Primary Rosai-Dorfman disease (sinus histiocytosis with massive lymphadenopathy) of the pancreas: Comparison of cytomorphology in conventional smears, liquid-based preparations and histopathology

Chih-Yi Liu¹, Feng-Chuan Tai², Shih-Hung Huang³

¹ Division of Pathology, Sijhih Cathay General Hospital, New Taipei City, Taiwan
² Department of Surgery, Cathay General Hospital, Taipei City, Taiwan
³ Department of Pathology, Cathay General Hospital, Taipei City, Taiwan

Background:

Rosai–Dorfman disease (sinus histiocytosis with massive lymphadenopathy) is a rare clinicopathologic variant of histiocytic disorders, and is characterized by an indolent clinical course and the proliferation of distinctive histiocytes within lymph node sinuses. Extranodal involvement in the Rosai–Dorfman disease (RDD) has now been documented in several organ systems. The gastrointestinal tract is rarely involved, and very few cases of primary involvement of the pancreas have been previously described. Herein, we report a rare case of primary Rosai–Dorfman disease of the pancreas. The cytomorphologic features in both liquid-based cytology and conventional specimens are introduced.

Case presentation:

A 71-year-old Taiwanese female was incidentally found to have a pancreatic tumor. Computed tomography of the abdomen revealed a 3.5-cm hypo-enhancing mass in the pancreatic tail. No radiographic evidence of lymphadenopathy or obvious metastatic disease was observed. Based on the clinical suspicion of malignancy, the patient underwent distal pancreatectomy and splenectomy. The surgical specimen demonstrated a well-defined, solid, and multinodular mass that measured 3.2 cm in its greatest dimension and that occupied the pancreatic tail. [Figure 1]

Histopathological findings:

The pancreatic tumor revealed diffuse proliferation of the histiocytoid cells accompanied with an inflammatory cell component and fibrotic stroma. The cellular infiltrates comprised pale areas formed by histiocytes and dark areas comprising plasma cells or small lymphocytes. The large histiocytes possessed abundant pale or eosinophilic cytoplasm, small round nuclei, and visible nucleoli. Some of the histiocytes contained intracytoplasmic lymphocytes, also known as emperipolesis or lymphophagocytosis. [Figure 2]

Cytological findings:

Touch imprint cytology of the resected tumor was performed for air-dried and alcohol-fixed smears. The tissue rinse cytology specimens were also prepared using the liquid based methods (SurePath and CellPrep). The cytologic specimens contained a polymorphous cellular component with numerous histiocytes, often with multinucleation and emperipolesis, in addition to lymphocytes and plasma cells. [Figure 3]

Discussion:

Until now, only six cases of extranodal RDD in the pancreas have been reported in the English literature. Review of the previous case series reports showed that a diagnosis of RDD is possible on FNA cytology, given adequate ancillary testing support. Despite the rare incidence in pancreas, the recognition of characteristic cytomorphologic features and immunophenotype contribute to the specific diagnosis. Touch imprint, as well as tissue rinse liquid-based cytology, can be effectively used for diagnostic purposes.