An 8-year-old, previously healthy, male child presented with pain in the lower back of 3 weeks' duration. There was no history of recent fever, rash, or weight loss. Physical examination was remarkable only for decreased muscular strength in the right lower extremity. Laboratory data for routine tests were unremarkable. A plain x-ray film and computed tomographic scan of the lumbar vertebra showed a lytic lesion with irregular edges at the level of the L1 vertebra (Figure, A, arrow). A computed tomographic-guided fine-needle aspiration cytologic examination, and concurrent core biopsy of the lesion were performed.

Cytology smears from the aspirate showed numerous large cells (approximately 20 μm each), distributed in loose aggregates. These cells contained abundant cytoplasm that was better observed with Diff-Quik stain (Figure, B, arrow, original magnification ×400). The nuclei were oval, markedly convoluted, and grooved. The nucleoli were inconspicuous. Nuclear detail was more apparent with Papanicolaou stain (Figure, C, original magnification ×400). Rare multinucleated giant cells were present. Eosinophils (Figure, C, arrow) were seen in the background. Immunocytochemical stains showed strong reactivity for CD1a (Figure, D, original magnification ×200) and S100 protein (not illustrated) in the lesional cells.

Subsequent histologic examination of the needle core biopsy material showed dyscohesive sheets of large cells similar to those seen in the aforementioned cytology material. Eosinophils were prominent in the background (Figure, E, hematoxylin-eosin, original magnification ×200).

What is your diagnosis?
Pathologic Diagnosis: Unifocal Unisystem Langerhans Cell Histiocytosis (Eosinophilic Granuloma)

Langerhans cell histiocytosis (LCH) is a rare disease known until recently under the rubric “histiocytosis X.”\(^1\)\(^2\) LCH represents a wide spectrum of clinical conditions that share common cytologic and histologic features. The etiology of LCH remains unknown, although the evidence indicates that LCH is a clonal proliferative disorder of Langerhans cells.\(^3\)\(^-\)\(^5\) It has also been variously characterized as a reactive disorder, a neoplastic process, and an aberrant immune response.\(^1\)\(^-\)\(^4\) LCH affects people of all ages, but it predominantly strikes children and young adults.\(^1\)\(^-\)\(^4\)

LCH is classified under class I of histiocytosis by the Writing Group of the Histiocyte Society.\(^3\) On the basis of clinical presentation, it is subdivided into 3 groups; multisystem multifocal, multifocal unisystem, and unifocal unisystem (eosinophilic granuloma). According to the Writing Group, a presumptive diagnosis of LCH is given when classic morphologic features are seen along with 2 or more of the following: a positivity for S100 protein, adenosine triphosphatase, or alpha D-mannosidase or binding with peanut lectin; a definitive diagnosis is made only if the lesion shows positivity for CD1a, as in our case, or the presence of Birbeck granules (electron-dense cross-striations) on electron microscopy.\(^3\)

The unifocal unisystem type accounts for most LCH cases.\(^5\) A single site such as bone, skin, lymph node, or lung is usually involved, with a predilection for bone. Among bone lesions, the skull and femur have a higher rate of involvement. Vertebral disease, as in our case, is seen only rarely. Symptoms and signs, mainly pain and tenderness, vary according to the site of involvement. Some patients are asymptomatic. There are no characteristic laboratory findings. Although some patients recover spontaneously, treatment is surgical by curettage or excision. The prognosis is excellent. Occasional recurrences may be encountered.\(^1\)\(^-\)\(^5\)

In cytologic as well as histologic material, LCH cells are large and polygonal with ample cytoplasm.\(^5\)\(^-\)\(^6\) Nuclei show longitudinal grooves produced by deep invaginations of the nuclear membrane. Nucleoli are inconspicuous. Scattered eosinophils and multinucleated giant cells are frequently present. Necrosis may be observed in a bone lesion. Mitosis is uncommon. As observed in our case, LCH cells are immunoreactive for CD1a and S100 protein.

Pathological differential diagnosis of LCH includes benign histiocytes (macrophages), sinus histiocytosis with massive lymphadenopathy, mastocytosis, and monocytic leukemia. Each of these entities has its characteristic clinical, laboratory, cytologic, histologic, and immunohistochemical features. In difficult cases, S100 protein and CD1a immunoreactivity are key to a confirmed diagnosis of LCH.\(^3\)\(^,\)\(^5\)

The diagnosis of LCH on fine-needle aspiration is possible in up to 85% of the cases.\(^5\) Correlation with clinical, laboratory, and radiographic findings is always prudent.

References