Unusual location of eosinophilic granuloma

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ABSTRACT

Eosinophilic granuloma (EG) is a well-recognized benign form of Langerhans cell histiocytosis, most commonly involving the skull bones. In this paper, we report an 8-year-old girl with EG of posterior element of vertebra; she had complete resolution with surgical curettage and bone grafting.

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Case Report

An 8-year-old female child, presented with a history of falling on her back while playing with her brother and had severe lower back pain, which resolved spontaneously over a few hours. After one week, the pain reappeared and became more severe, associated with stiffness of muscle, limitation of movement of the back and walking difficulty. There was no history of fever and other systemic review was unremarkable. The physical examination revealed stiffness of the lower back at the lumbar region with left paravertebral tenderness at the level of L2 vertebra. There was no neurological deficit, and the other systemic examination was unremarkable. The laboratory investigations revealed a 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 an osteolytic lesion of the left pedicle of 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 and 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 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 was normal. Open biopsy with curettage and bone...
Figure 1 - Radiograph of the lumbar spine
a) AP view showing a lumbar scoliotic attitude with left sided concavity. The centre of this concavity shows an ill-defined osteolytic lesion involving the left pedicle of 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 the pars interarticularis “neck of Scottie dog”.

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

<table>
<thead>
<tr>
<th>Type</th>
<th>Frequent</th>
<th>Rare</th>
<th>Exceptional</th>
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<tbody>
<tr>
<td>Benign</td>
<td>Osteoid osteoma</td>
<td>Eosinophilic granuloma</td>
<td>Chondroma</td>
</tr>
<tr>
<td></td>
<td>Osteoblastoma</td>
<td>Osteochondroma</td>
<td></td>
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<tr>
<td></td>
<td>Aneurysmal bone cyst</td>
<td></td>
<td></td>
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<tr>
<td>Malignant</td>
<td>None</td>
<td>Metastases from</td>
<td>Osteogenic sarcoma</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Neuroblastoma</td>
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<td></td>
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<td>Ewing sarcoma</td>
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Figure 2 - Enhanced axial CT (centered on L2) a) Showing an osteolytic process of the left pedicle of L2 with a small central bone density “button sequestrum”. b) Bone settings shows restitution of the bony architecture of the left pedicle (after 6 months)
graffing were performed. The patient subsequently was managed conservatively with simple analgesics and back support by body jacket and spinal brace. The patient was followed up regularly. Follow up CT scan at 6 months after the operation revealed complete healing of the lytic area (Figure 2b). Four years later the patient remained asymptomatic.

Discussion. Eosinophilic granuloma is a mild form of LCH, which is a rare disease, known until recently under the name histiocytosis X. It predominantly affects children less than 12 years old, other types are Hands-Schuler Christian disease a moderate form, and Letterer–Siwe disease the severe form.

Bone involvement is common. Fifty percent of the cases involved the skull bone, followed by mandible, ribs, pelvis and long bones. Vertebral diseases, as in this case are rarely seen, it usually affects the thoracic and lumbar spine; cervical spine is less frequently involved. Usually solitary lytic lesions involve the vertebral body, causing vertebral collapse producing "Vertebral plana" or "Coin–on edge" appearance. Posterior vertebral elements are rarely involved, but it has been reported recently and is similar to our case. Involvement of the sternum is another rare location. The clinical presentations of EG of vertebra are mainly pain and tenderness as in this case, limp and neurological deficits are rare. Some patients are asymptomatic and the diagnosis is usually determined from the radiological investigations. Posterior vertebral element involvement is difficult to diagnose by plain radiographs, CT scan is the investigation of choice in such case. Magnetic resonance imaging (MRI) usually indicates when there is a neurological deficit or other signs of spinal cord compression. Bone scan helps to detect any other bone lesions of EG or metastatic disease such as neuroblastoma. The different radiological investigations 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 includes 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 establish the diagnosis in 90% of the cases.

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, other modes 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 involve an inaccessible region such as the spine, and the dose usually used is between 500 to 1000 rads. Spontaneous resolution of osseous EG lesions have been reported. The prognosis is excellent in general, however, it is related to the age of onset, a younger age group had poor prognosis. Recurrence of the disease had been encountered occasionally. Regular follow up with CT scan and MRI is required to detect recurrence.

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

References