Sinus histiocytosis with massive lymphadenopathy.
Is the lymph node enlargement always massive?

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ABSTRACT
Sinus histiocytosis with massive lymphadenopathy (SHML) is a benign proliferating histiocytic disorder, predominantly of lymph nodes with extra-nodal involvement in some cases. It is a self-limiting disease and has a good prognosis; however some patients need steroid therapy. Therein lays a need to differentiate it from other lympho proliferative disorders of poorer prognosis. Clinically, it is suspected only when there is a significant lymph node enlargement in younger age group as the term signifies. Recently, we came across a case where a middle-aged female presented with a subtle swelling in submental region, clinically suspected of reactive or tubercular etiology. However on fine needle aspiration cytology, the smears showed many histiocytes, some of them showing lymphocytes and plasma cells in their cytoplasm, a morphological feature called as ‘emperipolesis’ classically seen in SHML. The present case is reported because of unusual clinical presentation and possibly the need for a revision of existing terminology.

Key words: Rosai-Dorfman disease, sinus histiocytosis, lymphadenopathy.
Rosai Dorfman disease is a rare, idiopathic histiocytosis. (1) The disorder is thought to occur as a result of immune dysregulation or response to a presumed infectious agent. The stimulation of monocytes/macrophages via Macrophage-Colony-Stimulating-Factor leads to immunosuppressive macrophages, which is a main pathogenesis of RDD. (4) Clinically mean age of onset is second decade; other features include fever, non-tender bilateral cervical lymphadenopathy, leucocytosis, elevated ESR and polyclonal hypergammaglobulinemia. However, in contrast our patient was a middle-aged woman with a subtle submental lymphadenopathy; though she had fever and a high ESR.

The morphological features include a histiocytic proliferation with presence of large histiocytes showing fine vacuoles in the cytoplasm and large vesicular nuclei. The phagocytosis of lymphocytes and plasma cell (emperipoleis) is characteristic. (5) The background population predominantly consists of lymphocytes, plasma cells, neutrophils and few eosinophils. Although the cytomorphological features are well described, diagnostic difficulties may sometimes arise. The common differential diagnoses (D/D) include infectious lesions, reactive lymphoid hyperplasia with sinus histiocytosis (RLHSH), Langerhans cell histiocytosis (LCH), hemophagocytic syndrome and malignant lymphoma. (5-8) A detailed clinical history and careful morphological assessment usually prevents misdiagnosis. The most important D/D is RLHSH and LCH; the features favoring SHML are numerous histiocytes with prominent emperipoleis. On IC histiocytes in RLHSH and SHML show strong positivity for S-100 and CD-68; however in LCH the histiocytes also show positivity for CD1a. Further confirmation of LCH can be done by demonstrating Birbeck’s granules on electron microscopy.

The course of RDD is usually benign, indolent and self-limiting in most of the patients. Treatment does not appear to be necessary in the majority of patients since the disease does not usually threaten life or organ function. Surgery is generally limited to biopsy to confirm the diagnosis or, to relieve obstructive symptoms. Patients with progressive disease have been treated with corticosteroids, chemotherapy or radiotherapy with variable results. (9) Our patient received corticosteroids to which she responded well.

To conclude, this case should alert the clinician and cytopathologist to the possibility of occurrence of SHML in minimally enlarged lymph nodes. Careful interpretation of FNAC slides along with appropriate marker study would help in early diagnosis and timely management. An increased awareness of such a possibility by the clinician and the pathologist is desired. Moreover, it is suggested that the terminology of massive lymphadenopathy in SHML may be revised to Sinus Histiocytosis with lymphadenopathy (SHL) or use of RDD should be encouraged.

**REFERENCES**