Sinus histiocytosis with massive lymphadenopathy Rosai-Dorfman disease: a unique case presentation

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Rosai and Dorfman first described sinus histiocytosis with massive lymphadenopathy (SHML) in 1969 with an article detailing 4 cases in which they differentiated this disease entity from the grouping of diseases categorized as histiocytosis X, where it was previously classified. Also known as Rosai-Dorfman disease (RDD), it is clinically characterized as massive, painless, bilateral, symmetric cervical lymphadenopathy, with fever and leukocytosis. An 11-year-old African American boy was referred to our clinic for extraction of a severely decayed tooth #30 and evaluation of a large right-sided neck mass. Initially, the patient had been seen by his general dentist who had diagnosed the mass as an odontogenic abscess. After 2 courses of different antibiotics, no changes in the mass were noted. Subsequently, the patient was sent to the emergency department where CT revealed multiple right-sided neck masses with the largest measuring 4×2 cm. The patient underwent an incisional biopsy by otolaryngology and a diagnosis of necrotic lymph tissue was made. Upon our examination, the carious tooth #30 was felt to be an incidental finding and fine-needle aspiration cytology of the largest mass was performed in 2 places. This also provided a diagnosis of necrotic lymph tissue. In concert with the patient and his mother, the decision was made to excise the mass because of psychosocial concerns. A massive right-sided lymph node attached to the submandibular gland was found and excised without complication. Histologic examination with S-100 stain confirmed a diagnosis of RDD. The patient healed well following surgery and has experienced no further lymphadenopathy. This case presentation and review of the literature is unique, as the patient presented with unilateral cervical lymphadenopathy only. Open biopsy and 2 fine-needle aspirations all returned as necrotic lymph tissue. Obtaining the correct diagnosis was additionally hampered by coincidental dental pathology on the affected side and final diagnosis was made only by excisional biopsy, which is not necessarily indicated in cases of RDD. (Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2011;112:e124-e126)

In 1969, Juan Rosai and Ronald Dorfman identified a benign disease entity they described as sinus histiocytosis with massive lymphadenopathy (SHML), now commonly known as Rosai-Dorfman disease (RDD).1 RDD is a well-described phenomenon characterized by overproduction of histiocytes that amass in the lymph nodes. Massive lymphadenopathy of the cervical neck lymph nodes is a common area of presentation; however, extranodal involvement has been documented in the skin, respiratory tract, bone, genitourinary system, oral cavity, central nervous system, eyes/orbit/ocular adnexa, salivary gland, tonsil, breast, soft tissue, and heart.2 We present a unique case of unilateral massive cervical lymphadenopathy without any other contralateral nodal or extranodal involvement.

CASE REPORT

An 11-year-old African American boy presented to our clinic for evaluation of a right-sided neck mass. Initially, the patient was seen by a dentist for evaluation of submandibular mass thought to be associated with a severely decayed tooth #30. He was treated for an odontogenic abscess with appropriate antibiotics. After several weeks and a change in antibiotics, he reported no pain, no drainage, and denied any change in the size of the mass.

With no resolution, the patient was taken to the emergency department for evaluation of the neck mass. Emergency room physicians evaluated the patient and consulted an otolaryngologist. Computed tomography of the neck with and without contrast was obtained, revealing multiple right-sided neck masses with the largest mass measuring 4×2 cm. At this point, the patient was taken to the operating room for an incisional biopsy. The pathology report described the specimen as necrotic lymph tissue.

At discharge, the otolaryngologist referred the patient to the Nova Southeastern University Department of Oral and Maxillofacial Surgery for further evaluation of the mass and for treatment of the lower right first molar. He was examined, and a mobile mass measuring 4×2 cm was palpated just below the right inferior border of the mandible within level II (Fig. 1). The
A carious lower-right first molar was felt to be an unrelated, coincidental finding, as there was no pain associated with the affected tooth and radiography showed no periapical pathology (Fig. 2). Fine-needle aspiration of the mass was performed in our clinic and cytologic examination provided a diagnosis of necrotic lymph node with no malignancy noted.

