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CASE REPORT

Rosai-Dorfman Disease: A Rare Cause of Cervical Lymphadenopathy

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ABSTRACT

Rosai-Dorfman disease is also known as sinus histiocytosis with massive lymphadenopathy (SHML) is a rare clinicopathological condition. It is a benign condition which causes significant cervical lymphadenopathy in children and young adults. The clinical presentation varies from isolated nodal involvement to significant extranodal manifestations. The clinical features are usually mild, but rarely life-threatening complications can occur in some individuals depending on the site of involvement. Here, we present two cases of Rosai-Dorfman disease, both diagnosed on fine needle aspiration cytology (FNAC) and histopathology and responded well to steroids. One of the patients had extranodal site involvement in the form of bilateral nasal mass which is very rare.

Keywords: Rosai-Dorfman disease, Cervical lymphadenopathy, Nasal mass.

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INTRODUCTION

Rosai-Dorfman disease is a rare histiocytic disorder affecting various groups of lymph nodes in the human body. Rosai and Dorfman in 1969 described this disorder under the term sinus histiocytosis with massive lymphadenopathy (SHML).[1] Majority of patients are children or young adults.[2] Males are more commonly affected.[3] Cervical lymph nodes are most commonly affected;[4] however, other lymphnodal groups, like axillary, inguinal and mediastinal may also be involved. Extranodal involvement is seen in 25 to 40% cases. Various extranodal sites have been reported including the upper respiratory tract, gastrointestinal tract, paranasal sinuses, orbit and even meninges.[4] In most of the cases, patients are usually asymptomatic except for cervical lymphadenopathy; however, they may present with symptoms due to extranodal involvement and infiltration of vital organs. The disease usually has a self-limiting, benign course and may not need any treatment. Episodes of remissions and exacerbations are characteristics; however, few patients may die from their disease.

Etiology is not exactly known however like any other histiocytic disorder responds well to systemic steroids. Histologically, it is characterized by pericapsular fibrosis with dilated sinuses, heavily infiltrated by large histiocytes, lymphocytes and plasma cells. Emperipolesis is characteristic of lymphnodal involvement. Poor prognosis in the disease is due to wide spread dissemination and involvement of vital organs like kidney and liver or presence of immunological abnormalities. Otherwise, the disease has a very stable and benign course.

CASE REPORTS

Case 1

A 12-year-old male presented to us with complaints of slowly progressive bilateral neck swellings since 1 year (Fig. 1). He also complained of bilateral nasal obstruction, insidious in onset and gradually progressive, since last 6 months. It was associated with intermittent nasal bleeds, spontaneous and self limiting. There was no history of fever, pain in throat, difficulty in deglutition, chronic cough, loss of appetite or loss of weight. On examination, there was 3 × 4 cm swelling present on both sides of

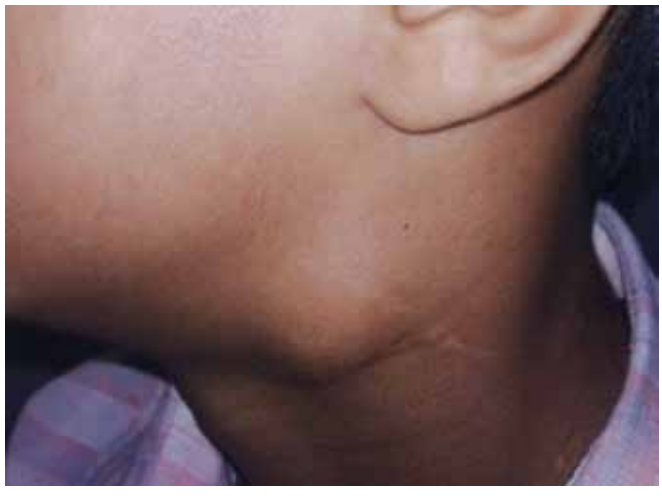

Fig. 1: Patient with left-sided cervical lymphadenopathy (patient had bilateral lymphadenopathy)

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neck involving both anterior and posterior triangles. The swellings were firm in consistency, nontender, nonfluctuant, nonmatted and smooth in outline. The mass was freely mobile and not fixed to skin or underlying structures. Examination of the nasal cavities revealed bilateral pinkish, insensitive mass which bleed on touching. The nasal mass was more prominent on right side compared to left side. Routine hematological examination showed anemia with Hb% of 7 gm%. Total leukocyte count (TLC) and differential leukocyte count (DLC) were within normal limits. Erythrocyte sedimentation rate (ESR) was normal, and Mantoux test was negative. Ultrasonography (USG) neck showed bilateral cervical lymphadenopathy without caseation or matting. Radiograph of chest and ultrasonography of abdomen were within normal limits. Fina needle aspiration cytology from the cervical mass showed lymphocytes, plasma cells and histiocytes showing emperipolesis suggestive of Rosai-Dorfman disease (Fig. 2). Endoscopic biopsy of the nasal mass also showed similar features and was typical of Rosai-Dorfman disease. S-100 stain was done to confirm the diagnosis.

