2 was performed by neurosurgeons. The postoperative course was uneventful and the patient's symptoms improved.

Six months later he developed progressive myelopathy that, within a month, led to spastic paraplegia. From that time and for the next 22 months the patient underwent repeated admissions due to severe cervical lordosis and acute cervico-thoracic junction kyphosis secondary to vertebral massive osteolysis and collapse (Fig. 3). He underwent a multistage extensive cervical spine surgery by J. K. W. That included removal of vertebral bodies for cord de-

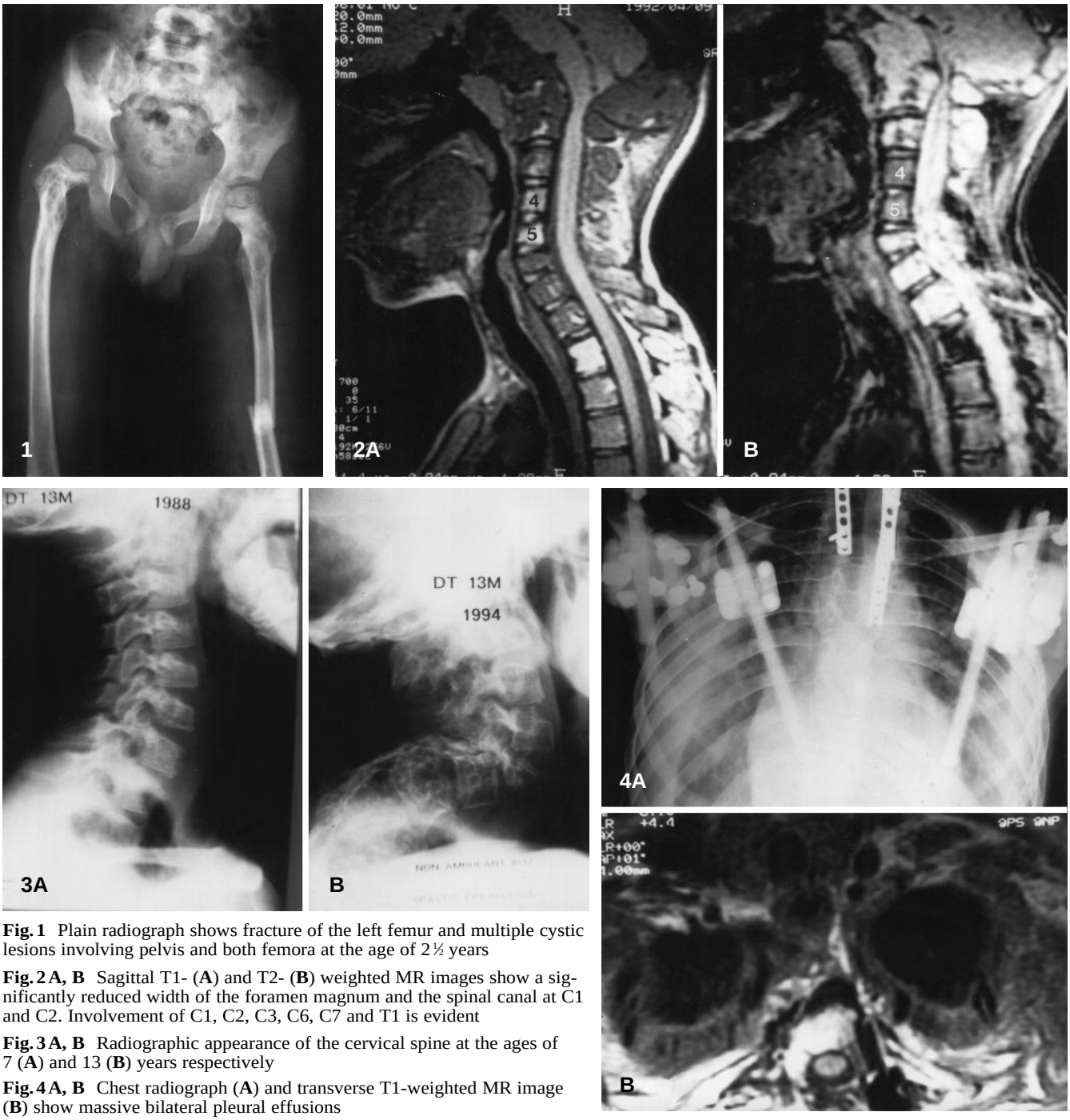

Fig. 1 Plain radiograph shows fracture of the left femur and multiple cystic lesions involving pelvis and both femora at the age of 2½ years

Fig. 2 A, B Sagittal T1- (**A**) and T2- (**B**) weighted MR images show a significantly reduced width of the foramen magnum and the spinal canal at C1 and C2. Involvement of C1, C2, C3, C6, C7 and T1 is evident

Fig. 3 A, B Radiographic appearance of the cervical spine at the ages of 7 (**A**) and 13 (**B**) years respectively

Fig. 4 A, B Chest radiograph (**A**) and transverse T1-weighted MR image (**B**) show massive bilateral pleural effusions

compression as well as stabilization through inserting bilateral mass plates from the occiput to T5 and grafting with cortico-cancellous iliac bone.

