



Langerhans' Cell Histiocytosis of the Thoracic Spine Diagnosed by Percutaneous Transpedicular Biopsy under Fluoroscopic Guidance in a 9-Year-Old Child

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We present the case of a 9-year-old girl who had complained of mid-back pain for two months. The plain X ray of the thoracic spine revealed vertebra plana (collapse of the vertebral body), and magnetic resonance imaging of the thoracic spine showed that the T-7 vertebral body had collapsed, and there was slight compression of the spinal cord. The patient underwent a fluoroscopy-guided percutaneous transpedicular biopsy and the histology led to a diagnosis of Langerhans' cell histiocytosis. After low-dose radiotherapy and six months immobilized in a spinal brace, the mid-back pain subsided but the plain X ray of the thoracic spine revealed collapse of the vertebral body. Therefore, we report it as a rare case and discuss the clinical significance of this disorder.

Key words: Langerhans' cell histiocytosis, percutaneously transpedicular biopsy, vertebra plana

INTRODUCTION

Langerhans' cell histiocytosis (LCH) is a rare disorder of the reticuloendothelial system and is characterized by a proliferation of cells causing local or systemic effects¹. It can affect patients of any age, but primarily affects children and young adults². The clinical symptoms of LCH vary in terms of the site and the extent of involvement. In addition, the symptoms may be focal or systemic^{3,4}.

Its primary lesion is usually in bone, but it may be found in the skin, gingival, salivary glands, lung, liver or lymph nodes⁵. The skull, ribs and long bones are frequent sites for the skeletal lesions⁶. Spinal involvement is less frequent, but is the most common cause of vertebra plana in children⁷.

We report the case of a 9-year-old child with LCH of the T-spine and a slight compression of the spinal cord. The disease was diagnosed using a fluoroscopy-guidance percutaneous transpedicular biopsy. We also review the literature regarding this disease.

CASE REPORT

For two months, a 9-year-old girl complained of a dull and deep pain in her mid-back region, and that the pain was severe during extension or flexion of the trunk. She had no pain elsewhere and no history of trauma, fever, or night sweats. In addition, she had no past medical history. Initially, she was treated by her general practitioner but analgetic treatment showed no effect. As the symptoms persisted, she visited our clinic.

The physical examination revealed local tenderness over the mid-thoracic region and a mild humpback. The neurological examination revealed no numbness, weakness or paresthesia. The plain X ray of the T-spine showed a collapsed T-7 vertebral body (vertebra plana) (Figs 1a & 1b). The magnetic resonance images of the T-spine showed collapse of the T-7 vertebral body altogether, with paraspinal and epidural soft-tissue mass enhanced by gadolinium diethylenetriamine pentaacetic acid (Gd-DTPA). This extended the vertebral body height, whether in the upward or downward direction, and the spinal curvature became a kyphosis that slightly compressed the spinal cord (Figs 2a & 2b). The whole-body bone scan revealed an increased uptake over T-7. The extensive skeletal survey did not reveal other bone lesions. Laboratory evaluation, including complete cell count, liver function tests, and alkaline phosphatase, were all within normal ranges.

The patient underwent a percutaneous transpedicular biopsy of T-7 under fluoroscopic guidance, and diagnostic tissue was obtained from the vertebral body lesion via a

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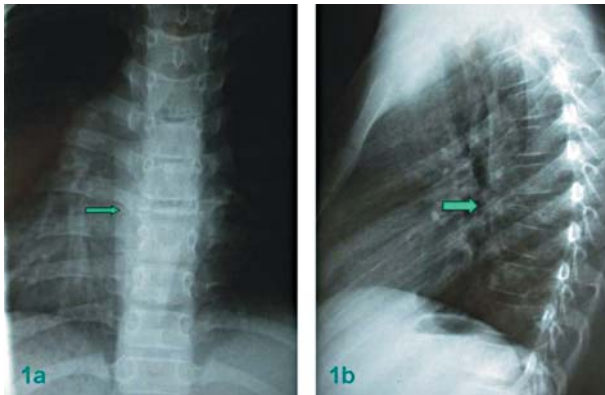


Fig. 1 (a) Anteroposterior, (b)lateral plain radiographs of the T-spine showed vertebral plana of T7 (arrow).

bilateral transpedicular biopsy. The histological examination revealed a picture of LCH with hemorrhaging, and mixed inflammatory cells and histiocytes infiltration that showed microscopic nuclear grooves and kidney-shaped nuclear contours (Fig. 3a). Langerhans' cells express a membrane-based immunoreactivity for CD1a (Fig. 3b).

