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Langerhans cell histiocytosis of the sacrum

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Langerhans cell histiocytosis is a rare disease with a wide spectrum of clinical presentations. It is a multisystemic disease with organ system involvement ranging from simple—where it involves only one organ—to widespread progressive disease. Although it can affect any age group, the peak incidence is between 1 and 3 years of age.

Case report

A 27-year-old man presented with an 8 months' history of back pain radiating to his leg. Imaging revealed an expansile lytic lesion in the body of the first sacral vertebra (S1) (Figs. 1-7). CT-guided biopsy was subsequently performed (Fig. 8), and histology confirmed the lesion to be Langerhans cell histiocytosis.

Discussion

Langerhans cell histiocytosis (LCH), a disease characterized by proliferation of histiocytes, affects multiple systems (1). Histiocytes are derived from bone marrow and proliferate in association with leukocytes, neutrophils, lymphocytes, plasma cells, and multinucleated giant cells. This proliferation leads to tissue infiltration and destruction (2).

The clinical features of LCH depend on the site and extent of involvement. LCH can present as a simple lesion that regresses spontaneously with time, or as a diffuse, progressive, multisystem disease (3). It can affect any body organ system. When it involves bones, it most commonly affects the skull and proximal femur (4).

LCH is a very rare disease, and accounts for fewer than 1% of all osseous lesions (5). The incidence rate of LCH is not well established, but it is estimated to be about 2-5 cases per million inhabitants per year (2). LCH can affect any age

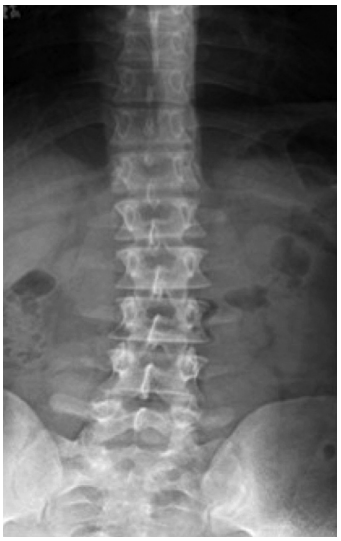

Figure 1. Plain dorsal and lumbar spine x-ray of 27-year-old male patient 8 months before CT-guided biopsy. It is not easy to find the expansile lytic sacral lesion on this image.

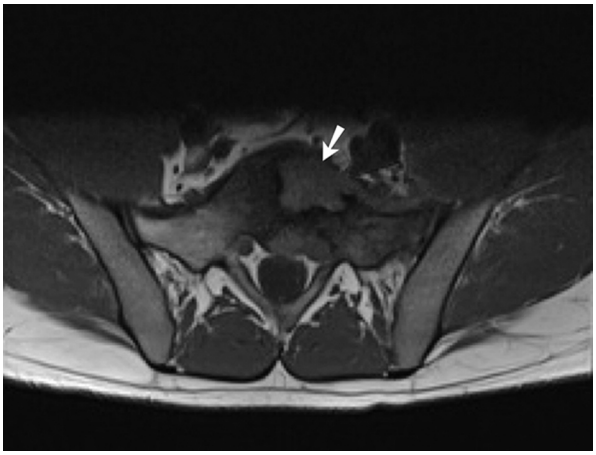

Figure 2. Plain Axial FSE T1W image of first sacral vertebra shows slightly increased signal in the sacral lesion (arrow).

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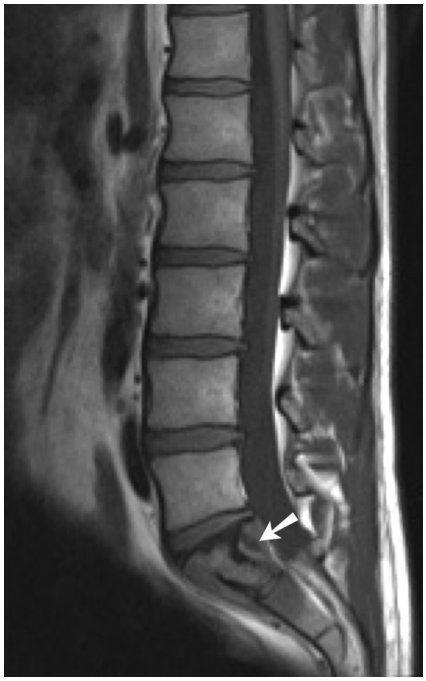

Figure 3. Sagittal FSE T1W image of the first sacral vertebra shows slightly increased signal in the lesion (arrow).

group (2), but the peak incidence is between 1 and 3 years of age (3).

Plain radiographs demonstrate solitary or multiple punched-out lesions. In the skull, this may present as a geographic pattern. When affected, long bones present with endosteal scalloping, cortical thinning, and periosteal reaction. In the spine, LCH can present as a "vertebra plana," that is, a crush fracture in which the anterior and posterior height of the vertebra is lost.

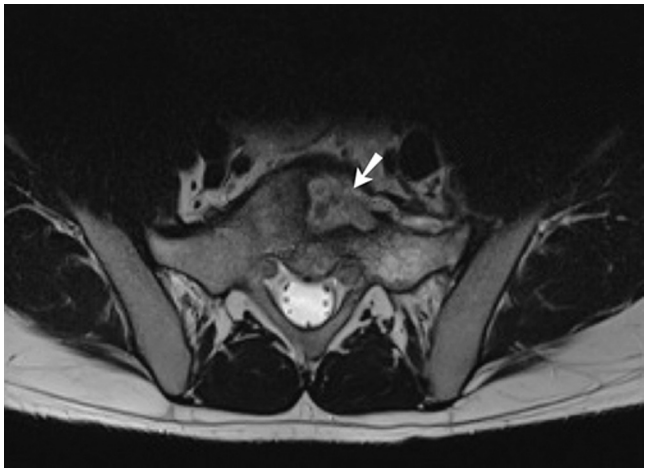

Figure 4. Axial FSW T2W image of the first sacral vertebra shows slightly increased signal and heterogeneity (arrow).

