

Multifocal nodal and extranodal Rosai-Dorfman disease initially diagnosed as histiocytic lymphoma.

ABSTRACT

Rosai-Dorfman disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy (SHML), is a systemic disease involving nodal and extranodal tissues. We report a 48-year-old female with recurrent nasal obstruction due to polypoidal masses involving the nasal sinuses, turbinates and septum bilaterally, and lumps in the right infra-orbital region and region of the right lacrimal sac. A 4 cm right upper neck mass was also noted, which was initially diagnosed as histiocytic lymphoma. Histopathology of the nasal and infraorbital lesions revealed fibro-inflammatory masses containing histiocytic cells with large vesicular nuclei and abundant foamy cytoplasm exhibiting emperipolesis and lymphophagocytosis, admixed with scattered plasma cells and lymphocytes. These histiocytes revealed immunohistochemical positivity for S-100 protein and CD68, but were negative for CD1a. The findings supported a diagnosis of RDD. This report serves to remind pathologists and clinicians of the extranodal manifestations of RDD and its potential confusion with lymphomas.

Keyword: Rosai-Dorfman disease (RDD); Histiocytosis; Sinus; Lymphoma; Nasal obstruction.