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Case report

ROSAI-DORFMAN DISEASE WITH CERVICAL LYMPHADENOPATHY AND ORBITAL INVOLVEMENT: A CASE REPORT

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ABSTRACT

Rosai-Dorfman disease (RDD), which is also called as sinus histiocytosis with massive lymphadenopathy (SHML), is a rare histiocytic disorder which occurs due to the over-production of non Langerhans sinus histiocytes. It is a nonmalignant disorder that most frequently affects children and young adults and typically presents with fever, night sweats, nonpainful cervical lymphadenopathy, leukocytosis and an elevated ESR. Extranodal involvement may also occur, thus a variety of organs in the body can be affected. Although some viral etiology has been implicated, the disease generally is considered to have an unknown cause. RDD can often be misdiagnosed as lymphoma, leukemia or tuberculosis, so it is imperative to distinguish it from these conditions as well as other forms of histiocytosis because of difference in the modes of treatment. Diagnosis of Rosai-Dorfman disease is based on biopsy of affected tissue. Biopsy showing the presence of emperipolesis, or the engulfment of lymphocytes and other immune cells by histiocytes that express S-100 antigen is diagnostic of Rosai-Dorfman disease. Once diagnosed, further workup including imaging studies are undertaken in order to determine the extent of the disease. In majority of cases, the disease resolves on its own however, treatments including corticosteroids, chemotherapy, surgical treatment or radiotherapy are carried out in severe or persistent disease or when organ function is at stake (e.g. breathing obstruction, kidney failure, visual problems). The case we report is that of a 16 year old girl who presented with a 6 month history of gradual onset drooping of left upper eyelid with mild proptosis of the left eye alongwith mild drooping of right upper eyelid, low grade fever, night sweats and cervical lymphadenopathy. Blood workup showed increased ESR, CT scan of orbits showed superior orbital masses and diagnostic biopsy revealed Rosai-Dorfman disease.

Keywords: Rosai Dorfman disease, Cervical lymphadenopathy, Emperipolesis, Histiocytes, Proptosis

INTRODUCTION

Rosai-Dorfman disease (RDD) or sinus histiocytosis with massive lymphadenopathy (SHML), is a rare disease resulting from increased production of non Langerhans sinus histiocytes.^[1,2] Rosai and Dorfman first described this disease in 1969.^[3] Lymph nodes are commonly involved but it can involve any organ.^[1] Its main features are painless cervical lymphadenopathy, fever and elevated ESR.^[4] Different sites of extranodal involvement have been reported, particularly the skin, orbit, and upper

respiratory tract.^[5, 6] Central nervous system involvement with no nodal disease has also been reported.^[7] RDD mostly occurs in children and young adults with a slight male predominance and an increased incidence in African descent.^[8] There are some viruses thought to be involved in the pathogenesis of RDD however, the disease is considered to have an unknown cause^[9, 10, 11] The presentation of RDD resembles a number of different conditions including leukemia, lymphoma, TB and

other causes of histiocytosis. The definitive way to diagnose this disease is via a biopsy of the affected tissue. The disease is usually a self-limiting one however, on case to case basis depending upon the organ involvement and severity of the disease, different treatment modalities ranging from medical to surgical or radiotherapy can be instituted to fight this disease. [12, 14, 15]

The purpose of this case report is to highlight this rare disease that we have encountered and to educate clinicians about the fact that the disease can present with a variety of symptoms and can involve different organs that may often distract or misguide clinicians into making a wrong diagnosis.

CASE REPORT

A 16 year old girl presented to the outpatient department of Khyber Teaching Hospital, Peshawar, Pakistan in February 2015 with a six month history of gradual onset, painless left upper eyelid drooping along with low-grade fever and night sweats plus a 20 day history of gradual onset right upper eyelid drooping. She also noticed a few lumps in her cervical region. She had no significant past medical history of any major illness and no family history of tuberculosis or blood disorders was found. Her vitals were as follows, BP 120/80 mm Hg, pulse 90/min, respiratory rate 15/min and temperature 100.2 F. On examination she had bilateral painless cervical lymphadenopathy and bilateral superior orbital masses on palpation. Her vision was 6/6 in both eyes. Ptosis was seen in both right (3mm) and left (5mm) eyes. Mild left eye proptosis was also seen. Extraocular movements were restricted in upper gaze of both eyes, more so of the left eye. There was no evidence of any visceromegaly and the rest of the general physical and systemic examinations were unremarkable. Lab investigations: Hb 11.1 g/dl, RBC 4.27 million/cmm, Hct 32.4 %, MCV 76 fl, MCH 25.9 pg, MCHC 34.2 g/dl, Platelet count 328000/cmm, TLC 11000/cmm, normocytic normochromic picture with DLC showing 80% neutrophils, 15% lymphocytes and 5% monocytes on peripheral smear, ESR 70 mm/1st hour, negative HbS and HCV screening, negative PPD and sputum AFB, normal Liver function tests and normal Renal function tests. Chest X-ray was normal, U/S and CT scan of the abdomen and pelvis was normal. CT scans

of the orbits showed bilateral superior orbital masses (Fig 1) and mild proptosis of the left eye (Fig 2). Incisional biopsy of the left Superior orbital mass was performed that revealed the diagnosis of Rosai-Dorfman disease.

