Limp as unusual presentation of Langerhans’ cell histiocytosis

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Abstract An isolated eosinophilic granuloma involving the posterior elements of a lumbar vertebra is reported in a 3-year-old boy presenting with progressive limp. Radiological investigations revealed osteolysis of the L5 right pedicle. MRI showed a well-defined homogeneous mass with nonspecific signal intensity. An unusual feature was the paravertebral muscular location of the largest part of the tumour indicating a possible soft tissue origin. Immunohistochemical studies were typical for Langerhans’ cell histiocytosis.

Conclusion A limp can be due to lumbar and paravertebral muscular location of Langerhans’ cell histiocytosis.

Key words Eosinophilic granuloma · Histiocytosis · Langerhans’ cell · Spinal diseases · Radiography

Abbreviations EG eosinophilic granuloma · LCH Langerhans cell histiocytosis

Introduction

Eosinophilic granuloma (EG) represents the localized and most benign form of bone Langerhans’ cell histiocytosis (LCH) [2, 7, 10]. Its classical vertebral manifestation is “vertebra plana” described in 1924 by Calve [2, 8, 10]. Involvement of the posterior vertebral elements is unusual with only a handful of cases reported previously [1, 2, 5, 6, 8, 10]. Soft tissue changes may be associated and usually represent inflammatory reaction or extension of the primary osseous lesion [2, 10].

Case report

A 3-year-old boy was admitted to our hospital because of a limp of a few weeks duration. He started to complain of pain in the anterior part of the right thigh 6 weeks before admission. The pain was usually mild and ceased spontaneously, but at times the child became hyperalgic and even awakened by this pain during the night. According to his mother, this pain was associated with mild limping. He was seen in another hospital where radiographs of both hips and knees, and hip echography were performed. All these tests were considered normal. A trial with Ibuprofen syrup administration for 10 days gave no positive result. He was then referred to our hospital by his general practitioner for further evaluation.

On examination, the child was not in acute distress, but obviously uncomfortable. The rectal temperature was 37.6°C. Mobilisation of the legs was not limited and there were no signs of inflammation at any articulation. Percussion of the spine raised no particular pain. When the child was walking, an external rotation of the left foot was observed, without obvious limp. The neurological examination was completely normal, there were no pyramidal signs, and the osteo-tendinous reflexes were symmetrical. The remainder of the physical examination was normal.

Laboratory investigations were as follows: haemoglobin 11.5 g/dl, white cell count 9.800 cells/mm³, platelets 264.000/mm³, CRP...
2.5 mg/dl (normal 0.5 mg/dl). Urea, creatinine, α-feto protein, CEA and neuron-specific enolase were normal.

A technetium bone scan showed a small increase in radioactivity in the right side of the fifth lumbar vertebra. An X-ray of the lumbar spine revealed osteolysis of the corresponding pedicle (Fig.1) and MRI examination disclosed an associated paravertebral muscular mass (Fig.2).

A CT scan guided biopsy was performed. The histology was quite characteristic of LCH. Immunohistochemically, these typical cells expressed S-100 protein and CD 1A.

After surgery via a posterior approach to the lumbar spine, the child made an excellent recovery without spinal instability. X-rays were otherwise normal without any other lesion, in accordance with the isotope bone scan. A close follow up with no adjuvant treatment was proposed to the family. No particular complaints were noted, and the physical examination showed only a slight decrease of the right rotular reflex during the first outpatient visits. One and a half years after surgery, the child is living normally and free of symptoms.

Discussion

Langerhans’ cell disease of bone is a disorder of histiocytic proliferation with variable and often unpredictable evolution [7]. It typically presents in children, accounting for 1% of all paediatric tumoural or tumour-like lesions involving the bone; although rare, adult cases have been reported [7]. The most common complaint at presentation is pain [2, 7, 10, 11], often worse at night [2, 7, 10]. In case of vertebral involvement, neurological signs and symptoms due to cord or nerve root compression may occur [2, 3, 8, 11].

It has been reported in many skeletal sites, most commonly in the skull, but also in the mandible, spine, ribs, and long bones, particularly the femur and the humerus [2, 10]. In the spine, the thoracic vertebrae are most often involved, followed by lumbar and cervical vertebrae [2, 10].

The lesions usually develop in the vertebral body and are of the lytic type [2, 8, 10]. Varying degrees of vertebral collapse may be associated leading to a flattened vertebral body, classically known as “vertebra plana” [1, 2, 3, 5, 6, 8, 10, 11]. However, this finding is not pathognomonic for the disease.

Isolated involvement of the posterior elements of the vertebrae is unusual in Langerhans’ cell disease with only a few cases reported previously (Table 1). The lesions can arise from the pedicles [1] or from the spinous process [5] and are classically lytic.

Soft-tissue abnormalities may be present: inflammatory changes or haematoma during the early phases of the disease, extradural expansion of the osseous lesion leading to neurological compromise, or extradural non-osseous mass. The incidence of muscular paravertebral involvement is unknown, but is probably more frequent in cases of posterior vertebral lesions [1, 6]. It may represent the extension of the osseous tumour, but it could also be a primary disease of the muscle with secondary extension in the epidural space and erosion of the

Fig. 1 Anteroposterior X-ray of the lower lumbar spine shows loss of the right pedicle and transverse process of L5 (black arrows)

Fig. 2a Noncontrast axial T1-weighted MR image of L5 (450/14) shows a paravertebral soft-tissue mass of low signal intensity involving the right paraspinal muscles (curved black arrow) and extending by the foramen of L5 into the extradural space. b Axial T1-weighted MR image after intravenous injection of gadopentetate dimeglumine demonstrating homogeneous enhancement of the mass (black arrows)