After confirming the clinical diagnosis, a six-day course of low-dose radiotherapy (150 cGy/day; total 900 cGy) was performed and six months of immobilization with a spinal brace suggested. After a series of treatments, the symptoms subsided and no progressive lesion was found. At the four-year follow-up, the plain radiograph of the T-spine did not show any reconstitution or change in the height of the T-7 vertebral body (Figs 4a & 4b), but the symptoms were absent.

DISCUSSION

The prevalence of LCH is 1:2,000,000/year, and usually presents as a solitary painful lesion in children between 3 and 10 years of age^{8,9}. Up to 90% of the lesions occur in bone, and the incidence of spinal involvement is 7 to 15%^{3,10}. If only children are noted, the incidence rate can rise to 20 to 30%¹¹.

The symptoms and signs of LCH can be presented with pain, limited motion, local inflammatory signs, or mild fever¹². LCH of the spine normally only presents with localized neck or back pain^{11,13}. Neurologic symptoms are extremely rare and are usually limited to mild paresthesias or radicular pain^{2,5,8,11}.

Some lesions of LCH are often asymptomatic and accidentally found on skeletal studies, such as radiographs or bone scans⁸. The classic radiographic findings of LCH

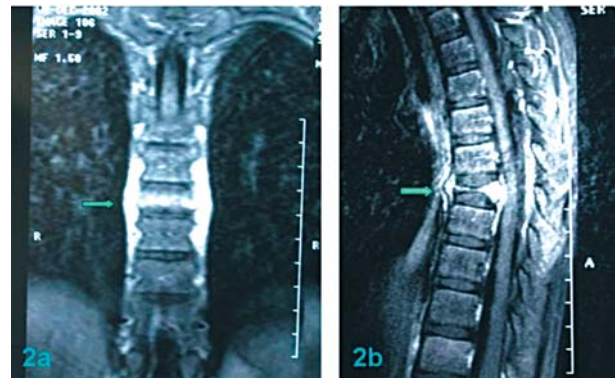


Fig. 2 (a) Coronal, (b) sagittal magnetic resonance image of T-spine showed collapse of T-7 vertebral body together along with paraspinal and epidural soft tissue mass (arrow) enhanced by Gd-DTPA which extend a vertebral body height no matter in upward and downward, and the spinal curvature becomes kyphosis that compressed spinal cord slightly.

of the spine include vertebral collapse (vertebra plana), which causes symmetrically flattened vertebral bodies, preservation of disc spaces, and lack of extraspinal spread and soft-tissue mass^{11,12,13}. MRIs of the spine may be useful to demonstrate clearly the preservation of disc space that helps discriminate the lesion from nonspecific or tuberculous vertebral infection, and to rule out soft-tissue masses or aggressive lesions with epidural soft-tissue extensions^{3,8}. MRI is also useful for evaluating spinal cord compression and for planning biopsies of the spine if involvement is limited to this region¹⁶.

LCH of the spine may present a focal osteolytic vertebral lesion, with or without collapse of the vertebral body^{12,16}. Patients with these lesions should be carefully differentiated because other disease processes, such as aneurismal bone cysts, osteogenesis imperfecta, Ewing's sarcoma, lymphoma, leukemia, malignant solid tumors, neuroblastoma, infection, hemangioma, myeloma, osteogenic sarcoma, metastatic lesions, and Gaucher's disease, may cause similar findings^{14,16}. For vertebra plana, the diagnosis, including metastatic lesions, lymphoma leukemia, or trauma causing vertebral body collapse, should be differentiated¹⁶.