The main histologic abnormality from the resected fragments of bone was the presence of large vascular channels in the marrow spaces, resembling angiomatosis of bone.

The patient's neurologic status improved, he developed increasing function in his limbs and regained the ability to stand and walk independently.

During the same period the patient had bilateral sterile pleural effusions, the right greater than the left (Fig. 4). These were primarily drained by pleural taps and, when they were larger, causing progressive dyspnoea, by chest tubes. Analysis of the fluid was not typical of a chylothorax.

Following his neurologic recovery, the patient suffered a significant new deterioration of the neurological status. MRI scans revealed compression on the cord due to progression of the cervical spine collapse. The patient became virtually paraplegic, with loss of bowel and bladder control. The spinal cord was decompressed by J. K. W.

Postoperatively, the patient developed large infected pleural effusions, showed severe dyspnoea and cyanosis and finally died at the age of 15 years.

Discussion

A combination of radiographic and histopathologic characteristics is always necessary to confirm the diagnosis of cystic angiomatosis and rule out similar conditions such as polyostotic fibrous dysplasia, hyperparathyroidism and histiocytosis X [1].

In children, diagnosis usually follows skeletal pain and pathologic fractures. Clinical examination for involvement of abdominal or thoracic organs is essential. On plain radiographs the lesions are sharply defined with sclerotic borders, while those in the vault of the skull lack a sclerotic rim. Periosteal reaction is associated only with pathologic fractures. The disorder never spreads from bone to the surrounding soft tissues. Results of isotope bone scan are normal, because no significant osteoblastic activity is seen [2]. The MR findings are highly suggestive of the disease: sharply defined lesions with low signal intensity relative to muscle on T1-weighted images and extremely bright signal – much brighter than that of fat – on T2-weighted, STIR and gradient echo pulse sequences [5].

Biopsy is always necessary to confirm the diagnosis, but it often yields nonspecific histologic material, so the most satisfactory specimen is an involved rib [1]. The characteristic pathologic tissue consists of cavities lined by endothelium and containing blood or lymph. Depending on whether vascular or lymphatic tissue predominates in the lesional tissue, the disease is characterized as haemangiomatosis or lymphangiomatosis respectively, and it is termed angiomatosis when both are present [12].

In the reported case no extraskeletal soft tissue or visceral lesions were detected. The patient showed an almost stationary disease process for approximately 10 years. The dramatic change to massive osteolysis and collapse of the cervical spine after the first decompressive operation at the region remains unexplained.

Massive osteolysis – also known as disappearing bones or Gorham's disease – is a histologically similar clinical entity, but in destructiveness it far surpasses angiomatosis. Moreover, the two disorders have distinct clinical differences [6, 12]. Two cases of skeletal lymphangiomatosis with or without visceral involvement, which changed to resemble massive osteolysis in the cervical spine, have previously been described. The course of the disease process in those patients was similar to the one presented in this paper [4, 6]. It is important to note that in all cases this transformation concerns only the cervical part of the spine.

The incidence of chylothorax with massive osteolysis or lymphangiomatosis of bone has already been reviewed. In the former, it seems likely that there is thoracic duct destruction caused by the same bone-destructive angiomatous malformation, while in the latter, the source of chyle may be either an undiagnosed mediastinal lymphangioma or the abnormal chest wall lymphatic channels. In both clinical entities, conservative treatments such as dietary manipulation or thoracentesis have not been successful, and unabated losses of chyle may lead to death by inanition or respiratory compromise [3, 10]. In our patient surgical treatment was rejected, since the effusions were larger only after respiratory infections or postoperatively. It has been shown that, in angiomatosis, the malformation does not spread to soft tissue around bone [7]. However, no other source of the fluid could be found than the angiomatous bones of the thoracic cage.

It must be emphasized that this is the only known case of skeletal angiomatosis without definite visceral involvement that developed non-chylous pleural effusions, and changed to massive osteolysis of the cervical spine. However, questions remain as to whether this was a true change to massive osteolysis, how it was elicited and why it was confined to the cervical spine.

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