Rosai-Dorfman Disease Isolated to the Nasal Septum

Wm Greg Young MD, Danny Meslemani MD, Jamie Segel MD, Tamer Ghanem MD
Department of Otolaryngology Head and Neck Surgery
Henry Ford Health System, Detroit, Michigan

Abstract

Objectives: To describe the typical presentation, pathology findings, and treatment options for patients presenting with Rosai-Dorfman disease.

Study Design: This case report was completed using chart review for data collection.

Methods: The case report was generated through chart review looking at physical findings found at presentation, intra-operative findings, and pathology. A literature search was completed, investigating typical presenting symptoms, surgical management, pathology findings, and treatment options.

Results: The patient was a 38 year old gentleman who presented with a six month history of nasal congestion and recurrent epistaxis. He was found to have significant enlargement of the nasal septum with no cervical lymphadenopathy. Surgical excision of the lesion via open rhinoplasty approach revealed surgical pathology consistent with Rosai-Dorfman disease. Due to the lack of involvement of other head and neck sites, no other intervention was necessary but close observation was recommended.

Conclusion: Rosai-Dorfman disease can present with isolated head and neck extranodal involvement at sites including the nasal septum. While no ideal treatment has been determined, interventions should correspond to the degree of symptoms experienced. Adjutant therapies may be indicated beyond surgical excision.

Introduction

Rosai-Dorfman disease, a rare benign disorder first classified in 1969, is an extranodal manifestation of Sinus histiocytosis with massive lymphadenopathy (SHML). SHML, classically seen in children, presents as bilateral massive non-tender lymphadenopathy located in the cervical region in ~90% of patients.[1] The lymphadenopathy may be accompanied by features of fever, leukocytosis, elevated ESR, and hypergammaglobulinemia. SHML is thought to be a disorder of the mononuclear phagocyte and immunoregulatory effector (M-PIRE) system with a speculated association with Epstein-Barr virus and Human Herpesvirus 6.[2, 3] Other proposed mechanisms include an occult chronic infection, an aberrant exaggerated immune response to an infectious agent, or an antigen that causes a proliferation of histiocytes.

Forty-three percent of patients with SHML have extranodal manifestations of the disease which demonstrate similar histopathology morphology to the nodal lesions. Importantly, twenty-three percent of cases of SHML have only extranodal involvement; this manifestation of disease is referred to as Rosai-Dorfman disease.[1] Unlike SHML, Rosai-Dorfman disease is more common in older individuals, lacks a classical presentation, and symptomatology varies depending on the location of the extranodal involvement. The most frequent extranodal sites in decreasing order are skin and soft tissue (16%), nasal cavity and paranasal sinuses (16%), ocular (11%), bone (11%), salivary gland (7%), central nervous system (7%), and oral cavity (4%).[1] Half of the head and neck manifestations are most commonly seen within the nasal cavity. Other common areas of involvement in this region include the pharynx, paranasal sinuses and trachea.

Case Report

GA is a 39 year old male with Crohn's disease and a 6 month history of progressive nasal obstruction and recurrent epistaxis. Physical exam demonstrated midline symmetrical swelling of the anterior septum which was obstructing the nasal airway. The overlying mucosa was normal, demonstrating no telangiectasias or ulceration. In addition, there was no lymphadenopathy present.

MRI of the nasal lesion demonstrated an 5.3 x 2.7 cm enhancing and encapsulated soft tissue mass centered in the nasal septum which had no communication with the skull base. Pathology from the biopsy specimen demonstrated a mixed inflammatory, granulomatous process with prominent S-100 positive histiocyte proliferation with emperiplois.

Due to the extensive size of the nasal mass and its obstructive nature, a combined endoscopic as well as external open septorhinoplasty approach was used. Intra-operative findings were significant for a large cm spongy midline nasal mass located between the mucoperichondrial flaps of the septum with a near complete obliteration of the cartilaginous septum. The mucosa of one side of the septum was preserved. In order to reconstruct the resulting saddle nose deformity, a L-strut of bone was harvested from the perpendicular plate of the ethmoid bone and sewn into place between the lower medial cartilages and nasal bones. In addition, a remnant piece of cartilage was used as a columellar strut.

Post-operatively, the patient’s nasal obstruction and periodic epistaxis resolved. Final pathology was obscure since the patient’s known Crohn’s disease could not definitely rule out other inflammatory conditions. Consequently, other inflammatory markers C-ANCA, P-ANCA, and rheumatoid factor were checked and were negative. Patient is now one year post procedure without any evidence of recurrence.

Discussion

Varying presentations of Rosai-Dorfman disease make its diagnosis quite challenging. Although imaging may be obtained, a pathologic evaluation is required for confirmation. The characteristic histopathology finding is lymphocytophagocytosis (emperiplois) where histiocytes have phagocytosed lymphocytes, plasma cells, erythrocytes, or polymorphonuclear leukocytes; however, this is found much less commonly in the extranodal lesions.[1] Immunohistochemical staining will demonstrate the presence of the S-100 protein and CD68, as well as the absence of the CD1a antigen.[2] Extranodal lesions lack the architecture of true nodal tissue and contain a greater proportion of fibrosis when compared to the nodal lesions. With the absence of the classic lymphadenopathy, the diagnosis may be overlooked and confused with diseases similar immunohistological characteristics such as Langerhans cell histiocytosis, Hodgkin's disease, Non-Hodgkin's lymphoma, metastatic carcinoma, and metastatic malignant melanoma.[2]

The benign nature and high rates of spontaneous remission of Rosai-Dorfman disease means patients typically require no treatment. Surgical management may be required if there are obstructive or compressive symptoms, as in our patient with the nasal obstruction. In addition to surgical excision, there has been reported success with CO2 laser excision for disease in the nasal cavity.[4] Medical management of Rosai-Dorfman disease, although not typically used, includes steroids, chemotherapy, radiation and low-dose interferon. In addition, close follow-up in patients with SHML and RDD is important because there have been case-reports showing an association with malignant lymphomas.[5] Renal, liver, lower respiratory, or widespread dissemination is correlated with a poor prognosis.

Conclusions

Rosai-Dorfman disease is an extranodal manifestation of sinus histiocytosis with massive lymphadenopathy which can be isolated to the head and neck. As this case demonstrates, the diverse presenting symptom possibilities directly relate to the wide variety of possible tissue involvement. With its subtle histopathology, especially in the extranodal lesions, Rosai-Dorfman disease must be included in the differential diagnosis of isolated masses in the head and neck.

References