Given the diagnosis provided by the previous biopsies, and because of the concerns of the patient and family, surgery was performed to remove the neck mass. Operative findings revealed a large 4 × 2-cm node, which was firmly attached to the submandibular gland. Scalloping of the inferior border of the mandible owing to pressure from the mass was noted. The specimen was submitted to pathology for final review (Fig. 3). On the first postoperative day, the patient was discharged and experienced an uneventful postoperative course.

At 1 week, the patient returned to our department for his postoperative evaluation with no complaints. Final report from the pathologist described sinus histiocytosis with massive lymphadenopathy, with numerous phagocytized acute inflammatory cells within the cytoplasm of the histiocytes. Immunohistochemical staining with anti-S100 protein antibody showed a strong cytoplasmic and nuclear positive reaction; CD 68 tested positive as well. This, combined with large cells demonstrating emperipolesis, is consistent with RDD (Fig. 4).

Fig. 1. Patient upon presentation to our clinic with a large rightsided mass in level II; a smaller mass is noted further posterior.

Fig. 2. Panoramic radiograph shows coronal radiolucency affecting tooth #30 with no periapical pathology present.

Fig. 3. Specimen sectioned displaying affected node attached to submandibular gland.

Fig. 4. (A) Lymph node displaying emperipolesis, histiocytes containing neutrophils within the cytoplasm (arrow; hematoxylin-eosin stain; magnification ×400). (B) Immunohistochemical staining with S-100 displaying positive cytoplasmic and nuclear reaction (magnification ×400).
DISCUSSION

RDD is clinically characterized as massive, painless, bilateral symmetric cervical lymphadenopathy, with fever and leukocytosis. This disease displays a predilection for males, which is slightly more prevalent in those of African descent. Usually presenting in the first and second decades of life, patients may complain of fever and recent weight loss, and display leukocytosis and neutrophilia. Aside from the cervical nodes, RDD is known to occur in mediastinal, hilar, retroperitoneal, axillary, and inguinal nodes as well. In more than half of all patients with nodal SHML, extranodal involvement is also present, with most affected individuals experiencing lesions in the head and neck region.1,4,5

Histopathologically, SHML displays enlarged lymph nodes with dilatation of the subcapsular and medullary sinuses. Cells within the sinuses are mostly non-neoplastic histiocytes, often containing phagocytosed lymphocytes. This finding is described as lymphophagocytosis or emperipolesis, which is characteristic of RDD.1,6,7

Currently, the etiology of SHML is unknown, but is thought to be a disorder involving the immune response related to a presently unknown pathogen. Efforts to determine a possible pathogen as the cause have been unsuccessful. Foucar et al.8 noted in their review of the SHML registry an association with immune dysfunction, and observed that patients with clinically apparent immunologic disorders have a higher mortality rate. A familial relationship appears to be possible. A registry of 423 cases of SHML established by Rosai noted the entity appeared in 2 sets of twins, 2 other sets of siblings, and 4 individuals reported blood relatives who had masses that were never diagnosed.2,8

Treatment of RDD is usually conservative, observing the patient until spontaneous resolution occurs. One case report discusses a patient who rapidly improved after being placed on acyclovir. Surgical treatment is necessary in instances where there are functional or life-threatening issues. Although surgical treatment is not necessary, it may be advised in cases where the cosmetic deformity is causing psychosocial harm to the patient, as is often the case in children. Nonsurgical treatment may consist of steroids, radiation therapy, chemotherapy, or acyclovir. No single treatment has been proven to be more beneficial than another.

The presentation of this particular case is unique. Whereas most patients with RDD present with bilateral, symmetric, cervical lymphadenopathy, this patient presented with a large unilateral mass with multiple biopsies of necrotic lymph tissue. In this particular case, the patient had no need for further treatment, as the excisional biopsy at 1 year showed no further masses or complications of this disease (Fig. 5).

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REFERENCES


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