The patient was started on tablet prednisolone 1 mg/kg/day for a period of 2 weeks which was then tapered over next 2 weeks period. The patient was kept on low dose oral steroids for a period of 4 months. He responded very well to the treatment, with both the nasal mass and cervical lymphadenopathy showed drastic reduction in size. The nasal mass almost completely regressed in a months period and the cervical swelling became negligible (Fig. 3). Patient is under regular follow-up and not showing any signs of recurrence.

Case 2

A 10-year-old female child, presented to us with complaints of gradually progressive bilateral cervical swelling since last 8 months. There was no history of pain in the swelling, fever, difficulty in respiration, difficulty in deglutition, throat pain, loss of appetite or loss of weight. Local examination revealed bilateral 6 × 4 cm swellings involving the anterior triangle of the neck. The lesion was firm in consistency, nontender, nonmatted, smooth and freely mobile. Ultrasonography of neck revealed bilateral cervical lymphadenopathy with matting. Radiograph of chest and ultrasonography of abdomen were nonsignificant. Computed tomography (CT) neck was done and it showed bilateral, non-caseating cervical lymphadenopathy involving upper deep cervical nodes. Routine hematological investigations were normal. Mantoux test was negative. Fine needle aspiration cytology from cervical lymph node was highly suggestive of Rosai-Dorfman disease, however biopsy was advised for confirmation. After ruling out tuberculosis, cervical lymph node biopsy was done under general anesthesia and histopathology was diagnostic of Rosai-Dorfman disease. Patient was started of systemic oral steroids 1 mg/kg/day for a period of 1 month and then low dose oral steroids for further 4 months. She responded well to treatment with drastic reduction in size of the swelling. She is under regular follow-up and doing well.

DISCUSSION

Rosai-Dorfman disease is a distinct benign histiocytic disorder, which presents in younger age group with massive cervical lymphadenopathy. The disease is worldwide in distribution and males are commonly affected with male to female ratio of approximately 2:1. Other lymph node groups, like axillary, mediastinal, inguinal and retroperitoneal, may be involved. Other systemic symptoms include fever, leukocytosis, increased ESR, hypergammaglobulinemia and sometimes anemia. The

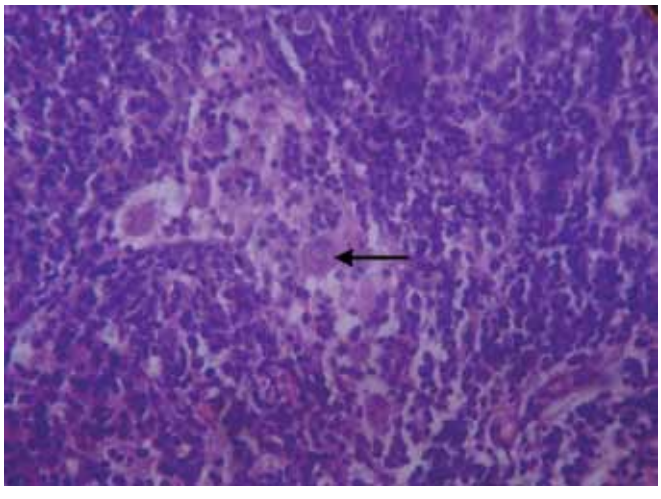

Fig. 2: High magnification histopathology showing the typical emperipolesis and dense lymphocytic infiltrate

