Fine-Needle Aspiration Cytology of Rosai-Dorfman Disease of Bone

Xin Jing, M.D.,* Jonathan B. McHugh, M.D., and Robert T. Pu, M.D., Ph.D.

Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease) is a rare, benign self-limiting condition of unknown etiology. Less than a quarter of cases have only extranodal involvement and a few cases of skeletal involvement of Rosai-Dorfman disease without associated lymphadenopathy have been reported in the literature. We herein report cytohistologic findings in a case of sole skeletal Rosai-Dorfman disease in a 51-year-old woman who presented with an expansile, heterogeneous lesion at T11 with cord compression and edema. A CT-guided fine-needle aspiration of T-11 lesion was performed and the sample was processed by ThinPrep technique. The ThinPrep smear showed characteristic features of Rosai-Dorfman disease including hypercellularity with moderate number of histiocytes in a background of lymphocytes, plasma cells, and neutrophils. The histiocytes possessed abundant, pale and vacuolated cytoplasm, rounded nuclei with smooth nuclear membranes, fine chromatin, and distinct nucleoli. The histiocytes showed emperipolesis of lymphocytes and neutrophils. The diagnosis was confirmed by concurrent biopsy with immunhistochemical study. Our case highlighted the role of fine-needle aspiration with ThinPrep technique in the diagnosis of Rosai-Dorfman disease. Diagn. Cytopathol. 2008;36: 516–518.

Key Words: FNA; ThinPrep; skeletal Rosai-Dorfman disease

Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease) was first described by Rosai and Dorfman as a rare, benign self-limiting condition of unknown etiology in 1969.[1] It mainly involves pediatric and young adult patients with a male predominance. The classical presentations include bilateral cervical lymphadenopathy, fever, leukocytosis, elevated erythrocyte sedimentation rate, and hypergammaglobulinemia.[2] However, one-third of cases present with extranodal involvement. Further, less than a quarter of cases have only extranodal involvement[3,4] and the exclusive extranodal involvement is more commonly seen in elderly patients.[5] The most common extranodal sites include head and neck organs, respiratory tract, skin, and bone. A few cases of skeletal involvement of Rosai-Dorfman disease without associated lymphadenopathy have been identified.[6–8] We herein report cytohistologic findings in a case of sole skeletal Rosai-Dorfman disease with associated spinal cord compression.

Case Reports

A 51-year-old woman presented with 1 month history of back and hip pain, progressive tingling and weakness of bilateral lower extremities. The patient also noticed a mass on her right clavicle. MRI images of lumber spine revealed a heterogeneous lesion at T11 with cord compression and edema. A CT-guided fine-needle aspiration and a concurrent core needle biopsy of the T11 lesion were performed. The fine-needle aspiration sample was submitted in the Cytolyt solution from which a ThinPrep smear and a cell block slides were prepared. The core biopsy sample was fixed in formalin, embedded in paraffin, and stained with H&E. Special stain for acid fast bacilli (Fite stain), and immunohistochemical studies for pancytokeratin, S-100, and CD1a were performed.

The ThinPrep smear showed hypercellularity with moderate number of histiocytes, abundant lymphocytes and some plasma cells, as well as some neutrophils. The histiocytes possessed abundant, pale and vacuolated cytoplasm, rounded nuclei with smooth nuclear membranes, fine chromatin, and distinct nucleoli (Fig. 1). The histiocytes showed emperipolesis of lymphocytes and neutrophils (Fig. 2). The cell block revealed clusters of mixed inflammatory cells with crush artifact. As no additional material was available for further studies, a cytological diagnosis of "acute and chronic inflammation; negative for neoplasm" was made.

