INTRODUCTION

Rosai-Dorfman disease (RDD) otherwise known as sinus histiocytosis with massive lymphadenopathy, was first described by Rosai and Dorfman in 1969. RDD is a rare disease whose pathogenesis is poorly understood. The typical presentation of RDD is massive lymphadenopathy in the cervical neck; however extranodal involvement is not uncommon and is found up to 43% of cases. Documented sites of disease include paranasal sinuses, eye, kidneys, central nervous system, upper respiratory tract, gastrointestinal tract, genitalia, salivary glands, and temporal bone. RDD occurs mostly often in children and young adults. There is a slight male predominance. The differential diagnosis is broad and diagnosis is dependent on histopathology. Treatment of this infrequently encountered disease is varied and no consensus is reached on therapeutic measures.

METHODS

We report a unique case of RDD found in the bilateral nasal cavities and review the current literature and treatment recommendations for this disease.

CASE REPORT

A 27 yo female presented with nasal obstruction and 6 month history of enlarging left neck mass. Past medical history is significant for chronic sinusitis, asthma, esophageal reflux, and hypothyroidism. On clinical examination a 2.5 X 2.0 cm palpable level I lymph node was appreciated. Fine needle aspiration of the submental neck mass demonstrated a reactive node. The patient was taken to the operating room for nasal endoscopy and excisional biopsy of the neck mass. Intraoperatively a tan irregular ovoid lymph node was excised measuring 3.0 x 2.5 x 2.0 cm. On nasal endoscopy, the nasal cavities were evaluated. Multiple masses arising from the floor of the nose as well as the septum were visualized. The nasopharynx was obstructed secondary due to the masses and was unable to be thoroughly evaluated. Endoscopic biopsies were taken of a mass along the anterior floor of the nasal cavity.

Pathology of the lymph node showed nodal sinus histiocytosis with massive lymphadenopathy and nasal biopsies showed extra nodal sinus histiocytosis with massive lymphadenopathy consistent with Rosai-Dorfman disease. Flow cytometry was negative for lymphoma. Pathological analysis showed emperipolisis and staining positive for S100 and negative for CD1a.

REFERENCES


CONCLUSIONS

RDD is a rare and difficult disorder in diagnosis and treatment. No published therapeutic guidelines are currently used to treat RDD. Treatment is largely based on the individual patient. Studies are still ongoing to delineate RDD and its relationship to autoimmune proliferative disorders. This disease should be considered in the differential when encountering patients with nasal masses and reactive lymphadenopathy.

DISCUSSION

The etiology of RDD is not conclusive and several theories have been proposed. RDD has been suggested to result from an immune response to infectious agent or an aberrant response of macrophages to cytokines. However no single antibacterial or antiviral agent has been associated. Infectious agents that have been studied in association with RDD include Epstein Barr Virus (EBV) and Human Herpes Virus 6 (HHV-6). EBV has been largely discounted as an infectious agent due to the lack of evidence of EBV found in histocytes in RDD. HHV-6 has been found within DNA in histocytes of RDD in 7 cases.

Diagnosis is primarily dependent on pathology. Emperipolisis also known as lymphphagocytosis is pathognomonic for RDD. Lymphocytes penetrate and move within the histocyte. Immunohistochemical staining for S100 is also paramount. S100 is also found in dendritic cells in lymph nodes and Langerhan cells in skin. The main histologic challenge is differentiating sinus histiocytosis proliferation from RDD and Langerhan cell histiocytosis. However, Langerhan cell histiocytosis is distinguishable by its small grooved nuclei and association with eosinophilic microabscesses.

The majority of patients with RDD have a favorable prognosis. For most patients the disease is self-limiting. Poor prognosis is associated with extranodal involvement found in the kidneys, liver or lower respiratory tract. Treatment for RDD is variable. Patients with symptoms are more likely to be treated with steroids, which has been suggested to retard disease progression and prevent major organ involvement. Surgical excision may play a role, particularly in sinusonal disease. Endoscopic sinus surgery may be beneficial in preventing recurrent complications. Observation has been used in asymptomatic patients. No role for chemotherapy has been found and radiation therapy has been only been documented to be effective in case reports.

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