Unusual location of eosinophilic granuloma

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ABSTRACT

Eosinophilic granuloma (EG) is a benign self-limiting disease, which belongs to the spectrum of Langerhans' cell histiocytosis (LCH). The etiology of LCH remains unknown, although the evidence indicates that it is a clonal proliferative disorder of Langerhans cells, it has also been characterized as reactive disorder, neoplastic process and a berrant immune response. Eosinophilic granuloma is characterized by single or multiple skeletal lesions occurring predominately in children, adolescents and young adults, it accounts for 70% of LCH. It is more common in males, and the common sites are the skull, mandible, ribs, spines and long bones particularly the femur and the humerus. The estimated incidence of EG is 3-4 per million of the population. Vertebral bone involvement is rarely seen and usually affects the vertebral body. We are reporting an unusual case of EG in a female child presented with a solitary lesion at posterior element of lumbar vertebra.

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Eosinophilic granuloma (EG) is a benign self-limiting disease, belongs to the spectrum of Langerhans' cell histiocytosis (LCH). The etiology of LCH remains unknown, although the evidence indicates that it is a clonal proliferative disorder of Langerhans cells.[1] It has also been characterized as reactive disorder, neoplastic process and a berrant immune response.[2,3] Eosinophilic granuloma is characterized by single or multiple skeletal lesions occurring predominately in children, adolescents and young adults, it account for 70% of LCH. It is more common in male, and the common sites are the skull, mandible, ribs, spines and long bones particularly the femur and the humerus.[4] The estimated incidence of EG is 3-4 per million of the population.[5] Vertebral bones involvement is rarely seen and usually affect the vertebral body. We are reporting an unusual case of EG in a female child presented with a solitary lesion at posterior element of lumbar vertebra (L2).

Case Report. An 8-year-old female child, presented with a history of fell down on her back while playing with her brother and got severe lower back pain, which resolved spontaneously over few hours. One week later the pain re-appeared and became more severe associated with stiffness of muscle, limitation of movement of back and walking difficulty. There was no history of fever and other systemic review were unremarkable. The physical examination revealed stiffness of the lower back at the lumbar region with left paravertebral tenderness at the level of the lumbar vertebra (L1 and L2). There were no neurological deficit, and the other systemic examination were unremarkable. The laboratory investigations revealed normal white blood count, hemoglobin level, platelet count, erythrocyte sedimentation rate, C-reactive protein, renal and bone profile. Radiological investigation revealed mild scoliotic attitude with osteolytic lesion of the left pedicle of

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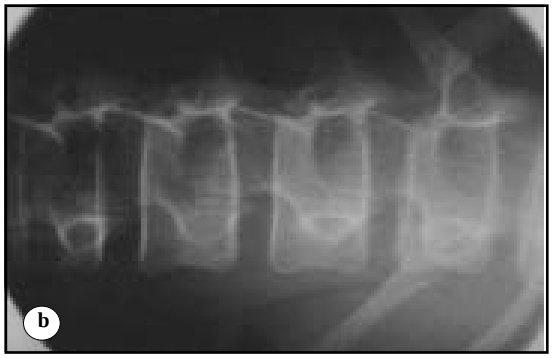
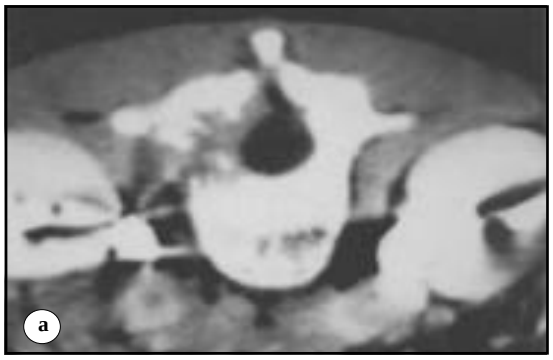


