Solitary eosinophilic granuloma of the adult lumbar spine

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Abstract

Introduction Eosinophilic granuloma (EG) is a benign bone tumor that rarely occurs in adults. It is usually found occurring in flat and long bones, but spine is often affected too. EG is of unknown aetiology, and the course of the disease is unpredictable.

Materials and methods Two cases of EG of the adult lumbar spine are reported, representing a difficult challenge for diagnosis and treatment.

Conclusion CT scan guided trocar biopsy allowed to achieve a definitive diagnosis, and CT scan guided intralesional infiltrations of corticosteroids were the successful treatment.

Keywords Eosinophilic granuloma · CT-scan · Intralesional infiltrations of corticosteroids

Introduction

Langherans' cell histiocytosis (LCH) of the bone is a disorder of unknown origin, characterized by proliferation of abnormal Langerhans' cells. This entity represents a spectrum of disease that includes eosinophilic granuloma, Hand-Schueller-Christin syndrome and Letterer-Siwe disease. Localised LCH of bone is a benign tumor-like condition, often affecting the skull and spine, which is commonly referred to as eosinophilic granuloma (EG).

Solitary EG was first described by Lichtenstein and Jaffe in 1940 [1, 2] as a very rare benign osteolytic lesion that usually occurs in flat and long bones of children in the first

two decades of life. The course of the disease is unpredictable and can be associated with important morbidity due to pain, limited activity and pathologic fracture [3].

In children, EG usually involves the thoracic spine, and vertebrae appears as “flattened” (vertebra plana). EG is rarely observed in adults, especially in the spine, in the adults the vertebrae usually do not collapse, and only small areas of the vertebral body are affected by the disease.

Due to the uncommon occurrence and the aspecific lytic imaging, mimicking all the pure lytic condition (giant cell tumor, myeloma, lymphoma, Ewing sarcoma, metastatic disease), EG in the adults is rarely suspected, lately diagnosed and represents a difficult challenge for treatment.

In this paper two cases of eosinophilic granuloma in the adult spine are reported.

Case reports

Case 1

A 35-year-old woman with a 2-month history of severe lumbar pain was admitted to our hospital. The patient had no history of fever, weight loss or systemic symptoms. The back pain had developed spontaneously, and at the beginning she was treated with oral analgesics without benefits. There were no neurological signs or symptoms. Laboratory studies were within normal limits.

X-rays and CT-scan done elsewhere showed a lytic lesion involving the right side of the vertebral body of L4 with breakage of the lateral cortex. MRI demonstrated a lesion of the vertebral body of L4 with partial collapse of the right side and involvement of the adjacent soft tissue (Fig. 1). The lesion had a C–D layers involving sectors 7–10 according to Weinstein–Boriani–Biagini classification [4].

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Fig. 1 **a** Plain LL X-rays, **b** CT scan and **c** MRI of the lumbar spine show the presence of a lytic lesion involving the right side of the vertebral body of L4

Bone scan showed an increased radio-trace uptake over L4 vertebra.

An incisional transpeduncular biopsy was performed. A soft grayish-yellow granulation tissue affected the adjacent soft tissue, involving the ipsilateral muscle psoas. The combination of imaging and microscopic findings suggested the diagnosis of eosinophilic granuloma. Two following intralesional infiltrations of corticosteroids under a CT scan were performed (Fig. 2). At a 4-year follow-up the patient was asymptomatic, and the imaging showed a

Fig. 2 First intralesional infiltration of corticosteroids under a CT scan at 1 month after diagnosis

decrease in size of the bony lesion and the complete reformation of the right lateral cortex of the vertebral body (Fig. 3).

Case 2

A 30-year-old woman presented with a 4-months history of predominantly nocturnal lumbar pain. Her symptoms were unresponsive to medical and physical therapy. She was previously healthy and the laboratory studies were normal. On physical examination pressure over the spinus process of L2, L3 and L4 produced acute pain. No neurological symptoms were observed. Lumbar X-ray examination showed a lytic lesion involving the upper part and the posterior wall of the L3 vertebral body.

