Rosai Dorfman Syndrome With Extranodal Manifestation In The Nasal Cavity

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Citation

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Abstract

The term Sinus histiocytosis with massive lymphadenopathy (SHML) was introduced by Rosai and Dorfman in 1969 and is now widely known as Rosai Dorfman disease or syndrome (RDS). It is a benign condition, usually manifesting as massive enlargement of cervical lymph nodes and often confused with lymphoma. Extranodal involvement is observed in 25 to 40 % of cases. This pathology is very rare and involvement of the nasal cavity as an extranodal site is exceptional. A 22-year-old male reported progressive right nasal obstruction accompanied by frequent epistaxis and massive bilateral cervical lymphadenopathy. A FNAC from cervical lymph node and biopsy from nasal mass was compatible with Rosai Dorfman syndrome.

INTRODUCTION

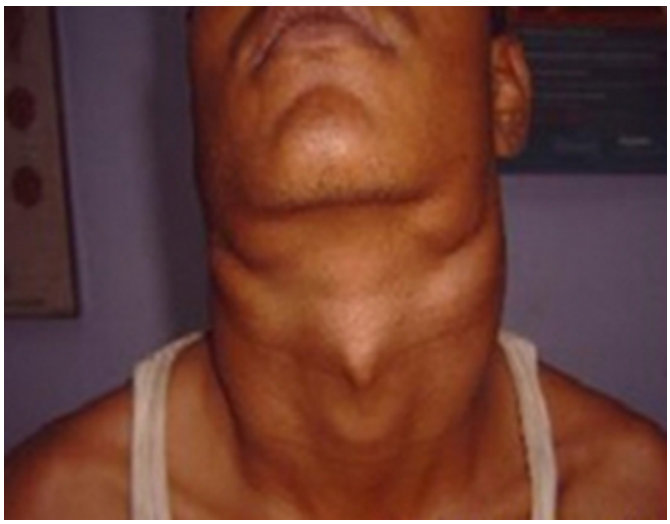
In 1969, Rosai and Dorfman described 4 cases of a disease they called sinus histocytosis with massive lymphadenopathy. Later in 1972, they analyzed 30 additional cases, establishing SHML as a well-defined clinicopathologic entity[1]. RDS is a non-malignant disease generally occurring in children or young adults with massive cervical lymphadenopathy, fever, leukocytosis, an increased erythrocyte sedimentation rate, and hypergammaglobulinemia. Other lymphatic groups, such as mediastinal, axillary and inguinal lymph nodes can also be affected. In about 25 to 40 % of cases, one or more of the following extranodal sites are affected: eyelids, orbit, respiratory tract, salivary glands, skin, bone, testis, lung, kidney, central nervous system, thyroid, and gastrointestinal tract [2,3,4]. Extranodal involvement is often responsible for the most important clinical manifestation of the disease. The cause of RDS has yet to be established. The clinical course of the disease is variable. We report a case of RDS with extranodal involvement exclusively confined to the nasal cavity and pertinent literature is reviewed.

CASE REPORT

A 22-year-old male presented with bilateral, massive, painless neck swelling for past six months. He complained of gradually increasing nasal obstruction on right side with frequent episodes of epistaxis for past three months. On examination bilateral cervical lymph nodes were grossly enlarged in both anterior and posterior triangle which were multiple, firm, non-tender and non-matted with smooth surface (Figure-1).

Figure 1

Figure 1: Clinical photograph of the patient showing bilateral massive cervical lymphadenopathy

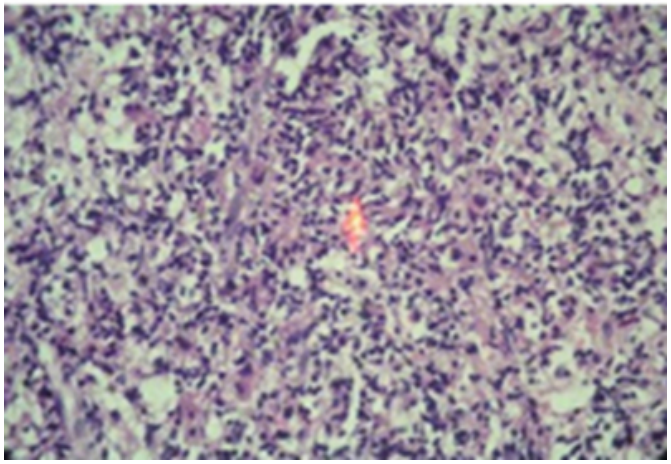


Anterior rhinoscopy showed pinkish, irregular and friable mass filling the right nasal cavity, which was soft, non sensitive and bled when manipulated. Posterior rhinoscopy showed the mass in the right choana. Rest of ENT and systemic examination revealed no abnormality. Investigations showed HB-11 gm/dl, total WBC count -13730/cu mm, Differential WBC count -N 72, L14, E14, ESR- 122 mm 1st/hr. Peripheral blood film showed microcytic, hypochromic red blood cells. X-rays Paranasal