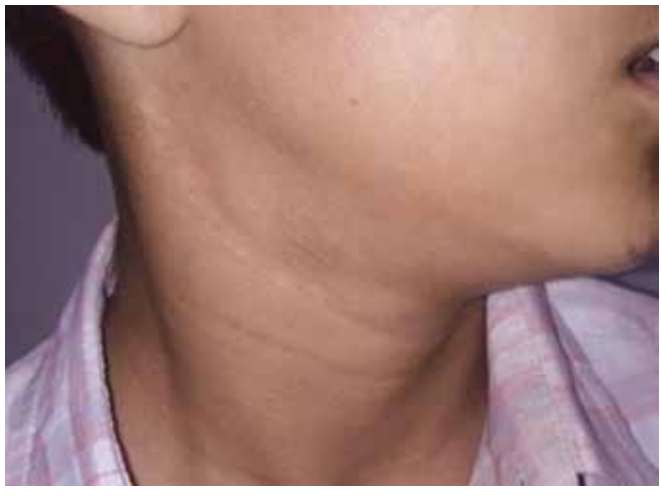

Fig. 3: Complete regression of cervical lymphadenopathy after oral steroid therapy

systemic manifestations may be seen in 60 to 90% patients. The systemic symptoms were not seen in our patients, except for anemia in first case. This finding may be secondary to recurrent epistaxis due to nasal mass. Extranodal involvement is seen in 25 to 40% and commonly involved extranodal sites include skin, subcutaneous tissue, respiratory system, genitourinary system, bones, orbit, central nervous system and breasts.[4] Rarely, in less than 20% cases, isolated extranodal involvement without lymphadenopathy is seen. Head and neck region is involved in 22% cases, nasal cavity being most common.[2] Our patient had extranodal involvement in the form of bilateral nasal mass and nodal involvement in the form of bilateral cervical lymphadenopathy.

Diagnosis of Rosai-Dorfman disease is based on clinical suspicion and histopathological confirmation. Histologically, there is infiltration of the tissue by lymphocytes, histiocytes and plasma cells. Presence of emperipolesis, i.e. engulfment of lymphocytes and erythrocytes by histiocytes, is usually diagnostic of Rosai-Dorfman disease. Immunohistochemistry is usually necessary for confirmation of diagnosis. Characteristically, S-100 is always positive. Also, some other markers, like CD68, CD163, α1 antichymotrypsin and α1 antitrypsin, may also be positive.[2] Systemic symptoms in this disease may be related to enhanced production of such cytokines.[5] In general, the disease has a benign course and is self limiting. However, massive lymphadenopathy and multisystemic involvement, especially vital organs, like central nervous system (CNS), liver, kidney and lungs, are usually associated with poorer prognosis.

The treatment in Rosai-Dorfman disease is nonspecific and depends on the site of involvement. Isolated lymphadenopathy may not be treated at all except for cosmetic reasons. However, if any vital organs are involved or if the lesion is causing some obstructive symptoms or pressure symptoms, aggressive treatment may be indicated. The medical treatment includes corticosteroids, chemotherapy, low dose interferon, antibiotics and radiation therapy. But, response to treatment is highly variable with repeated remission and exacerbation episodes. Surgical treatment may be in the form of partial or total resections. However, surgery is usually limited to biopsy for confirmation. Debulking or excision may be reserved for compressive symptoms involving the upper respiratory tract, orbit or CNS. However, the best treatment for Rosai-Dorfman disease is yet to be established. In our patients, surgery was limited to diagnostic biopsy and both our patients responded very well to systemic steroids.

CONCLUSION

Rosai-Dorfman disease should be kept as a differential diagnosis in young patients presenting with massive cervical lymphadenopathy. High degree of clinical suspicion with typical histopathological features and immunohistochemistry are diagnostic. These patients respond well to systemic steroids and aggressive surgical treatment is needed only in life-threatening complications. However, it is necessary that both clinicians and pathologist keep this entity in their list of differential diagnosis for massive lymphadenopathy.

REFERENCES

1. Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy. Arch Pathol 1969 Jan;87(1):63-70.
2. Juskevicius R, Finlay JL. Rosai-Dorfman disease of the parotid gland, cytologic and histopathologic findings with immunohistochemical correlation. Arch Pathol Lab Med 2001; 125:1348-1350.
3. Lauwers GY, Perez-Atayde A, Dorfman RF, et al. The digestive system manifestations of Rosai-Dorfman disease (sinus histiocytosis with massive lymphadenopathy): review of 11 cases. Hum Pathol 2000 Mar;31(3):380-385.
4. Foucar E, Rosai J, Dorfman R. Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease): review of the entity. Semin Diagn Pathol 1990 Feb;7(1):19-73.
5. Foss HD, Herbst H, Araujo L, et al. Monokine expression in Langerhans' cell histiocytosis and sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease). J Pathol 1996 May;179(1):60-65.