Case Report
Extranodal Rosai-Dorfman Disease Presenting as an Intranasal Mass

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Abstract Objective. We report the case of a 40-year-old female with biopsy-proven cutaneous extranodal Rosai-Dorfman who presented with symptoms of nasal obstruction and sinusitis. Rosai-Dorfman disease, also known as sinus histiocytosis with massive lymphadenopathy (SHML), is a rare, idiopathic disorder characterized by a benign proliferation of histiocytes initially thought to be limited to cervical lymph nodes. However, between 30% and 50% of patients identified with SHML have extranodal manifestations. Study design. Case report and literature review. Results. The patient presented to the otolaryngology clinic with nasal obstruction. A CT scan demonstrated lesions of the left inferior turbinate and anterior nasal septum with bony erosion. The patient underwent a biopsy of the nasal mass. The specimen was evaluated with flow cytometry, microscopy, and stained for immunohistochemical markers which confirmed a diagnosis of Rosai-Dorfman. Conclusion. Rarely, Rosai-Dorfman disease may manifest in the nasal cavity with symptoms mimicking that of sinusitis. Otolaryngologists should be familiar with the pathophysiology of the disease, the variability of extranodal involvement, radiologic findings, and the immunohistochemical analysis of Rosai-Dorfman to aid in diagnosis. This report emphasizes the radiological, pathological, and clinical appearance of the disease.

Keywords Rosai-Dorfman disease; immunohistochemistry; intranasal mass

1. Introduction
Rosai-Dorfman disease, also known as sinus histiocytosis with massive lymphadenopathy (SHML), was first described by Rosai and Dorfman in 1969 [1]. This rare, idiopathic disorder is characterized by a benign proliferation of histiocytes initially considered to be limited to cervical lymph nodes [2,3]. However, some reports note that between 30% and 50% of patients have extranodal manifestations [3,4]. Histochemical staining is necessary for diagnosis when suspicious extranodal lesions are identified [2]. We present a case of extranodal Rosai-Dorfman disease occurring in the nasal cavity and describe the subsequent steps required to confirm the diagnosis. IRB exemption was obtained from the Committee for Human Research (CHR) at UCSF prior to chart review.

2. Case presentation
A 40-year-old female with a ten-year history of biopsy-proven cutaneous Rosai-Dorfman presented with a nasal mass concerning for a new extranodal manifestation of her disease. The patient was previously asymptomatic with regards to her Rosai-Dorfman disease and was not on any systemic treatment. She first noted a nasal obstruction in the setting of an upper respiratory infection (URI) which failed to respond to medical management. Her primary physician subsequently discovered an intranasal mass. CT imaging revealed lesions of the left inferior turbinate and nasal septum causing bony erosion suggestive of lymphoma or a new extranodal dissemination of Rosai-Dorfman (Figure 1). On endoscopic examination, hypervascular lesions were identified emanating from the septum bilaterally and the left inferior turbinate. Given the vascularity of the lesions, it was recommended that the patient undergo operative biopsy. Operative findings included a submucosal mass along the septum bilaterally and a left inferior turbinate lesion (Figure 1). Using a KTP laser, these lesions were removed and sent for pathologic study which demonstrated...
Figure 2: H&E stain, 200× magnification. Notice the numerous histiocytes and mature, small lymphocytes. Positive S-100 staining (upper left) confirmed the diagnosis of Rosai-Dorfman.

a histiocyte-rich, chronic inflammatory environment that could not be distinguished from other chronic infections. H&E staining demonstrated numerous histiocytes with abundant pink cytoplasm admixed with mature small lymphocytes (Figure 2). Also scattered throughout the tissue were multinucleated giant cells, focal collagenous fibrosis, and focal organizing fibrin accumulation. These findings portray an active inflammatory process, but for accurate diagnosis immunohistochemical stains were necessary. Specimens were stained for CD3, CD20, CD56, EBV, CD30 (lymphoma markers), CD68 (found in histiocyte rich tissues), and S-100. Flow cytometry was also performed to rule out a lymphomatous process. Our specimen stained positive for CD68 and S-100, whose positivity supports the diagnosis of Rosai-Dorfman disease (Figure 2). It was planned for the patient’s case to be discussed at our tumor board with necessary referrals made for subsequent treatment.

3. Discussion

The diagnosis of extranodal Rosai-Dorfman disease can be challenging. It has a tendency to involve numerous extranodal sites with a wide variety of presentations. In the head and neck, it often manifests as an infiltrating submucosal lesion [4]. Though it often shows evidence of tissue invasion suggesting a neoplastic process, its indolent course and histopathological findings convey a more reactive type etiology [1]. While the cause is unknown, multiple infectious agents, including both Epstein-Barr virus and herpesvirus, have been proposed in its pathogenesis [1, 4]. With little insight into the disease pathophysiology, the differential diagnosis remains diverse and includes various malignancies and inflammatory processes such as lymphoma, sarcoidosis, Wegener’s, rhinoscleroma, and atrophic rhinitis. Therefore, a definitive diagnosis often requires a histopathological and immunohistochemical examination of surgical specimens. A characteristic histopathologic feature is the presence of prominent histiocyte proliferation and activation [1, 2]. As noted in other studies, positive immunohistochemical stains for S-100 and CD68 confirm the presence of histiocytes which makes the diagnosis of Rosai-Dorfman more likely [2]. Once a diagnosis is made, treatment options include oral steroids or chemotherapeutic agents, radiation or surgery depending on the location and extent of the disease.

Head and neck manifestations can be seen in up to a quarter of patients with Rosai-Dorfman. Second only to the skin, the nose and paranasal sinuses have the greatest predilection for extranodal involvement. Still, extranodal manifestations of Rosai-Dorfman disease involving the nasal cavity have been infrequently reported [2, 3, 4, 5]. The cases that have been described often report synchronous lesions found in addition to the nasal involvement, with isolated nasal lesions uncommon [2, 3, 4, 5]. Our study reports the case of an isolated nasal lesion in a patient with CT and clinical photographic documentation consistent with extranodal Rosai-Dorfman disease. Correlating this with the pathology and immunohistochemical properties of the surgical specimen, a template is provided to guide the physician from clinical discovery to diagnosis.

Conflict of interest The authors declare that they have no conflict of interest.

References