Department of Pathology, University of Michigan Health System, Ann Arbor, Michigan

*Correspondence to: Xin Jing, M.D., Department of Pathology, University of Michigan Health System, 1500 E Medical Center Drive, Ann Arbor, MI 48109. E-mail: xinjing@umich.edu

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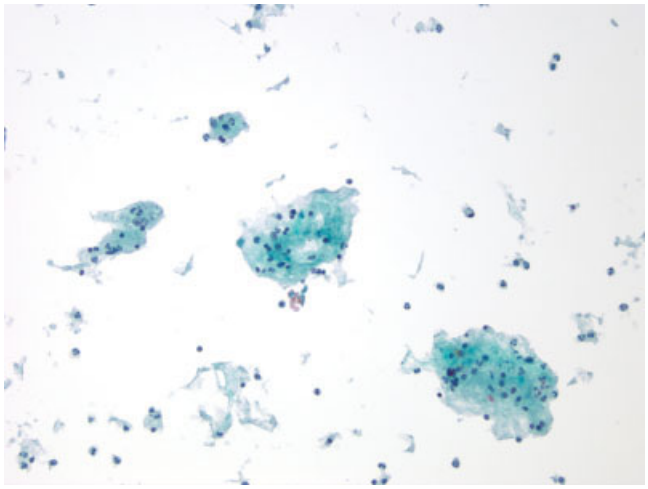

Fig. 1. ThinPrep smears showing large histiocytes in a background of mixed inflammation (Papanicolaou stain, ×200). [Color figure can be viewed in the online issue, which is available at www.interscience.wiley.com.]

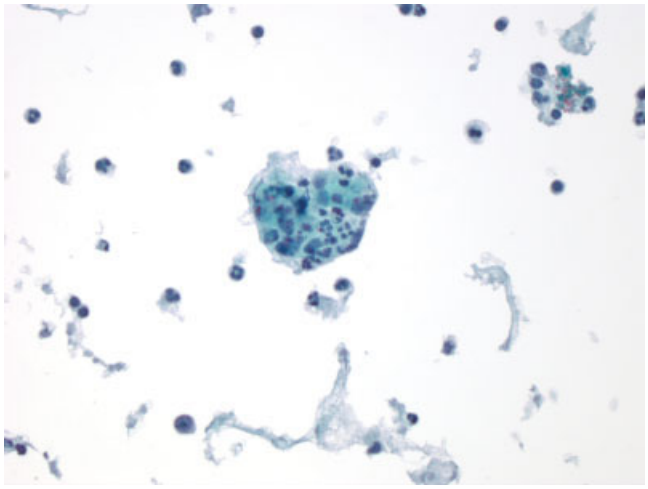

Fig. 2. The histiocytes showed emperipolesis of neutrophils and lymphocytes (Papanicolaou stain, ×400). [Color figure can be viewed in the online issue, which is available at www.interscience.wiley.com.]

The H&E of the concurrent core needle biopsy showed numerous histiocytes interspersed with various number of lymphocytes, plasma cells, and neutrophils (Fig. 3). The histiocytes revealed emperipolesis of lymphocytes and neutrophils. Fite stain for acid fast microorganisms was negative. Immunohistochemical studies showed that the histiocytes were positive for S-100 (Fig. 4) and were negative for pancytokeratin and CD1a. A histologic diagnosis of "extranodal Rosai-Dorfman disease" was made. Subsequently, curettage of the spinal lesion with right semihemilaminectomy and right transpedicular vertebral body decompression was performed. Biopsy of the right clavicle mass was also performed and histologic changes were consistent with extranodal Rosai-Dorfman disease.

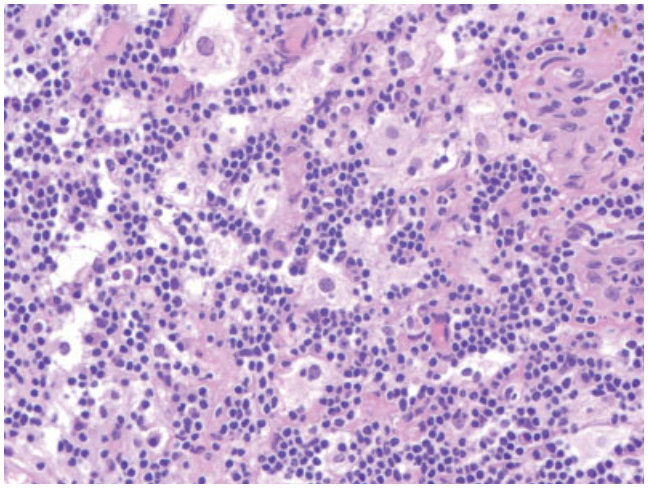

Fig. 3. H&E of current biopsy shows similar findings (H&E, ×400). [Color figure can be viewed in the online issue, which is available at www.interscience.wiley.com.]