CT demonstrates the same features as those seen on plain radiographs, although with better visualization of the cortex and of soft-tissue involvement. MRI images demonstrate low signal on T1W images, isointensity or hyperintensity on T2W images, and enhancement on T1W fat-saturated images following the administration of a gadolinium-containing contrast agent. The appearance of LCH on radionuclide bone scans is quite variable, and may show increased or decreased radiotracer uptake depending on the histological appearance (3).

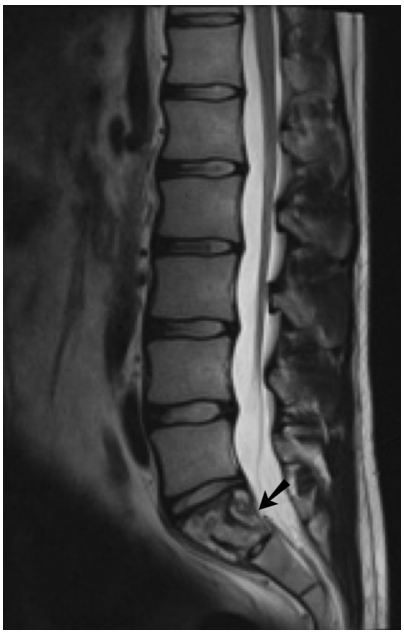

Figure 5. Sagittal FSE T2W image of the first sacral vertebra shows slightly increased signal in the lesion with heterogeneity (arrow).

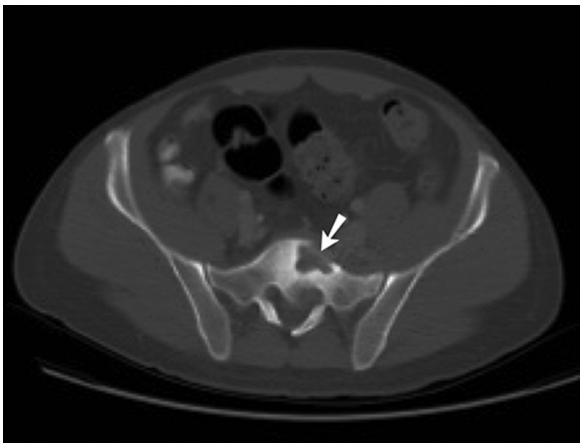

Figure 6. Axial CT scan image with bone windowing shows an expansile lytic lesion in the first sacral vertebra (arrow).

The treatment of LCH is variable and depends on the extent of the disease. Corticosteroids with or without cyclophosphamide or busulphan represent one treatment option (4). Radiotherapy represents another effective and safe treatment with minimal side effects (6).

The clinical course of osseous LCH is quite variable and difficult to predict (7). Patients with multisystem involvement have a poor prognosis, and their disease progresses despite treatment (4). A young age at presentation, throm-

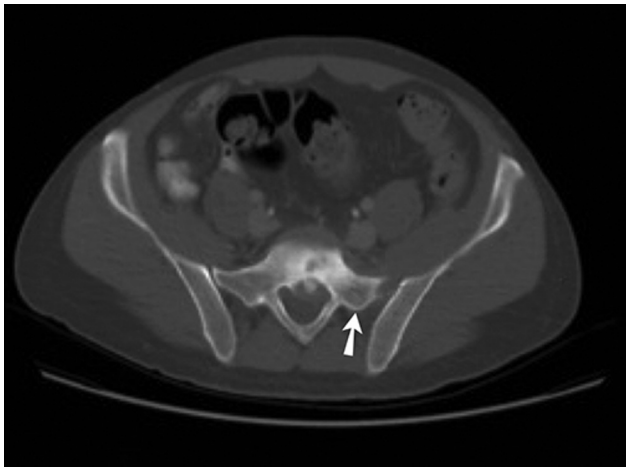

Figure 7. Axial CT scan image with bone windowing shows another focal expansile lytic lesion in the first sacral vertebra (arrow).

bocytopenia, hepatosplenomegaly, and multiosseous disease are all poor prognostic factors (7).

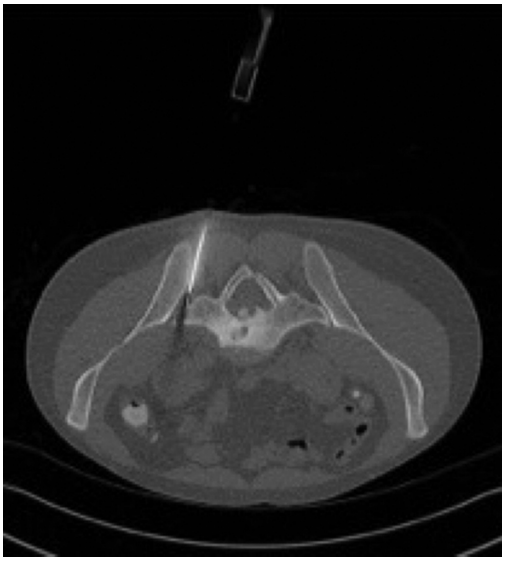

Figure 8. CT-guided biopsy: Axial image shows the trochar and needle tip in a lytic bone lesion of the sacral vertebra.

The usual differential diagnosis of lucent, expansile osseous lesions includes primary bone tumors, metastasis, osteomyelitis, leukemia, and lymphoma (3). While Langerhans cell histiocytosis is a rare condition, it should also be considered in the differential diagnosis of expansile lytic bone lesions.

References

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