Langerhans Cell Histiocytosis of the Orbit Diagnosed by Fine Needle Aspiration: Two Case Reports

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Abstract

Purpose: Fine needle aspiration (FNA) cytologic findings of Langerhans cell histiocytosis (LCH) have been well described, but using this method in the diagnosis of orbital lesions is a recent experience. We hereby report two cases of orbital bone LCH diagnosed by FNA that later was confirmed by routine H&E histopathology and immunohistochemistry (IHC) methods.

Case reports: The first case was a one-year-old boy with a left upper lid mass. Radiologic findings were suspicious for malignancy. The other case was a three-year-old boy with right lower lid edema. Radiographic study revealed a mass with peripheral condensation and orbital bone defect. FNA of lesions in both patients showed a mixed cell population of eosinophils, neutrophils and lymphocytes admixed with neoplastic histiocytes with folded nuclei. Consequently, the cytological diagnosis was "Langerhans cell histiocytosis" that was confirmed by the routine histopathologic examination. In both cases staining for CD1a by IHC method was also performed that showed positive reaction.

Conclusion: LCH should be considered in differential diagnosis of primary orbital bone lesions specially in childhood. FNA method may be useful in the diagnosis of orbital lesions suspicious for LCH.

Keywords: Langerhans Cell Histiocytosis, Orbit, Fine Needle Aspiration

Introduction

Eosinophilic granuloma, Hand-Schuller-Christian disease and Letterer-Siwe disease are three disorders classified within the more recently general entity known as Langerhans cell histiocytosis (LCH). LCH accounts for less than 1% of all orbital tumors, although orbital involvement in LCH is not uncommon.1,2 Early diagnosis of orbital eosinophilic granuloma is essential to prevent visual loss as a consequence of rapid tumor enlargement and eyeball dislocation.3 There are a few studies about performing fine needle aspiration (FNA) cytology in diagnosis of LCH or other orbital lesions.

We hereby report two cases of orbital LCH who were initially diagnosed by FNA.

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Diagnosis was confirmed by routine H&E and immunohistochemistry (IHC) methods afterwards, in order to emphasis the accuracy of FNA cytology in the diagnosis of orbital mass lesions specially LCH.

**Case reports**

Our study was performed in Farabi eye hospital, an ophthalmology referral center related to Tehran University of Medical Sciences, during 2 years; 2005 and 2006.

**Case 1**

A one-year-old boy presented with painless progressive swelling of left lower lid and proptosis from 15 days before, was referred to our center in April 2005. Physical examination showed a firm swelling over the superior and lateral aspect of left orbit measured 3 cm. Ocular movement was limited in the left eye. His visual field and acuity were normal. CT scan showed a solid tumor located in left temporal rim of orbit with bone invasion and erosion, suspicious for malignancy. Systematic physical examination including size of liver, spleen and lymph nodes, and laboratory tests such as complete blood count, liver function tests, urine analysis all were normal. Chest X-ray and skeletal survey also showed no abnormality. FNA and histopathology of the mass are described later.

**Case 2**

A three-year-old boy was admitted to our hospital on July 2006, complaining of right lower lid swelling from three months before. A tender fibrous mass in right inferolateral aspect of orbit was identified during physical examination. The remainder of ocular examination was unremarkable. In brain and orbital CT scan evaluation, a 3-cm homogenous mass with peripheral condensation centered in right lateral orbital wall with bone destruction and extracranial extension was discovered. There was also extension into infratemporal fossa and soft tissue around the temporal region as well. Right globe was normal with middle and upper displacement. Intracranial extension was not identified. Systematic physical examination, laboratory findings and X-ray surveys were also within normal limits.

FNA cytological smears orbital masses in both cases stained with PAP and Giemsa methods, showed same appearance that was a mixed cell population of eosinophils, neutrophils and lymphocytes accompanied with neoplastic histiocytes with folded nuclei; so called “coffee-bean” nuclei. Some of these cells were binuclear or multinucleated. The cytological diagnosis was “Langerhans cell histiocytosis” (Figure 1A). After surgical excision of the tumor, the diagnosis was confirmed by routine H&E method (Figure 1B). Both cases showed positive stain CD1a in IHC assessment.
Discussion
LCH, previously called histiocytosis X, is a remarkably variable clinicopathologic entity characterized by proliferation of Langerhans cells. The etiology of LCH remains unknown. A reactive origin has been suggested but some studies demonstrated clonality in some cases. The clinical features and behavior of LCH depend on the extent of organ involvement. LCH can present as solitary or multiple lesion in one organ (bone being the most common) or as a disseminated disease. Patient with disseminated disease may have lymphadenopathy, skin lesions or diabetes insipidus.4,5

Hidayat et al showed that orbital involvement in a series of 76 children with LCH was 23%.6 X-ray assessment reveals the lesion in orbit, characterized by osteolytic lesion with sclerotic margins, commonly involved the zygomatico-frontal suture.2 Nevertheless, involvement of temporal bone and lateral wall of the orbit were prominent in our cases.

Most cases of LCH present before the age of 20.7 However, other lesions specially malignancies such as lymphoma, rhabdomyosarcoma, neuroblastoma, Ewing sarcoma and chloroma needed to be excluded in children with primary orbital involvement.8

FNA cytologic findings of LCH have been well described, but using this method in diagnosis of orbital lesions is a recent experience. FNA cytology is a rapid, safe and useful method for early diagnosis of LCH9-11 and in our experience is a rather specific method for primary exclusion of other possible differential diagnosis. In a typical cytological appearance, as seen in our cases, LCH consists of abundant Langerhans cells having pale ill defined eosinophilic cytoplasm, lobulated nuclei with longitudinal groove, accompanied with many eosinophils and varying number of neutrophils, lymphocytes, macrophages and multinucleated giant cells.7 A definitive diagnosis is made by histopathologic examination of the tumor, the presence of Birbeck granules on electron microscopy or the positivity for CD1a antigen by IHC method12 as we did in our study.

Conclusion
It is important to perform a complete systemic survey in LCH patients with orbital involvement in order to rule out disseminated disease. The distinction between uni- and multisystem disease is very crucial in management and prognosis of the disease. In our two cases, the disease was limited to orbital region with no other systemic involvement.

We conclude that even if CT scan findings may indicate an aggressive disease, LCH should be considered in differential diagnosis of primary orbital bone lesions specially in childhood. FNA can be used as a useful method in the diagnosis of orbital lesions suspicious for LCH.

References