Langerhan’s Cell Histiocytosis of the Lumbar Spine during Pregnancy: A Rare Case with Literature Review

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ABSTRACT

Background: Langerhan’s cell histiocytosis (LCH), previously known as histiocytosis X, is a reactive proliferative dendritic cells of unknown pathogenesis characterized by the proliferation of Langerhan’s cells and is extremely rare in the lumbar spines of adults. This condition is most common among young males under the age of 15 years old (with a peak incidence at 2-4 years old), and the most frequent site of these osteolytic bony lesions of the spine is the thoracic region.

Purpose: To highlight an interesting and rare presentation for Langerhans cell histiocytosis of the spinal cord in pregnant woman.

Study design: This is a case report of a single patient in whom a Langerhans cell histiocytosis was resected from the lumbar spine in pregnant woman with return to normal functioning.

Patient sample: A 26-year-old pregnant woman at 20 to 22 weeks presenting with acute cauda equina syndrome, a 1-month history of pain and numbness and paraparese of right limb had gradually progressed to involve all the lower limbs.

Outcome measures: Frankel grading of neural function and Visual Analogue Score are included to evaluate the therapeutic efficiency.

Methods: Magnetic resonance imaging revealed the widespread involvement of an extradural contrast-enhancing mass in the lumbar spine of L2-L5.

Results: The patient underwent decompression and surgical resection of the tumor in a three-quarters prone position, fetal heart monitoring was performed by our obstetrician; there was no fetal distress during the surgery. The diagnosis was confirmed by histological analysis. She entered spontaneous labor at 36 to 37 weeks and birthed a baby weighing 3000 gm. The child had gradually progressed to involve all the lower limbs.

Conclusion: Our case’s unique presentation involves LCH in an adult pregnant patient at the lumbar spine with no osteolytic activity. The management of pregnant women and maintenance of fetal well-being should be coordinated among the spinal neurosurgeon, obstetrician and anesthesiologist.

Keywords: Langerhan’s cell histiocytosis, Pregnancy, Cauda equina syndrome, Three quarter prone position.


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INTRODUCTION

Langerhan’s cell histiocytosis (LCH) or histiocytosis X is a rare disease complex caused by large numbers of Langerhan’s cells (LCs), lymphocytes, plasma cells, eosinophils, and neutrophils that generate local or systemic effects. The accumulation of these cells leads to classic lytic bone lesions, skin rashes, lymphadenopathy, splenomegaly, and organ dysfunction of the pituitary gland, lung, liver and bone marrow. LCH is a disease of reactive proliferative dendritic cells with unknown etiology. The most frequent sites of these bony lesions are the skull (particularly in the parietal and frontal bone), mandible, femur, pelvis and spine.6,7,8,10,15,16 LCH can affect patients of any age, but males under 15 years old are more commonly affected.3,11,19 The general estimated incidence falls between 0.2 and 2.0 cases per 100,000 children under 15 years of age (with a peak incidence at ages 2-4 years old).10,20

In 95% of cases, LCH of the spine presents as a focal osteolytic vertebral lesion, with or without the collapse of the vertebral body. Regarding location, LCH at the level of spinal involvement varies among different ages. Whereas it often occurs in the thoracic spine (54%) in children, in adults, 47% of the reported cases involved the cervical spine. A further 20% of the reported cases affected the lumbar spine with no osteolytic involvement.20 The involvement of a single organ, such as the bone, skin or lymph nodes, usually suggests a favorable prognosis, with the patient requiring minimal or even no treatment. In cases of multiple organ involvement (multisystem disease), the patient may be at risk for a poor outcome, including a 10 to 20% mortality and a 50% risk of life-imparing morbidity.1,6,10
CASE REPORT

A 26-year-old housewife at 20 to 22 weeks of her gestational pregnancy presented with a 5-month history of progressive back pain associated with tingling, numbness and paraparesis in both lower limbs, inability to pass urine and constipation lasting 3 weeks (cauda equina syndrome). She was first evaluated at a primary health center and then was treated with analgesics. There was no history of spinal trauma. She also reported generalized weakness, weight loss and low-grade fever. There was no loss of sphincter control, but she required support to walk. She denied any contact history with a tuberculosis patient. Family and medical history were also unremarkable. The physical examination revealed tenderness over the spinous processes at the lumbar vertebrae 2-5 (L2-5). Sensation was reduced over the right side at the L2-L5 level. Voluntary power in all muscle groups was fairly normal. Perirectal examination revealed no abnormal findings. No lymph adenopathy was found.

Magnetic resonance imaging (MRI) is our choice of modality with which to diagnose spine disorders in pregnancy; the patient’s MRI with contrast revealed a well-defined extradural enhancing mass lesion at the L2-L5 level (Fig. 1). All laboratory findings (blood counts and differentials, serum electrolytes, liver and thyroid function test) were within normal limits. Exceptions included a slight elevation of the total white cells. Because a neurological deficit spinal cord compression were noted, in addition to accompanying clinical and imaging findings, the decision to operate was made immediately. Due to her pregnant state, we decided to operate upon her at the three-quarter prone position (Fig. 2), which allowed us to maintain the blood flow and welfare of the fetus during surgery.