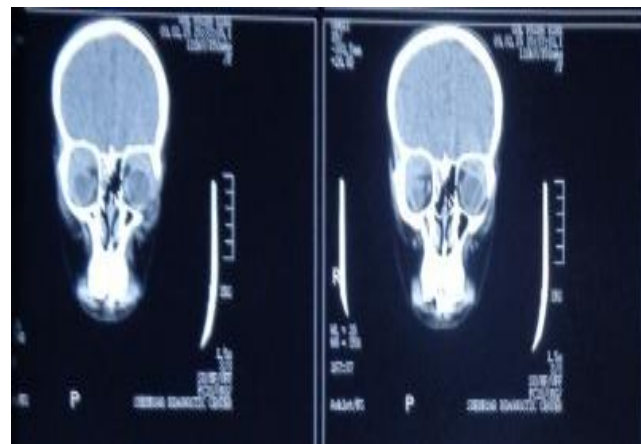


Fig 1: CT scan orbit (Coronal View): Bilateral Superior Orbital Masses

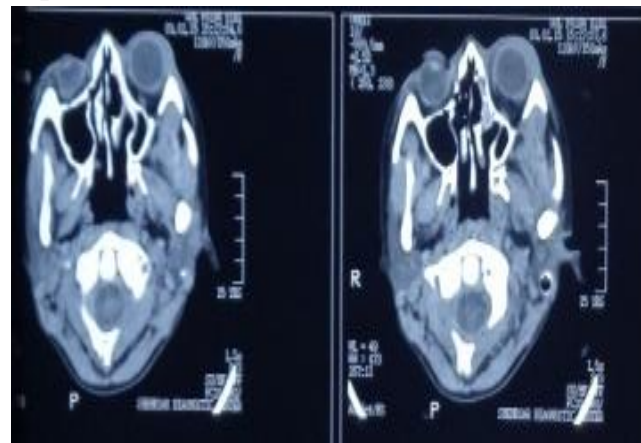


Fig 2: CT scan Orbit (Axial View): Mild Proptosis of the Left Eye

Treatment: The patient was counseled by the attending physician about the nature of the disease and administered Inj. Methylprednisolone 1gm x OD for 3 days followed by Tab Prednisolone 1mg/Kg body weight x OD and advised follow up after 4 weeks. On follow up visit, examination showed that her ptosis and cervical lymphadenopathy had improved. She was also assessed for side effects of steroid therapy. No side effects were noted. She was advised follow up after 8 weeks.

DISCUSSION

Rosai-Dorfman disease (RDD), which is also called as sinus histiocytosis with massive lymphadenopathy (SHML), is a rare histiocytic disorder which occurs due to the over-production of non Langerhans sinus histiocytes. [1, 2] Rosai and Dorfman were the first

people to identify this disease as a unique clinicopathologic condition in 1969. [3] RDD commonly affects lymph nodes however, it has the propensity to involve any organ. [1] Cardinal features include painless cervical lymphadenopathy, fever and elevated ESR. [4] Extranodal involvement has been reported in diverse anatomic sites, particularly the skin, orbit, and upper respiratory tract. [5, 6] Central nervous system involvement without nodal disease has also been reported. [7] Rosai-Dorfman disease though quite rare, is distributed worldwide with 80% cases occurring in children and young adults with a slight male predominance (58%) and has a general predilection for individuals with African descent. [8] The etiology of RDD is unknown, however certain viruses like Human Herpes virus 6 and Epstein-Bar virus via causing immune dysregulation have been implicated in the pathogenesis of this disease. [9, 10, 11] The diagnosis of Rosai-Dorfman disease is not easy since its presentation can mimic a number of other non-malignant as well as malignant conditions ranging from bacterial or viral infections to malignancies including leukemia and lymphoma. Biopsy of the lymph node or affected tissue is required for the diagnosis of this disease. Proliferating S100 and CD68 antigens positive histiocytes exhibiting emperipolesis i.e. phagocytosis of intact lymphocytes and other immune cells, is the classical histologic finding on biopsy in Rosai Dorfman disease.

No specific treatment protocol is established for Rosai Dorfman disease because the disease is rare and its course is mostly self limiting. [14] However, patients with severe, persistent disease or in cases where organ function is compromised steroid therapy, chemotherapy, surgical resection or radiotherapy can be instituted with varying success rates. [12,13, 14, 15]

CONCLUSION

Rosai-Dorfman disease shares many of its presenting features with leukemia, lymphoma, tuberculous lymphadenitis and other causes of histiocytosis, so it should be considered in the differentials of patients, especially children and young adults who present with painless cervical lymphadenopathy. It is also important for physicians to recognize that the disease can have a myriad of clinical manifestations depending upon the tissue involved, as was the case in our patient we presented in this case report having

cervical lymphadenopathy with orbital involvement in whom lymphoma, leukemic deposits, orbital pseudo tumor, Langerhan's cell histiocytosis and hemangioma were also amongst the list of differentials until biopsy confirmed the diagnosis of RDD. It is essential for pathologists as well to look for the histopathologic features of this disease in biopsy specimens, since if promptly diagnosed and managed, can reduce unnecessary diagnostic workups and mismanagement due to misdiagnosis of this disease.

Conflict of Interest: Nil

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