10.5005/jp-journals-10039-0000

CASE REPORT

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

[1]Rully H Dahlan, [2]Farid Yudoyono, [3]Sevline E Ompusunggu, [4]Akhmad Y Pramatirta, [5]Agung B Sutiono
[6]Ahmad Faried, [7]Muhammad Z Arifin

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 lumbal 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 began crying immediately and had Apgar scores of 8 and 10 in the 1st and 5th minutes, respectively. No residual disease or recurrence was noted at follow-up longer than 1 year.

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, Qauda equina syndrome, Three quarter prone position.

How to cite this article: Dahlan RH, Yudoyono F, Ompusunggu SE, Pramatirta AY, Sutiono AB, Faried A, Arifin MZ. Langerhan's Cell Histiocytosis of the Lumbal Spine during Pregnancy: A Rare Case with Literature Review. J Spinal Surg 2014;1(2):0-0.

Source of support: Nil

Conflict of interest: None

[1-7].....

[1-3]Department of Neurosurgery, Faculty of Medicine, Spine Research Working Group, Universitas Padjadjaran, Dr Hasan Sadikin Hospital, Bandung, Indonesia

[4]Department of Obstetrics and Gynecology, Faculty of Medicine, Universitas Padjadjaran, Dr Hasan Sadikin Hospital Bandung, Indonesia

[5-7]Department of Neurosurgery, Faculty of Medicine, Universitas Padjadjaran, Dr Hasan Sadikin Hospital, Bandung, Indonesia

Corresponding Author: Farid Yudoyono, Spine Research Laboratory, Department of Neurosurgery, Faculty of Medicine Universitas Padjadjaran, Dr Hasan Sadikin Hospital, Jl Pasteur No. 38, Bandung 40161, Indonesia, e-mail: faridense@gmail.com

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.[6,7,9,10,15,16] 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 lumbal 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-impairing morbidity.[1,6,10]