A L2-5 decompressive laminectomy was performed. We found that the tumor wall was easy to separate from the dura, and the lesion was well circumscribed, firm and moderately vascular. The tumor was removed in toto (Fig. 3), and the cord was not tense at the end of procedures. The patient was extubated immediately after the operation, with no neurological deficits. Before, in between and after the surgery, fetal heart monitoring was performed by our obstetrician; there was no fetal distress during the surgery.

The histopathological examination of the specimen showed an infiltration of lymphocytes, eosinophils, multinucleated giant cells and Langerhan’s histiocytes (Fig. 4), based on which a final diagnosis of LCH was made. Her postoperative MRI reflected the complete excision. The postoperative course was smooth, and complete pain relief was achieved. After the operation, she was ambulatory with walking stick, and her bowel and bladder symptoms had improved. Further examination has revealed an improvement of her paraparesis from grade II to grade IV, sensory impairment 1 week after surgery, and brisk reflexes with no clonus. She had to strain during micturition and required laxatives for almost 14 days. The patient was subjected to neurorehabilitation with the help of a physiotherapist and occupational therapist. Her lower limb power improved progressively, and by postoperative day 14, she was able to walk without a supportive device.
including eosinophilic granuloma, Hand-Schuller Christian syndrome and Letterer-Sewe disease; all of these terms have been replaced at present by the term LCH, as agreed upon by the Histiocytosis Society in 1987.20

LCs are nonpigmented bone marrow-derived dendritic cells. They function as antigen-presenting cells and are vital to the epidermal component of the immune system. LCs share certain properties with monocytes and macrophages but differ from other histiocytes due to their pale clefted nuclei, abundant pale eosinophilic cytoplasm, and pentalaminar Birbeck granules. LCH manifests clinically in various ways, ranging from solitary bone lesions to polysystemic disease. The term histiocyte was originally used to designate a large cell normally found in the lymph nodes and spleen. Such cells were morphologically nonspecific but had a voluminous, granulated cytoplasm; some contained ingested particles and one or more round to irregularly shaped pale nuclei. Subsequently, the term histiocyte was taken as synonymous with the fully differentiated end cells of the monocyte/macrophage lineage, including the spleen sinusoidal macrophages, the lung alveolar macrophages, and the Kupffer cells in the liver. Still more recently, the term has been extended to include another group of cells comprised of the LCs in the skin, the interdigitating dendritic cells in the lymph nodes, thymus, and spleen, and the dendritic reticular cells found principally in the germinal centers of lymph nodes. Consequently, the term histiocyte, in its current usage, includes cells of both the monocyte-macrophage series and the LCs dendritic cell series. Sometimes the cellular system that encompasses both macrophages and LCs is called the mononuclear phagocyte and immunoregulatory effector system (M-PIRE).5,17,18 LCH resembles a malignant disease due to its progressive, invasive growth and dissemination.

Depending on its classification and clinical course, LCH can be treated with many different therapeutic strategies, including observation, local treatment, immunomodulation, radiation, chemotherapy and allogenic stem cell transplantation. For progressive multisystemic disease, various combinations of three- or four-agent chemotherapy regimens are used to treat higher risk lymphoma. Stem cell transplantation also benefits patients with bone marrow involvement.4,8,12-14,20

The surgical options for spinal disease during pregnancy are challenging and require a carefully selected strategic approach.5 During the pregnancy, the blood flow increases at a low impedance into the uteroplacental circulation, reaching up to 500 ml/min at term when measured in the supine position and even higher when measured in the left lateral decubitus position. In the supine position, the uterus may obstruct the abdominal aorta, iliac arteries and inferior vena cava. This compression is relieved by shifting the patient to the left lateral decubitus position.18 The three-quarter prone
position is recommended for back surgery during pregnancy because it allows blood to flow to the uteroplacental region during surgery while still providing clear visibility to the neurosurgeon. Among the other benefits is a lower risk of venous air embolism, but this approach can also increase the risks of bleeding, brachial plexus injury, pressure sores and macroglossia.

Langerhan’s cell histiocytosis in pregnancy remains a rare case of spinal lesions, and our case was unique in several aspects: (1) Adult female pregnancy patient, making fetal blood flow and welfare during surgery a high priority; (2) Presence of cauda equina syndrome, which is the main indicator for urgent treatment during pregnancy; (3) Involvement of the lumbal spine posterior element; and (4) Absence of osteolytic activity. Ultimately, we sustained the pregnancy after surgery; at gestational age 36 to 37 weeks, a healthy baby was born at full term via normal spontaneous vaginal delivery.

**CONCLUSION**

Surgical options should be offered in the presence of neurological deficits. To provide optimal results for both the pregnant woman and her fetus, a team of a spinal neurosurgeon, obstetrician and anesthesiologist is necessary. The choice of a surgical approach is mandatory for pregnant woman to maintain fetal blood flow and welfare during surgery. Patient should be observed closely after the surgical procedures and the baby’s delivery. Relapse has not been observed as of 1 year after surgery. Radio- and chemotherapy can be offered in the cases of recurrent disease.

**CONSENT**

Informed consent was obtained from the patient for publication of this case report and any accompanying images. Her family was present at the time.

**AUTHORS’ CONTRIBUTIONS**

RHD, FY, SEO, AYP, ABS, MZA and AF had examined, treated, observed, and followed up the subject of this case. RHD, FY and SEO performed the operation on the patient. AYP examined, treated, observed, and followed up both the pregnant patient and her baby well-being. All authors participated in writing the manuscript. All authors has read and approved of the final manuscript.

**REFERENCES**