A work-up for vertebra plana must include chest X rays and laboratory studies, along with simple radiographs, MRI of the spine and whole-body bone scans⁸. These studies can rule out other lesions, except of the spine, and can confirm the extent of the disease. When an isolated vertebral lesion is noted, the necessity for a vertebral biopsy is controversial¹⁷. Some authors suggest that spinal

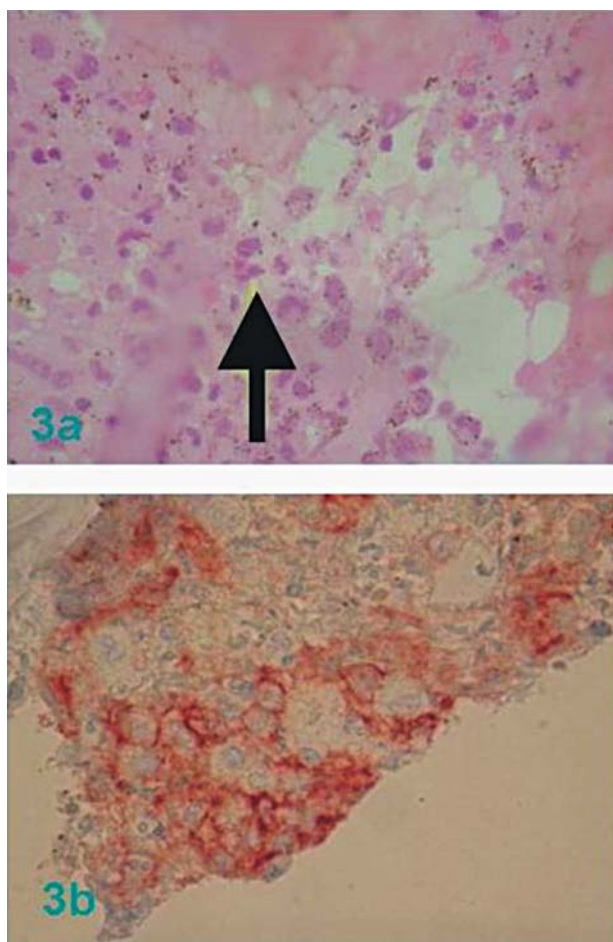


Fig. 3 (a) The pathology revealed hemorrhage and mixed inflammatory cells and histiocytes infiltration that showed nuclear grooves and kidney-shaped nuclear contour. (b) Langerhans' cells express membrane based immunoreactivity for CD1a.

biopsies are unnecessary in typical vertebral lesions that have classic radiologic characteristics, mild to moderate local pain, typical manifestations, and a benign and nonprogressive clinical course. This is because the apophyseal plates of the vertebra, nerves or vessels may be damaged during the biopsy procedure^{17,18}. However, a tissue diagnosis is necessary when there is a suspect malignancy or if there are any atypical manifestations and clinical courses, such as a soft-tissue mass, disc-space involvement, significantly persistent pain, and neurologic symptoms^{12,14,17}. When extravertebral involvement is found, a biopsy at those sites provides an appropriate confirmation of the diagnosis. Therefore, to establish a histological diagnosis, a corporal biopsy through the pedicle would be imperative if no other sites were available¹⁷. The biopsy of

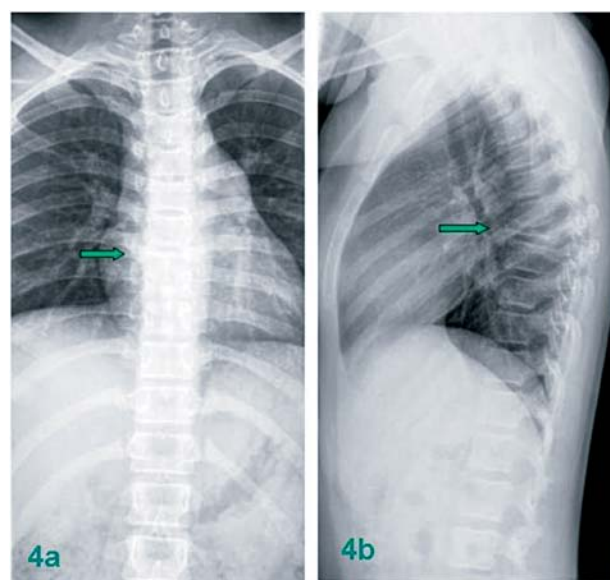


Fig. 4 (a) Anteroposterior, (b) lateral plain radiographs of the T spine, 4 years following diagnosis, did not show reconstitution or change of T7 (arrow) vertebral body height.

