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 infraorbital 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.