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Editorial

Cytology is a powerful tool for diagnosis of Langerhans cell hyperplasia/sarcoma

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EDITORIAL

Langerhans cells (LCs) derived from the CD34+ hematopoietic precursor cells of the bone marrow are mobile, dendritic, antigen-presenting cells. The characteristic cytoplasmic marker, the Birbeck granules, is found ultrastructually in 10-week-old embryo. The expression of a more characteristic immunohistochemical and immunocytochemical marker, CD1a, is completed by 12 to 13 weeks of estimated gestational age. Dr. Paul Langerhans, who also discovered pancreatic islets, first described Langerhans cells (LCs) in 1868, after making it visible by means of a gold chloride technique.

There are two types of tumors of LCs. They are classified into Langerhans cell histiocytosis (LCH) and Langerhans cell sarcoma (LCS). LCH is the commonest disorder of the phagocytic system. The term LCH was introduced as an alternative to histiocytosis X by Dr. Nezel of in 1973. Eosinophilic Granuloma, Hand-Schuller-Christian Disease and Letterer-Siwe Syndrome are the three conditions that are believed to represent different expressions of the same disorder, now known as LCH. Due to the relative rarity of the disease, estimating 0.5-5.4 cases per million persons as an annual incidence, its diagnosis is often delayed. Also, many questions regarding etiology, pathogenesis and therapy are unanswered. LCH is characterized by clonal proliferation of pathogenic LCs. LCH occurs at any age, although the majority of the cases are diagnosed in children (newborn to 15 years). There is no significant gender difference. The clinical spectrum varies from a solitary lesion to multifocal unisystem to multisystem lesions with related symptoms.

Recently several reports have been published in which fine needle aspiration (FNA) cytology, endoscopic ultrasound-guided FNA (EUS-FNA) cytology or cell block with immunocytochemistry is helpful in making a rapid and correct diagnosis of LCH and LCS. The authors suggested that highlighting diagnostic cytological features will help the pathologist in rendering a rapid and accurate cytological diagnosis, avoid unnecessary biopsy, and guide for an early and appropriate management. The classical cytologic findings include high cellularity composed of sheets/clusters and many isolated LCs admixed with numerous inflammatory cells, including eosinophils, neutrophils, lymphocytes, plasma cells, multinucleated giant cells and macrophages. The key to the diagnosis is to identify the LC through its characteristic features, such as nuclear grooves and nuclear pseudo-inclusions.

According to the WHO classification, LC tumors have been difficult to differentiate from other malignancies, such as non-Hodgkin lymphoma, melanoma, sarcoma and undifferentiated carcinomas due to their similarities. Immunostaining for CD1a, S-100, CD163, and CD207 (langerin) is useful for diagnosis of LC tumor. Although diagnosis is difficult due to the similarity of cytological and histological features, mitotic activity would differentiate between LCH (variable) and LCS (>50 per 10 high-power fields). BRAF mutations in histiocytic proliferative diseases are known to be restricted to LC lesions of LC, the positive rate of BRAF mutations in LCS is lower than that in LCH.

Although neoplastic cells suggesting a histiocytic neoplasm are observed in cytology, it is hard to diagnose LCH or LCS only by cytological findings. Immunocytochemistry using FNA, EUS-FNA or cell block materials may be useful for an accurate preoperative diagnosis. Additionally, cytology can be very useful for staging and follow-up of the LC neoplasm.