sinuses showed radio-opaque shadow in right nasal cavity with clear sinuses. Chest x-ray and USG scan of the whole abdomen was revealed normal. Nasal endoscopy showed mass present in floor of nasal cavity, middle meatus and corresponding septum area on the right side. A nasal biopsy was obtained under local anaesthesia and histopathology showed subepithelial tissue heavily infiltrated with large numbers of histiocytes,which were having abundant clear foamy cytoplasm containing ingested lymphocytes and other blood cells(emperipolesis) (Figure-2).

Figure 2

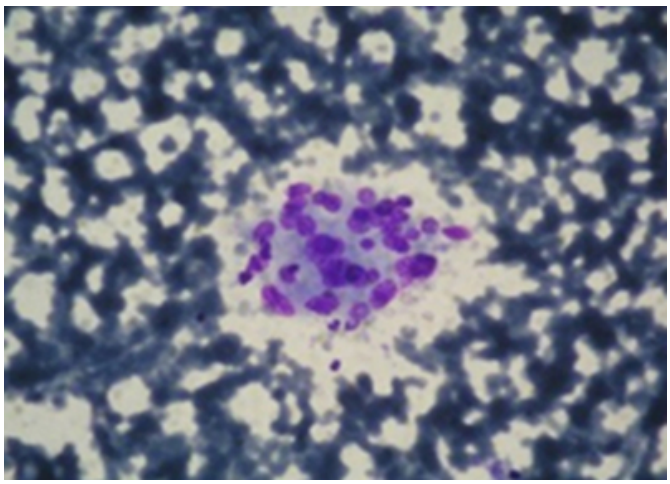
Figure 2: Microphotography of Nasal mass shows respiratory epithelium with subepithelial infiltration of histiocytes containing ingested lymphocytes and other blood cells (emperipolesis).



FNAC of right and left cervical node both showed many small and large lynphocytes, plasma cells, immunoblasts along with many histiocytes. Many of these histiocytes show lymphocytophagocytosis and erythrophagocytosis(emperipolesis)(Figure-3).

Figure 3

Figure 3: Microphotograph (High power) of the lymph node showing characteristic emperipolesis



These features were consistent with a diagnosis of Rosai Dorfman syndrome. Bone marrow examination revealed normal. The patient was advised surgery for nasal mass under general anesthesia and the mass was excised endoscopically. Intraoperatively, the mass was found to have arisen from the nasal septum. The patient was treated with oral prednisolone 60 mg / day for a period of 8 weeks and then tapered off. Four months after surgery, the nasal cavity is free of any mass and the lymph nodes enlargements have regressed in size and are still minimally palpable. The patient is under follow up.

DISCUSSION

RDS is a well-recognized benign syndrome but a rare cause of lymphadenopathy in the third decade of life. It usually presents with cervical lymphadenopathy and should be considered in the differential diagnosis. The disease shows a worldwide distribution and can manifest in any age group. 81% of reported cases occur during the first and second decades of life and has a 2:1 male-to-female ratio[3]. Our patient was a male who reported the occurrence of the disease during the third decade of life. The predominant clinical manifestation of the disease is massive cervical lymphadenopathy (87.3% of cases) that, in most cases, is painless and bilateral, affecting one or all-cervical chains. Lymph nodes are isolated, mobile, and small during the initial stages but becomes adherent with disease progression, forming a voluminous multinodular mass. The axillary (23.7%), inguinal (25.7%), and mediastinal (14.5%) regions can also be affected, but always to a lesser extent than cervical involvement[2]. In our case bilateral cervical group of lymph nodes in anterior and posterior triangle were