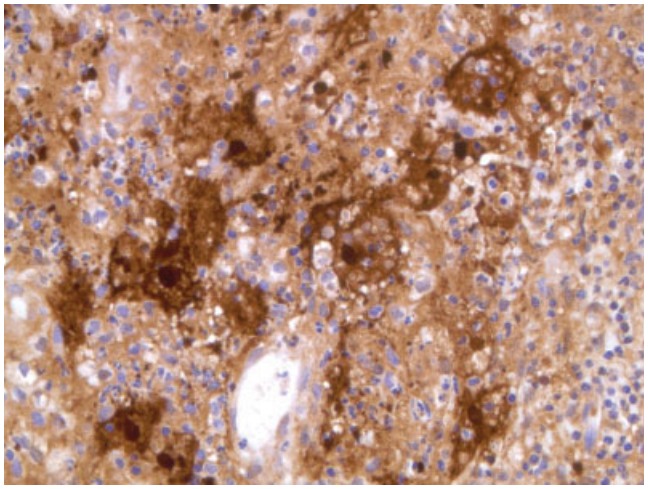

Fig. 4. The histocytes are positive for S-100 (Immunostain, ×400). [Color figure can be viewed in the online issue, which is available at www.interscience.wiley.com.]

Discussion

There are a limited number of reports in the English literature on fine-needle aspiration cytological features of Rosai-Dorfman disease. Commonly, conventional smears are made from fine-needle aspiration samples. Cytological features have been characterized as cellular smear with numerous large histiocytes in a lymphoplasmic background and the histiocytes show phagocytosis of lymphocytes, plasma cell, and occasional neutrophils.[9,10] Although the cytologic features were not diagnostic for the disease, fine-needle aspiration is helpful to differentiate it from other lesions. Differential diagnoses include sinus histiocytosis, hemophagocytic syndrome, Langer-

hans cell histiocytosis, reactive lymphadenitis, and malignant lymphoma. To make a definitive diagnosis of Rosai-Dorfman disease, special stains for organisms and immunocytochemical stains should be performed on an optimal cell block. The positive staining results for S-100 and CD68 and negative staining results for CD1a support the diagnosis of Rosai-Dorfman disease. Unfortunately, an optimal cell block was not available for our case and further classification of the lesion was thus rendered on the concurrent biopsy. Despite of its limitation, fine-needle aspiration is a useful tool, as being aware of and/or recognizing this rare entity may help in the triage of specimen and prevent unnecessary surgical procedures.

Skeletal involvement as a sole manifestation of Rosai-Dorfman disease is extremely uncommon and it can involve solitary or multiple bones. Rarely, patients present with symptoms of spinal core compression as a consequence of Rosai-Dorfman disease of spine.[11,12] The skeletal lesions of Rosai-Dorfman disease are typically lytic with either poorly or sharply defined margins and are rarely sclerotic.[7] Whereas nodal Rosai-Dorfman disease which needs to be distinguished from lymphomas, major differential diagnoses of extranodal involvement of this entity includes various neoplasms such as lymphomas, metastatic carcinoma, and melanoma.

We report fine-needle aspiration cytologic features of Rosai-Dorfman disease with sole skeletal involvement and associated spinal cord compression, a rare entity. To our best knowledge, many comparison of conventional versus ThinPrep smears have been done with regard to cytologic evaluation of nongynecologic specimens. It remains controversial whether ThinPrep smear is as efficient as conventional smears and the nature of a lesions do play a role for the inconsistent results. Our case is unique in that instead of making conventional smears, the sample was processed with ThinPrep technique and was found that the cytological features showed in conventional smears are well reproduced in ThinPrep smear. It is thus thought that ThinPrep preparation may be as reliable as conventional smear in the fine needle aspiration diagnosis of Rosai-Dorfman disease. Diagnostic challenges arise when facing hypocellular samples with few histiocytes or if the entity is not considered. Regardless, immunocytochemical/immunohistochemical stains performed on an optimal cell block or a biopsy are certainly a key to confirm the diagnosis.

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