Figure 1



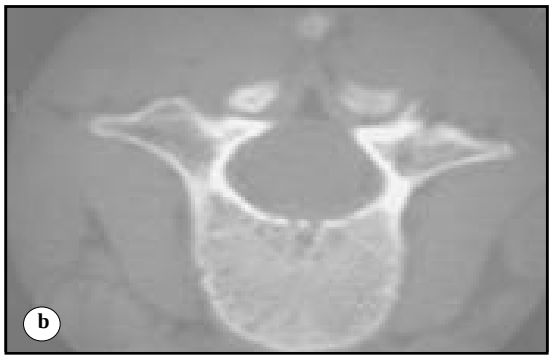


Figure 2

Table 1

Type	Frequent	Rare	Exceptional
Benign	Osteoid osteoma	Eosinophilic granuloma	Chondroma
	Osteoblastoma		
	Aneurysmal bone cyst	Osteochondroma	
Malignant		Metastases from neuroblastoma and Ewing sarcoma	Osteogenic sarcoma

Figure 1 - Radiograph of the lumbar spine **a)** anteroposterior view - showing a lumbar scoliotic attitude with left sided concavity. The center of this concavity shows an ill-defined osteolytic lesion involving the left pedicle of lumbar vertebra (L2). The outline of the left psoas muscle appears normal. **b)** left oblique view - the osteolytic lesion involves the left pedicle of L2 with loss of the cortical outline "eye of Scottie dog" and also the pars interarticularis "neck of Scottie dog".

Figure 2 - Computerized tomography of **a)** Enhanced axial (centered lumbar vertebra [L2]) - a heterogeneous enhancement with soft tissue mass extending into the spinal canal and displacing the thecal sac to the right. **b)** non-enhanced axial bone setting (centered on L2) - restitution of the bony architecture of the left pedicle.

Table 1 - Differential diagnosis of an osteolytic lesion of the posterior vertebral arch in children.

L2 and partially of the pars interarticularis (neck of the Scottie dog), the lesion is located exactly in the center of the concavity **(Figures 1a & 1b)**. Computed tomography (CT) scan, with contrast (**Figure 2a**) confirms the presence of an osteolytic lesion involving the left pedicle, pars interarticularis and partially the adjacent lamina with central bone density within the lesion "button sequestrum". The lesion shows a heterogeneous enhancement with soft tissue mass infiltrating the spinal canal and displacing the thecal sac to the right. The total body bone scan revealed a focus of increased tracer uptake at the level of the left pedicle of L2, the rest of the skeleton were normal. Open biopsy with curettage and bone grafting were performed. The patient was subsequently managed conservatively with simple analgesics and back support by body jacket and spinal brace. The patient was followed up regularly and the follow up CT scan at 6 months after the operation revealed complete healing of the lytic area. (**Figure 2b**). Over 4-years the patient remained asymptomatic.

Discussion.

Eosinophilic granuloma (EG) is a mild form of Langerhans' cell histiocytosis (LCH), which is a rare disease known until recently under the name histocytosis X.[1,2] It is predominantly affect children <12 years old, other types are Hands-Schuler Christian disease a moderate form and the severe form Letterer–Siwe disease. Bone involvement is common, the skull bone involved in 50% of cases followed by mandible, ribs, pelvis and long bones. Vertebral disease, as in this case is rarely seen, usually affect thoracic and lumbar spine, cervical spines less frequently involved. It is usually solitary lytic lesion involves vertebral body causing vertebral collapse producing 'Vertebral plana' or 'Coin-on edge' appearance. Posterior vertebral elements rarely involved, but it has been reported recently[6] similar to our case. Involvement of sternum is another rare location.[7] The clinical presentation of EG of vertebra are mainly pain and tenderness as in this case. Limp and neurological deficit were rare.[6,8] Some patients are asymptomatic and the diagnosis is usually made by radiological investigations. Posterior vertebral elements involvement are difficult to diagnose by plain radiographs, CT scan is the investigation of choice in such case. Magnetic resonance imaging (MRI) usually indicated when there is a neurological deficit or other signs of spinal cord compression. Bone scan help to detect any other bone lesions of EG or metastatic disease such as neuroblastoma. The different radiological investigations were carried out in this case revealed a single osteolytic lesion without signs of inflammatory process. The differential diagnosis of an osteolytic lesion of the posterior vertebral arch in childhood include many other disease processes as shown in **Table 1**. Definitive diagnosis in this case was made by open surgical bone biopsy; however, percutaneous needle biopsy can established the diagnosis in 90% of cases.[9]