the vertebral body via the pedicles could decrease the risk of neurovascular damage compared with extrapedicular procedures¹⁸. The percutaneous needle biopsy using computed tomography-guided fluoroscopy is a better choice than an open biopsy; however, the technique for a transpedicular biopsy would be difficult to perform in children because the diameter of their pedicles is much smaller than in adults. It is technically demanding, and frequently fails to provide an adequate specimen to establish a histological diagnosis¹⁷.

Once the diagnosis is established, adequate treatments may include local therapy, surgery, and radiotherapy or chemotherapy based on the extent of the disease, but these are still controversial^{16,19-21}. The treatment goals are spinal stability, preservation of neurologic function and eradication of the lesion²².

For local treatment for LCH of the spine, a spinal brace or collar has been suggested when there is no neurologic deficit^{18,20,23}. This bracing can last from a few months to two to four years. In these stable lesions, immobilization by spinal support is recommended until the clinical and radiologic healing processes are evident^{24,25}. This conservative treatment allows reduction in the mechanical load on the anterior spine and restoration of vertebral height^{19,20}.

The indications of spinal fusion for LCH are rare and mostly limited to significant spinal instability or a neuro-

logic deficit due to spinal cord compression from vertebral collapse or an extraosseous extension of the disease^{11,20,27}. This surgery, except for the biopsy, is not indicated for children with LCH of the spine¹¹. Spinal decompression and short fusion are performed to allow recovery of neural function and to avoid spinal instability^{22,27}.

Radiotherapy is only suggested when immobilization fails to relieve the symptoms, such as nonhealing or progressive spinal lesions, severe pain, and neurologic deficits^{18,20,24}. When a single vertebral lesion causes neural compression and irreversible damage, low-dose radiation therapy (500-1500 cGy) has been used^{3,20}. This limited dosage was proposed in the current protocol, and provides pain relief without limiting the potential for spinal growth^{18,20,24}. In general, this is a safe and effective treatment to achieve local control and to prevent the spread of disease²⁰. On the contrary, other studies have suggested that radiotherapy in children should be avoided unless the disease is unremitting. It may increase the potential of malignant change in adjacent tissues and there is a risk of damaging the vertebral growth plates²⁷. Our case, presenting LCH with a collapse of the T-7 vertebral body, together with soft-tissue mass extension, was treated with spinal brace immobilization and low-dose radiotherapy. The result showed a recovery and a complete resolution of the symptoms.

Chemotherapy is used for multisite bone lesions and multiple system involvements, but remains controversial. The most commonly used chemotherapeutic agents are vinblastine, etoposide (VP-16), prednisone, methotrexate, and 6-mercaptopurine^{3,18,20,24}. Recently, several studies reported good outcomes for children following chemotherapy. Tan et al.²⁷ treated four children with cervical LCH with prednisone, and they all completely recovered and retained normal function of the cervical spine. Tanaka et al.²⁸ treated two children with LCH of the atlas with prednisone and methotrexate, and the results showed that this was safe and effective. These studies concluded that chemotherapy now tends to be less intensive, and that this regimen results in satisfactory control of this more aggressive condition without marked complications^{8,27,28}.

In conclusion, the work-up of LCH should include history taking, physical examination, detailed image survey, and tissue diagnosis. When there is no neurologic deficit, conservative treatment with immobilization is suggested. There is still no clear evidence of the effects of radiotherapy or chemotherapy, but several recent studies following these managements showed good results. Surgery intervention is recommended for significant spinal instability or neurologic deficit.

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