Rosai - Dorfman Disease – A Rare Cause of Cervical Lymphadenopathy

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Case Report

Abstract: Rosai-Dorfman Disease also known as Sinus Histiocytosis with Massive Lymphadenopathy (SHML) is a rare clinic-pathological 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 extra-nodal 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 FNAC and Histopathology and responded well to steroids. One of the patients had extra nodal site involvement in the form of bilateral Nasal mass which is very rare.

Keywords: Rosai-Dorfman Disease, Cervical Lymphadenopathy, Nasal mass.

Introduction

Rosai-Dorfman Disease is a rare histiocytic disorder affecting various groups of lymphnodes in the human body. Rosai and Dorfman in 1969 described this disorder under the term Sinus Histiocytosis with Massive Lymphadenopathy (1). Majority of patients are children or young adults (2). Males are more commonly affected (3). Cervical Lymphnodes are commonly affected; however other lymphnodal groups like axillary, inguinal and mediastinal may also be involved. Extra-nodal involvement is seen in 25-40% 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 asyptomatic 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 characteristic 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 characterised by pericapsular fibrosis with dilated sinuses, heavily infiltrated by large hitiocytes, 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.

Cases

Case 1

A 12 yrs old male, presented to us with complaints of slowly progressive bilateral neck swellings since one year. 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 3X4 cm swelling present on both sides of neck involving both anterior and posterior triangles. The swellings were firm in consistency, non-tender, non-fluctuant, non matted 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 rt side compared to left side. Routine haematological examination showed anemia with Hb% of 7 gm%. TLC and DLC were within normal limits. ESR was Normal and Montoux Test was negative. USG Neck showed bilateral cervical Lymphadenopathy without caseation or matting. Radiograph of Chest and Ultrasonography of Abdomen were within normal limits. FNAC from the Cervical mass showed lymphocutes, plasma cells and histiocytes showing emperipolesis s/o Rosai Dorfman Disease. Endoscopic biopsy of the nasal mass also showed similar features and was typical of Rosai-Dorfman Disease. S100 stain was done to confirm the diagnosis.The patient was started on Tab. Prednisolone 1mg/kg/day for a period of 2 weeks which was then tepered 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 month's period and the cervical swelling became negligible. Patient is under regular follow up and not showing any signs of recurrence.

Case 2

A 10 yrs 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 6X4cm swellings involving the anterior triangle of the neck. The lesion was firm in consistency, non-tender, non-matted, smooth and freely mobile. Ultrasonography of neck revealed bilateral cervical Lymphadenopathy with matting. Radiograph of chest and Ultrasonography of abdomen were non-significant. CT neck was done and it showed bilateral, non-caseating cervical lymphadenopathy involving upper deep cervical nodes. Routine haematological investigations were normal. Montoux Test was negative. FNAC from cervical Lymphnode was highly suggestive of Rosai-Dorfman Disease; however biopsy was advised for confirmation. After rulling out Tuberculosis, Cervical Lymphnode Biopsy was done under General Anaesthesia and histopathology was diagnostic of Rosai-Dorfman Disease. Patient was started of systemic oral steroids 1mg/kg/day for a period of one 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 followup 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 ration of approximately 2:1. Other lymphnode groups like axillary, mediastinal, inguinal and retroperitoneal may be involved. Other systemic symptoms include fever, leucocytosis, increased ESR, hypergammaglobulineamia and sometimes anaemia. The systemic manifestations may be seen in 60-90% patients. The systemic symptoms were not seen in our patients, except for anaemia in first case. This finding may be secondary to recurrent epistaxis due to nasal mass. Extranodal involvement is seen in 25 – 40% and commonly involved extra-nodal sites include skin, subcutaneous tissue, respiratory system, genitor-urinary system, bones, orbit, central nervous system and breasts (4). Rarely, in less than 20% cases, isolated extra-nodal involvement without lymphadenopathy is seen. Head and neck region is involved in 22% cases, nasal cavity being most common (2). Our patient had extra-nodal involvement in the form of bilateral nasal mass and nodal involvement in the form of bilateral cervical Lymphanednopathy. 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. Demointration 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 S100 is always positive. Aiso 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 CNS, Liver, Kidney and lungs is usually associated with poorer prognosis. The treatment in Rosai-Dorfman Disease is non-specific 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 diffential 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.

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