

Cytological diagnosis of unifocal Langerhans cell histiocytosis in the temporomandibular joint

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To the Editor,

Unifocal Langerhans cell histiocytosis (LCH) presents as a solitary focal bone lesion characterized by clonal proliferation of CD1a (+) and langerin(+) dendritic cells, derived from the bone marrow. In 38-69% of cases BRAF-V600E mutation is revealed. It gains attention to maxillofacial surgeons due to involvement of the orofacial region and early onset of jaws symptoms. If overlooked, massive deconstruction of tissues, occurs.

We present a case of LCH of the temporomandibular joint in a 14 aged child where fine needle aspiration favorably contributed in an accurate diagnosis combined with the appropriate clinicoradiological findings. This can overrule unwarranted biopsy and monitor the treatment largely where approach to histopathology services is restricted.

A 14-year old boy presented with swellings, and pain in the left temporomandibular joint, without history of trauma, and uneventful medical history. There was limitation in mouth opening. He experienced tenderness on palpation of the joint that restricted jaw movements. Occlusal contact was absent on the first premolar area of the upper and lower dentition. Laboratory work up revealed an elevated alkaline phosphatase, reflecting bone pathology.

Contrast enhanced MRI showed (fig.1) a mass 4.7 cm in the left infratemporal region, with increased, heterogeneous signal intensity. It had obliterated the condyle and the coronoid process. It invaded the

masseter, the lateral and the medial pterygoid muscles. The borders of the mass were irregular with poor demarcation.

Fine needle aspiration biopsy was performed and yielded 1ml hemorrhagic fluid. The ThinPrep technique was applied. It achieves a diagnostic sensitivity as accurate as conventional preparations, especially for its excellent cell preservation and lack of background which decrease the amount of inadequate diagnoses. ThinPrep preparations are a valuable supplement, particularly for



Figure 1. MRI showing a 4,7 cm mass in the left infratemporal region.

immunocytochemical staining. Smears showed lymphocytes, mature and transformed, plasma cells, neutrophils, rare eosinophils, histiocytes not otherwise specified, and atypical polygonal cells with abundant cytoplasm, convoluted and grooved oval nuclei (with a coffee bean appearance), and inconspicuous nucleoli. Some showed intranuclear pseudoinclusions. Atypical cells were positive by CD1a (membrane staining) (fig. 2) and S-100 (cytoplasmic and nuclear staining). A cytological diagnosis of LCH was rendered. Because all of the examination results indicated only a single bone lesion, a trial of indomethacin was decided before administering chemotherapy, (2.5 mg/kg/day, for 5 months, twice daily).

The diagnosis of LCH is based on hematologic and histologic criteria (1).

Experience has gained in accurate cytological diagnosis of LCH in various body organs based on characteristic features in the appropriate clinical and radiological setting as reports show that cytology mirrors histomorphology (1).

The hallmark is the Langerhans cell (LC) with nuclear grooves and nuclear pseudoinclusions (1,3,4,5). LCs demonstrate pleomorphism and mitotic activity. Dendritic cytoplasmic processes are rare. Eosinophil infiltration varies from scant to abundant in cytology smears. Charcot-Leyden crystals containing eosinophil membrane protein from rupture of eosinophil's

granules indicate tissue eosinophilia towards LCH diagnosis (1,3,4,5).

LCs stain positive for S-100, PNA (peanut agglutinin), MHC class II, CD1a, and langerin (CD207) (1,3,4,5). Langerin was not employed in our case. The Birbeck granule is their ultrastructural hallmark. Electron microscopy was not performed in our patient and was not considered essential for diagnosis as also suggested by other authors (1).

Our LCH case was based on FNA cytological smears which pointed out LCs, in the proper radiology and clinical settings. Mandibular lesions tend to destroy alveolar bone, producing the radiologic appearance of "floating teeth" (2).

It is necessary to be familiar with cytological features of other differential diagnoses. Ewing's sarcoma and non-Hodgkin lymphoma are characterized by monotonous population of small round blue cells and lack features of LCs (1, 3, 4, 5).

Sinus histiocytosis with massive lymphadenopathy (SHML) involves cervical nodes. In SHML, histiocytes have abundant cytoplasm, with hematopoietic phagocytosis and prominent nucleoli (1).

Secondary hyperplasia of the LCs is associated with lymphomas, especially Hodgkin's disease and lung tumors (1).

Tumors with cells with nuclear grooves or pseudoinclusions should be ruled out (malignant melanoma, papillary thyroid carcinoma) (1).

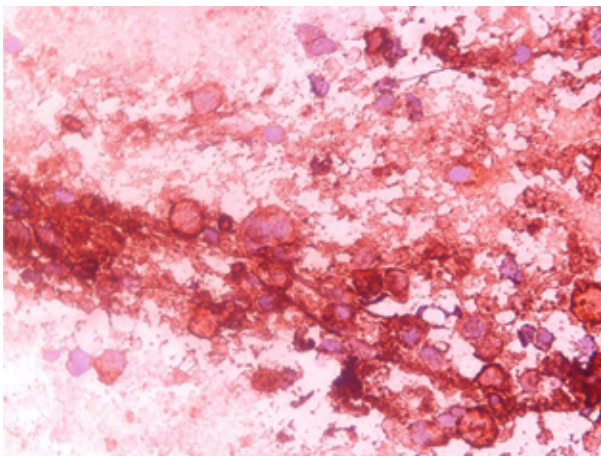


Figure 2. FNA smear. Langerhans cells show membrane positivity by CD1a. CD1a immunostain X400.

Conflicts of interest: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article.

References

1. Kumar N, Sayed S, Vinayak S. Diagnosis of Langerhans cell histiocytosis on fine needle aspiration cytology: A case report and review of the cytology literature. *Patholog Res Int.* 2011; 2011: 439518-439523
2. Stull MA, Kransdorf M, Devaney K. From the Archives of the AFIP. Langerhans Cell Histiocytosis of Bone. *RadlGraphlcs* 1992; 12:801-823

3. Park J, Chung J. Long-term treatment of Langerhans cell histiocytosis of the mandibular condyle with indomethacin. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod.* 2010 Apr;109(4): e13-21.
4. Tamiolakis D, Barbagadaki S, Proimos E, et al. Touch imprint cytological diagnosis of nodal Langerhans cell histiocytosis. *B-ENT* 2009; 5: 115-118
5. Hwang DS, Lee JS, Kim UK, et al. Langerhans cell histiocytosis of the mandible: two case reports and literature review. *J Korean Assoc Oral Maxillofac Surg* 2019; 45:167-72.

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