MRI of the spine was performed. The vertebral body and the left pedicle of L3 showed isointensity on T1-weighted images and hyperintensity on T2-weighted images. The MRI signal inside the lesion was heterogeneous. The adjacent discs were normal in shape and signal intensity, and there was no appearance of perilesional soft tissue edema. CT scan revealed a two-component osteolytic lesion of the vertebral body with breach of the posterior wall and superior cortex (Fig. 4).

Bone scan showed an increased uptake over L3 vertebra.

Fig. 3 **a** First axial CT-scan shows the osteolytic lesion with breakage of the right lateral cortex of L4. **b** Axial CT-scan performed 6 months after second steroid-infiltration. **c** Axial CT-scan at 4-year follow-up

Fig. 4 **a** Plain LL X-rays, **b** CT scan and **c** MRI of the lumbar spine show a two-component osteolytic lesion of the vertebral body of L3 with breach of the posterior wall and superior cortex

The lesion had a C–D layers involving sectors 5–8 according to Weinstein–Boriani–Biagini classification [4].

A biopsy was performed by trocar under a CT scan, and the reported diagnosis was eosinophilic granuloma. One week after the patient was treated with steroid infiltration. The patient recovered uneventfully without residual pain or deficits. Imaging did not correspond to clinical features. As a matter of fact, the following CT scan (performed after 2 months) did not show reduction or changes in shape of the vertebral lesion.

A second steroid infiltration under a CT scan was performed after 2 months. At a 1-year follow-up the patient was asymptomatic although the radiological picture was unchanged.

Discussion

Etiopathology of Langerhans' cell histiocytosis is still controversial although an immune-mediated reactive process, inflammatory or infectious diseases, viral infections and clonal proliferation of cells have been proposed as cause of the disorder [5–7].

EG of bone accounts for 50–60% of all cases of Langerhans' cell histiocytosis [8]. This lesion is most common in children; 80% of the patient are below 10 years old [9]. The diagnosis of spinal EG is based on clinical features, radiologic imaging studies and histopathologic examination. A wide spectrum of clinical features is reported: no symptoms to pain, swelling and tenderness. The most common radiologic sign is a lytic lesion of the vertebral body with a possible partial collapse. In children, full collapse of the vertebral body may lead to the so-called “vertebra plana” [10]. This image, once considered a pathognomonic sign of EG, is indeed the pattern of the late

evolution of the disease. On histological examination aggregation of histiocytes and eosinophilic leukocytes are typical features. Laboratory tests are usually normal.

Solitary EG of the spine involves differential diagnosis with osteoblastoma, aneurysmal bone cyst, Ewing's sarcoma, acute leukemia, Gaucher's disease and metastatic tumor [11]. Spondylitis can also be considered. Differential diagnosis is based on complete imaging, but it needs histologic confirmation.

In spinal EG the thoracic vertebrae are affected most often, followed by the lumbar and cervical vertebrae [12]. Solitary EG is usually a self-limiting disorder, and spontaneous healing of the lesion is possible [9]. In the literature, high-evidence studies about treatment of EO are not described. The treatment can be variable based on symptoms, extent of disease, morbidity and location of the lesion.

The two reported case were treated with intralesional infiltration of corticosteroids under a CT scan. We believe that this treatment is safe, minimal invasive and very effective. Furthermore, it is described that after steroid infiltration the pain can resolve within a week [13]. This kind of treatment was also recommended by Capanna et al. [14] while Egeler et al. reported results of treatment with intralesional steroids comparable to those of other forms of treatment described in the literature [13]. The conservative treatment such as clinical observation, casting or bracing is possible [15]. Some authors advocate operative treatment as surgical curettage and bone grafting of the lesion [16, 17]. Radiation therapy and chemotherapy are also reported, especially for lesions for which surgical complication would be high and for cases with numerous bone lesions [18, 19].

As a conclusion, EG should be always considered as diagnosis of a low growing pure lytic condition in the adult

spine. CT guided trocar biopsy should be first performed and if diagnosis is confirmed, corticoid injection by the same percutaneous approach can be a successful procedure.

Conflict of interest None of the authors has any potential conflict of interest.

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