Solitary Eosinophilic Granuloma in the Lumbar Spine: Case Report

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ABSTRACT
A case of a 3-year-old patient with osteolytic lesion in the lumbar region is presented. The clinic-radiological suspicion was infection, primary or metastatic tumor of spine. Subsequently, a CT guided biopsy proved an eosinophilic granuloma of bone. Following medical treatment after two month, there was improvement in patient medical status.

Keywords: Eosinophilic Granuloma; Inflammation; Tumor

INTRODUCTION
Eosinophilic granuloma of bones is the modest and commonest presentation of Histiocytosis-X malady. It affects male children in more than 90% of the cases. The lesion frequently presents as a solitary one and is located in flat and long bones, the vertebral spine and the skull. Inflammation, autoimmunity and loss of controlled proliferation of Langerhans cells are considered causes. This benign tumor is usually asymptomatic or may act as a palpable, tender mass over the involved bone. Uncommonly it results in epidural hematomas, suppression of bone marrow and pathological properties. When at skull, headache, neurological symptoms, chronic mastoiditis and exophthalmos may be related presentations. The diagnosis is based on the detection of the lesion with roentgenography, CT, MRI, scintigraphy and is set via histological evaluation after excision or guided biopsy. Eosinophilic granuloma can improve without treatment while passage from childhood to adulthood. If not, the recommended treatment is the surgical curettage of the tumor or local infusion of cortisone. Chemotherapy, radiotherapy and systemic use of cortisone are successful for multiple bone lesions.

We present a 3-year-old girl who developed eosinophilic granuloma at the vertebral body without having any previous history of bone tumors.

CASE PRESENTATION
A 3-year-old girl was admitted to our hospital with complain of acute pain due to falling down as a result of weakness in lower limbs. She was previously healthy and the laboratory studies were normal. No fever, elevated regional temperature nor lymphadenopathy was detected. Physical examination revealed lower limbs weakness and tender point in L2 and L3 level. There were no numbness, or remarkable abnormal tendon reflexes and muscular tone. On physical examination no pathological findings detected and erythrocyte sedimentation rate was normal.

MRI of the spine was performed. The vertebral body of L2 showed isointensity on T1- and T2-weighted images. The adjacent discs were normal in shape and signal intensity, and there was no appearance of perilesional soft tissue edema. CT scan revealed an osteolytic lesion of the vertebral body (Figure 2). Plain radiography of lumbar show L2 body collapse (Figure 3).

On the basis of the radiological features, there was a high possibility of a neoplastic lesion. A CT-guided needle biopsy of the L2 vertebral body was carried out. Histopathology exhibited proliferation of Langerhans cells with eosinophilic cytoplasm and round to oval neuclci arranged in sheets, with some eosinophils, lymphocytes
and plasma cells that replaced hematopoietic elements (Figure 4). Immunohistochemistry confirmed Langerhans cell histiocytosis with diffuse immunoreactivity of S-100 and CD68. The patient received only medical treatment and in follow up two month later medical condition was improved.

**DISCUSSION**

Eosinophilic granuloma is one of the most uncommon bone tumors responsible for less than 1% of them. In 90% of the reported cases it affects children less than ten. There is an evident predilection to males (2:1) ⁴. It usually presents as a monostatic lesion at flat and long bones (70%) ⁵, and the skull and the vertebral spine. However in this girl it presented as solitary lesion in lumbar spine that is less common. A unique cell, the Langerhans cell is detective ⁶. It includes Birbeck granules whose role is yet unrevealed. Eosinophils, lymphocytes, fibroblasts and foam cells may be also discovered but none of them are pathognomonic. The only definitive immunological marker is the OKT6 while the common S-100 protein is frequently positive as well ⁷. Eosinophilic granuloma can be asymptomatic or present as local swelling, pain or tenderness. Based on the site of the tumor, it may result in neurological symptoms such as numbness, limping, fracture, loosening of teeth, otitis media ⁸ or exophthalmos. A hematoma in skull after a mild injury is a usual presentation ⁹. No fever or other signs of inflammation have been reported. The blood tests display rise of leucocytes and eosinophils in about 7% of the cases ¹⁰. Erythrocyte sedimentation rate is more than the normal levels. The tumor’s material is sterile but there have been reports about the existence of staphylococcus and streptococcus ¹¹.

Radiological imaging of eosinophilic granuloma is essential to detect tumor activity and nature. Plain
radiograph exhibits its size and borders. The cortex of the affected bone may be seen as thin, eroded or thickened because of new bone generation. Mainly at skull there may be one or multiple osteolytic oval defects with regular or irregular borders. CT and MRI show the precise size and borders of the tumor as well as the location of the surrounding tissues and a probable hematoma. Radionuclide bone scan with technetium, gallium or thallium display an enhancing mass and easily detect other foci or recurrence points. Ultrasound is only carried out for guided biopsies. Diagnosis is based on histological evaluation.

The etiology of eosinophilic granuloma stays unknown but newly there have been some theories. The strongest ones are inflammatory processes, autoimmune disorders and an out of control proliferation of Langerhans cell. Eosinophilic granuloma does not result in malignant transformation. If it expands elsewhere is called Hand-Schuller-Christian disease and may present as diabetes insipidus, cerebellar, hypothalamic and with other central nervous system symptoms. The prognosis is estimated based on the age of diagnosis and the number of foci.

Figure 3. Plain radiography revealed uniform collapse of vertebral body.

Figure 4. Histopathology exhibited proliferation of Langerhans cells with eosinophilic cytoplasm and round to oval nuclei arranged in sheets.
This tumor may improve without treatment but there is probability of relapsing during the first year after diagnosis. Thus, even after surgical curettage a careful follow-up must be considered. If a secondary lesion seen then chemotherapy or further excision have to be performed. In our case solitary involvement of lumbar spine as a region that less affected especially in a girl is interesting.

REFERENCES