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 immunohistochemical study. Our case highlighted the role of fine-needle aspiration with ThinPrep technique in the diagnosis of Rosai-Dorfman disease. 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. The histiocytes showed emperipolesis of lymphocytes and neutrophils. The diagnosis was confirmed by concurrent biopsy with immunohistochemical study. Our case highlighted the role of fine-needle aspiration with ThinPrep technique in the diagnosis of Rosai-Dorfman disease.
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 semi-hemilaminectomy 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.

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. 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. The skeletal lesions of Rosai-Dorfman disease are typically lytic with either poorly or sharply defined margins and are rarely sclerotic. 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.

References