Extranodal Rosai-Dorfman Disease Presenting as an Intranasal Mass

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ABSTRACT

Rosai-Dorfman disease is a rare, idiopathic disorder characterized by a benign proliferation of histiocytes initially thought to be limited to cervical lymph nodes. However, between 30-50% of patients identified with the disease have extranodal manifestations.

A 40 year-old female with biopsy proven cutaneous extranodal Rosai-Dorfman presented with symptoms of nasal obstruction. A CT scan demonstrated lesions of the left inferior turbinate and anterior nasal septum.

The patient underwent a biopsy of the nasal mass. The specimen was evaluated with flow cytometry, microscopy, and stained for immunohistochemical markers whose results were all consistent with the diagnosis of Rosai-Dorfman disease. Clinical images, CT scans, and pathology slides are presented to familiarize clinicians with this disorder.

INTRODUCTION

Rosai-Dorfman disease, also known as sinus histiocytosis with massive lymphadenopathy (SHML), was first described by Rosai and Dorfman in 1969.

This rare, idiopathic disorder is characterized by a benign proliferation of histiocytes initially considered to be limited to cervical lymph nodes. However, between 30-50% of patients identified with Rosai-Dorfman have extranodal manifestations of the disease.

Histochemical staining of pathologic specimens is necessary to diagnose the disease when suspicious extranodal lesions are identified.

DISCUSSION

This case describes a woman with cutaneous extranodal Rosai-Dorfman disease with a new manifestation in the nasal cavity. The disease is rare and few case reports exist in the literature to guide clinicians.

The specimen demonstrated a histiocyte-rich environment that could not exclude other inflammatory processes. Immunohistochemical staining was obtained, showing a positive S-100 stain consistent with the diagnosis of Rosai-Dorfman disease.

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.

Though it often shows evidence of tissue invasion suggesting a neoplastic process, its indolent course and histopathological findings convey a more reactive type etiology. The differential diagnosis is quite diverse and includes various malignancies and inflammatory processes such as lymphoma, sarcoidosis, Wegener’s, rhinoscleroma, and atrophic rhinitis. Definitive diagnosis often depends on histopathological and immunohistochemical examination of a surgical specimen.

A characteristic histopathologic feature is the presence of prominent histiocyte proliferation and activation. Positive immunohistochemical stains for S-100 and CD68 confirm the presence of a histiocyte-rich environment that makes the diagnosis of Rosai-Dorfman more likely.

Treatment options vary based on location and extent of disease. Surgery is the mainstay treatment for life or function-threatening obstruction. In more indolent cases, chemotherapy and radiation have shown favorable response rates.

CONCLUSIONS

While uncommon, 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 in location of extranodal involvement, and the immunohistochemical analysis of Rosai-Dorfman to aid in diagnosis of this rare entity.

REFERENCES