The recommended treatment of solitary EG is surgical resection by curettage and bone grafting treatment with intralesional administration of methyl prednisolone had resulted in osseous healing,[8] other mode of treatment include chemotherapy and radiotherapy. When chemotherapy is used, prednisolone vinblastine and etoposide are the most common agents, used alone or in combination. Radiotherapy is only indicated when asymptomatic lesion involves in an accessible region such as the spine and the dose usually used is between 500-1000 rads. Spontaneous resolution of osseous EG lesions was reported.[10] The prognosis is excellent in general; however, it is related to the age of onset, younger age group had poor prognosis.[5] Recurrence of the disease had been encountered occasionally.[1-5] Regular follow up with CT scan and MRI is required to detect recurrence.

Eosinophilic granuloma should be considered in the differential diagnosis of lower back pain, and plain radiograph is recommended to demonstrate the osteolytic lesion. Finally, we recommend delaying surgical intervention to allow time for spontaneous resolution to occur in the absence of neurological deficit.

Reference

1. Willman CL, Busque L, Griffith BB, Favara BE, McClain KL, Duncan MH, et al. Langerhans' cell histiocytosis (Histiocytosis X) a clonal proliferative disease. ***N Engl J Med*** 1994; 331: 154-160.
2. Favara BE. Langerhans' cell Histiocytosis: pathobiology and pathogenesis. ***Semin Oncol*** 1991; 18: 3- 7.
3. Lieberman PH, Jones CR, Steinman RM, Erlandson RA, Smith J, Gee T, et al. Langerhans cell (eosinophilic) granulomatosis. A clinicopathologic study encompassing 50 years. ***Am J Surg Pathol*** 1996; 20: 519-552.
4. Resnick D, Niwayama G. Orthopedics: Diagnosis of bone and joint disorders. 3rd ed. Philadelphia (PA): W. B. Sauders; 1994. p. 2429-2439.
5. Velez-Yanguas MC, Warrier RP. Langerhans cell Histiocytosis. ***Orthopedic Clinic of North America*** 1996; 27: 615-623.
6. Bodart E, Nisolle JF, Maton P, Delos M, Trigaux JP, Gilliard C, et al. Limp as unusual presentation of Langerhans' cell Histiocytosis. ***Eur J Pediatr*** 1999; 158: 384-386.
7. Chiau JH, Chang TK, Hwang B, Tiu CM. Eosinophilic granuloma of sternum. Report of one case. ***Zhonghua Min Guo Xiao Er Ke Yi Xue Hui Za Zhi*** 1990; 31: 35-39.
8. Kamimura M, Kinoshita T, Itoh H, Yuzawa Y, Takahashi J, Ohtsuka K. Eosinophilic granuloma of the spine, early spontaneous disappearance of tumor detected on magnetic resonance imaging. ***J Neurosurg*** 2000; 93: 312-316.
9. Yasko AW, Fanning CV, Ayala AG, Carrasco CH, Murray JA. Percutaneous techniques for the diagnosis and treatment of localized Langerhans cell Histiocytosis (Eosinophilic granuloma of bone). ***J Bone Joint Surg Am*** 1998; 80: 219-228.
10. Oliveira M, Steinbok P, Wu J, Heran N, Cochrane D. Spontaneous resolution of calvarial eosinophilic granuloma in children. ***Pediatr Neurosurg*** 2003; 38: 247-252.