involved. Fever occurs in upto 30% of cases but was absent in our patient. Laboratory alterations are frequent and include anemia (65.7%), leucocytosis (59.1%), neutrophillia (68.4%), increased ESR (88.5%) and hypergammaglobulinemia (90%)[2]. Our patient showed anemia, neutrophillia and raised ESR. Extranodal manifestation of the disease is observed in 28-43% of cases, with preference for head and neck region [3,4]. The most common sites of extranodal involvement are skin (27.4%), nasal and paranasal cavities (26.8%), subcutaneous tissue (22.2%), orbit and eyelids (20.1%) and bone (18.4%). In extremely rare circumstances the central nervous system can be affected. Suprasellar involvement-mimicking meningioma has been reported [5]. According to one study the most frequent otorhinolaryngologic manifestations are found in the nasal cavity (50%), pharynx (25%), paranasal sinuses (18.7%), amygdala (12.5%) and trachea (6.3%)[6]. The concomitant involvement of one or more sites in the same individual is observed in up to 44.7% of cases. In 85% of cases, patients with RDS are in good general health without significant symptoms of disease [2]. The patient we have reported presented with extranodal disease confined to nasal cavity with cervical nodal involvement. The patient had nasal obstruction and epistaxis due to the involvement of nasal cavity. The diagnosis of RDS is made on the basis of clinical suspicion and confirmed by histopathology and immunohistochemical studies. In lymph nodes, the sinuses are markedly dilated and crowded with histiocytes, lymphocytes and plasma cells. Histiocytes show abundant foamy cytoplasm, some of which show small lymphocytes in cytoplasm (emperipolesis). Emperipolesis means lymphocytophagocytosis, which defers from phagocytosis in that lymphocytes taken up are not attacked by enzymes and appear intact within the histiocytes. The distinctive histiocytes of RDS that exhibit emperipolesis, their association with numerous plasma cell and the distension of lymph node sinuses or lymphatics in extranodal sites by these cells help distinguish RDS from a variety of benign and malignant disorders in which phagocytosis of cells may be a prominent feature. These findings are less characteristic in the extranodal form of the disease. Immunoreactivity of the histiocytes for S- 100 protein and CD-68 positive large histiocytes displaying lymphocyte phagocytosis are characteristically seen. However, these marker studies were not done in our patient. The pathogenesis of RDS has not been established so far but speculation relates the disorder to be an aberrant response to an unspecified antigen possibly an infective agent. The clinical and microscopic manifestation of RDS should be differentiated from malignant lymphoreticular neoplasias such as Hodgkin's disease and monocytic leukemia, histiocytosis, rhinoscleroma, tuberculosis, juvenile xanthogranuloma, dermatofibromas and eosinophillic granulomas, among others. Depending on the site affected, other pathologies also need to be excluded[7]. The epidemiologic, laboratory and histologic findings are similar among patients with nodal and extranodal involvement suggesting a common biologic basis. In general, extranodal involvement does not determine a more aggressive character or poor outcome; however, generalised lymphadenopathy, extranodal involvement of multiple organs (kidney, lungs and liver) and immunologic alterations lead to a poor prognosis[8]. Even though it is considered a benign disease, fatalities could occur due to cellular infiltrate. In a series of 14 known deaths due to RDS in a registry compromising of 215 patients. The causes of death due to RDS were reported to be a combination of cellular infiltration, mass forming ability of RDS and also may be due to defect in immune function[8]. Most often the RDS takes a benign course and treatment is not necessary. Treatment is necessary only when the disorder becomes life threatening or organ threatening. There is no specific treatment for RDS. The treatment modes include corticosteroids, chemotherapy with a combination of vinca alkaloids and alkylating agents, low dose interferon, antibiotics therapy, radiation therapy and surgical treatment with partial or total resection [9,10]. Surgical option may be reserved for compressive symptoms, like airway obstruction, neurologic or ocular compressions, or severe deformation. However, the best treatment for RDS has yet to be established. In our patient, surgical excision of nasal mass was done with satisfactory result, probably because the disease was restricted and limited to nasal cavity. Cervical lymphadenopathy responded favorably to oral prednisolone and the patient is under follow-up for four month with minimal palpable lymph node in the neck.

CONCLUSION

Extranodal manifestations of Rosai Dorfman Syndrome are rare and range from restricted to highly extensive forms. Because the head and neck region is the preferred site of extranodal form of disease, otorhinolaryngologist should always be aware of Rosai Dorfman Syndrome in making a differential diagnosis.

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