

Langerhans Cell Histiocytosis on Fine Needle Aspiration Cytology

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Langerhans cell histiocytosis (LCH) is a rare disease characterized by clonal proliferation of Langerhans cells.

We report a case of a five-year-old female presented with back pain and scoliosis. Radiological studies revealed lytic swelling with paravertebral soft tissue extension. Reports on cytology of paravertebral soft tissue extension of LCH are rare. CT scan guided fine needle aspiration from the spine was performed. A diagnosis of LCH was made based on the cytological and radiological findings. Subsequent histology and immunohistochemical stains also confirmed the diagnosis of LCH.

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LCH is a rare disease affecting predominantly children and young adults. LCH is considered a neoplasm of the mononuclear phagocytic immunoregulatory system. It is characterized by clonal proliferation of Langerhans cells (LCs). LCH has three categories: eosinophilic granuloma (solitary and most common), Hand-Schuller-Christian disease (multifocal unisystem form) and Letterer-Siwe syndrome (multifocal multisystem form)^{1,2}. The diagnostic cytological features of LCH include high cellularity composed of sheets of characteristic Langerhans cell with large kidney-shaped nuclei admixed with eosinophils, neutrophils, macrophages, and lymphocytes.

The cytological features facilitate rapid and accurate cytological diagnosis, avoid unnecessary biopsy and offer a guide for an early and appropriate management³. Immunohistochemistry if available could be performed on cell block.

The aim of this study is to highlight the cytological features of LCH and early management.

THE CASE

A five-year-old Bahraini female presented with four weeks history of back pain and scoliosis of two weeks duration. There was no history of fever, lower limb weakness, sweating, loss of appetite or loss of weight.

The patient's examination was normal except tilting to the left side. There were no neurological signs of sensory or motor deficit, no hepatosplenomegaly or lymphadenopathy. Plain X-ray revealed significant lateral compression of the T10. MRI revealed compression of T10 vertebral body with associated paravertebral and paraspinal soft tissue mass extending from right T9 down to T11 (approximately 2.5 cm). CT scan guided fine-needle aspiration (FNA) was performed and revealed Langerhans cells histiocytosis. Subsequent biopsy also confirmed the diagnosis of LCH and immunohistochemical stains (CD1a and S100) were positive.

Langerhans cells are large cells with prominent nuclear indentations, grooves, kidney-shaped coffee bean nuclei, fine chromatin and abundant pale cytoplasm, see figures 1 and 2². Spinal biopsy and further immunohistochemical stains were performed (including CD1a, CD68, and S-100) which confirmed the diagnosis of LCH, see figures 3 and 4.

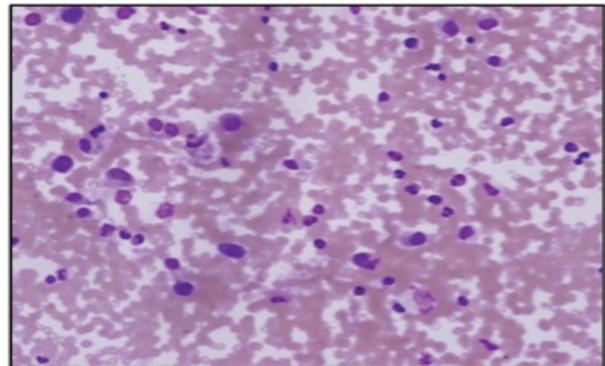


Figure 1: FNA Smear from Spine Showing Langerhans Cells Admixed with Neutrophils, Lymphocytes and Eosinophils (Giemsa Stain)

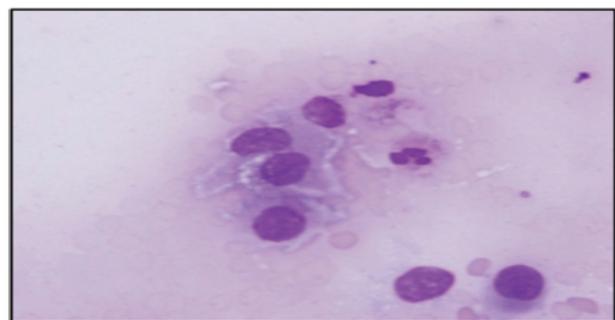


Figure 2: Langerhans Cells with Moderate to Abundant Cytoplasm and Prominent Nuclear Grooves (Giemsa Stain)

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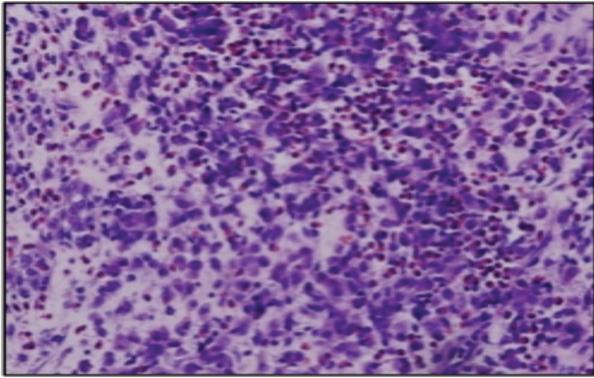


Figure 3: Sheets of Langerhans Cells Admixed with Numerous Eosinophils (Hematoxylin and Eosin Stain)

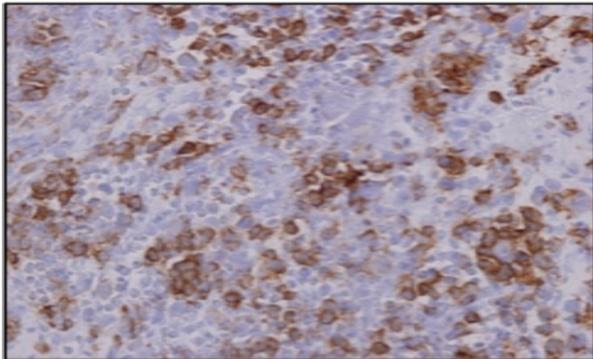


Figure 4: Langerhans Cells Showing Strong Membrane Immunoreactivity for CD1a (Immunoperoxidase)

DISCUSSION

LCH is a rare disease; the estimated annual incidence ranges from 0.5 to 5.4 cases per million². LCH may occur at any age; the majority occurs in children. There is no significant gender difference. Accurate cytological diagnosis of LCH could be achieved in combination with clinical and radiological features. The cytological features of LCH are numerous Langerhans cells admixed with many eosinophils, neutrophils, macrophages and lymphocytes.

The course of LCH is often unpredictable, varying from spontaneous regression to aggressive progression and organ dysfunction and potentially life-threatening complications. A patient with LCH needs careful staging of their disease to ensure that these are not part of a more extensive process. FNA could be used to establish the extent of the disease or recurrence of LCH. Multisystemic LCH has a worse prognosis compared to mono focal disease; multisystemic LCH has 10% to 30% mortality risk and 50% risk of life-impairing morbidity^{4,6}. For aggressive diseases, multiple chemotherapeutic regimens have been tried.

Treatment of LCH of the spine could be by bed rest, immobilization with cast and brace, hormone, chemotherapy, radiation therapy and surgery⁷⁻⁹. The treatment of LCH of the spine in children with soft tissue extension and neurological deficit is controversial¹⁰. Unfortunately, there was no clinical assessment or follow-up of the patient since discharge from the hospital.

CONCLUSION

Awareness of the cytological features of LCH is valuable for early management. We presented a rare case of a five-year-old female with LCH presented with back pain and scoliosis.

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