Diagnosis of Langerhans cell histiocytosis in a patient with vertebral lytic lesion and low back pain

Secilay Gunes, Haydar Gök, Sehim Kutlay

Mediterr J Rheumatol 2016; 27(3):108-10
Diagnosis of Langerhans cell histiocytosis in a patient with vertebral lytic lesion and low back pain

Secilay Gunes¹, Haydar Gök², Sehim Kutlay²

¹Kars Public Hospital Department of Physical Medicine and Rehabilitation, Kars, Turkey, ²Ankara University Faculty of Medicine, Department of Physical Medicine and Rehabilitation, Sıhhiye, Ankara, Turkey

ABSTRACT

The identification of lytic vertebral lesions on magnetic resonance imaging usually points to tumor metastasis in adults. We present a 47-year-old female complaining of back pain who had palpable breast mass and a vertebral lytic lesion on magnetic resonance imaging. Detailed investigation yielded the diagnosis of Langerhans cell histiocytosis rather than tumor metastasis.

Keywords: Langerhans cell histiocytosis, low back pain, vertebral lytic lesion.
INTRODUCTION
Langerhans cell histiocytosis (LCH) is a rare disorder caused by the clonal proliferation of specialized dendritic cells. The disease comes in 3 different forms: Eosinophilic granuloma, which is the solitary site localized form; the Hand-Schuller Christian disease characterized by bony lesions, exophthalmos and diabetes insipidus; and the most severe, Letterer-Siwe disease, which has a multi-system involvement. The etiology and pathogenesis of LCH remain unclear. There are two widely accepted hypotheses, one being the disorder due to immune regulation and other being neoplastic proliferation. Langerhans cell histiocytosis cases are frequently reported under the age of 15 years. The age of onset varies depending upon the form of LCH, and the disease is rarely seen in adults with an incidence around 1–2 cases/million persons per year.

CASE REPORT
A 47-year-old female presented complaining of back pain not relieved by NSAID therapy. The pain increased at midnight and used to wake the patient. There was no other complaint apart from back pain. Across physical examination, she only had a local tenderness on the thoracolomber junction on deep palpation. Other findings of neuromuscular examination were normal. Laboratory investigation revealed no abnormal findings except for an elevated CRP [7.4 (0-3)]. The lumbosacral spine X-Ray was normal. Magnetic resonance imaging (MRI) revealed a 15x10 mm contrast holding osteolytic lesion with a peripheral edematous rim on T12 vertebra corpus which was referred to the PET CT by the radiologists. (Figures 1-2) The PET CT revealed elevated fo- cal metabolism only on the T12 vertebra corpus (SUV-max: 17.4). The typical radiological features of the lesion suggested the possibility of a neoplastic lesion or metastasis. The patient was screened for a breast or thoracoabdominal cancer. Breast USG revealed 9.2x5.4 mm and 14.9x6.5mm mass at the upper outer quadrant on breast. Mammography was concordant with BI-RADS (Breast Imaging Reporting and Data System) Category 3 - which most likely means benign mass - but a biopsy was recommended. Thoracoabdominal pelvic CT revealed milimetric nodules which became denser on the upper lung. In concurrence with the lytic lesion on the T12 vertebra corpus, the appearance was found compatible with LCH. A needle biopsy was performed on the T12 vertebra and the histopathological diagnosis was concordant with fibrohistiocytic proliferation. Based on these findings, she was diagnosed with pulmonary Langerhans cell histiocytosis (PLCH). After treatment with local radiotherapy (200 cGy/day total: 2000cGy), the back pain was resolved. She decided to quit smoking and was under follow-up with no other treatment.

DISCUSSION
Langerhans cell histiocytosis is known as a pediatric disease, but recent studies have reported as many as 39% of diagnosed cases in adults. The etiopathogenesis of LCH is still unknown, but it is believed to be an immune-mediated reactive process accompanying viral infections, inflammatory and infectious diseases that could result in clonal proliferation of dendritic cells. Langerhans cell histiocytosis can involve any bone, but it has a predilection for the axial skeleton (skull, spine,
scapula, and pelvis). Langerhans cell histiocytosis in the spine has been reported to range between 6.5% and 25%. It usually involves the thoracic spine, followed by the lumbar and cervical vertebrae. Such involvement usually affects the vertebral body, with only 5% of cases associating with the posterior arch. The typical presentation of LCH of the spine is back pain without any neurological symptoms. Skeletal involvement can be demonstrated by lytic lesions on X-ray, but it must be kept in mind that lytic lesions become apparent only when 30–50% of the bone mineral density is already lost: it may be difficult to assess the vertebrae due to superposition of bowel gas. None of the adult cases reported had a radiological evidence of vertebra plana. In our case, direct radiography was normal. Magnetic resonance imaging presents typical single or multiple osteolytic lesions, round or oval-shaped. Although some authors suggested that vertebral biopsy is usually unnecessary, Ewing sarcoma, osteosarcoma, lymphoma, osteosarcoma, vertebral osteomyelitis, tuberculosis and hemangiomas may underlie lytic lesions.

Back pain in LCH usually increases at midnight. This must be kept in mind during the differential diagnosis of inflammatory lower back pain, which affects approximately 3% of adults. Inflammatory low back pain can be distinguished from other types of lower back pain by some features: young age at onset, typically seen in those under 40 years old, gradual onset of pain, symptoms of back pain that improve with exercise, pain does not improve with rest, pain at night, often waking a person in the second half of the night, morning stiffness that lasts for more than 30 minutes, pain lasting for more than 3 months, alternating buttock pain. Our patient did not have these symptoms, so we excluded inflammatory lower back pain.

Treatment includes observation, immobilization, NSAIDs, chemotherapy and radiotherapy. Radiation therapy is not suggested as a first option for some authors, as it has a potential for secondary malignancy, and it can be harmful for the vertebral growth plate in the skeletally immature patient. Gregenberg et al. reported secondary malignancy only in 4 patients (4%) in their studies. Since our patient had only one lytic lesion on vertebra with no disseminated LCH, a low dose radiotherapy was preferred. Low dose radiotherapy still appears a safe treatment option for LCH.

There are rare presentations of synchronous PLCH with lytic bone involvement. Pulmonary Langerhans cell histiocytosis usually presents with respiratory symptoms and occur primarily in middle-aged cigarette smokers. It is important to encourage patients to discontinue cigarette smoking during treatment. Concomitant breast nodules and bone involvement can also be seen in the Erdheim-Chester disease which is a rare non-Langerhans cell histiocytosis (nLCH).

The disease is characterized by the infiltration of tissues by foamy nLCH cells. It affects the bones, eyes, lungs, pituitary glands, breasts, etc. Distinctly, the characteristic radiographic findings are bilateral symmetric sclerosis of the long bones.

CONCLUSION

Although LCH is a childhood disease, it must be taken into consideration in the differential diagnosis of patients presenting with an osteolytic lesion in vertebra and lower back pain.

CONFLICT OF INTEREST

The authors declare no Conflict of Interest.

REFERENCES