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Case Report

Rosai Dorfman Disease Mimicking Lymphoma: A Case Report

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ABSTRACT

Rosai Dorfman Disease also known as Sinus Histiocytosis with massive lymphadenopathy is a very rare and benign self-limiting histiocytic proliferative disorder with unknown etiology. It typically manifests as massive painless bilateral cervical lymphadenopathy with or without extranodal involvement. It is mostly seen in young adults and children with a slight predilection for males. Clinically patients may be mistaken for lymphoma and other infectious disorders like tuberculosis which is more prevalent in developing countries like in India. Here we report a case of 40 yrs male who presented with massively enlarged cervical lymph nodes mimicking lymphoma. FNAC was performed and the patient was diagnosed with Rosai Dorfman Disease. This case is being reported for its rarity and its presentation in middle aged man.

Keywords: Rosai Dorfman Disease, Sinus Histiocytosis, emperipolesis, massive lymphadenopathy.

INTRODUCTION

Rosai Dorfman Disease (RDD) originally described as Sinus Histiocytosis with Massive Lymphadenopathy (SHML) is a very rare disorder with unknown etiology. It is a benign proliferative disorder of histiocytes. It typically manifests as massive painless bilateral cervical lymphadenopathy with or without extra nodal involvement. [1] These cases can be clinically misdiagnosed as lymphoma. It may occur at any age but mostly seen in young adults and children with a slight predilection for males (58%) and for individuals of African descent. Castleman's disease, dermatopathic lymphadenitis, mucocutaneous lymph node syndrome (Kawasaki's disease), histiocytic necrotising lymphadenopathy (kikuchi's disease), vascular transformation of lymph nodes and inflammatory pseudotumor of the lymph node are some other rare causes of lymph node enlargement. [2-5]

CASE REPORT

A 40 years male presented with painless bilateral submandibular swelling since 3 months. There was no history of fever, weight loss, night sweats or pruritus. There was no history of tuberculosis contact. The past history, personal history and family history of the patient was not significant. On examination patient had multiple, non-tender, firm and rubbery lymphadenopathy in submandibular region which were free from superficial and deep structures. There was no sign of bleeding, gum hypertrophy or bone pain. No hepatosplenomegaly or ascites found. General physical examination was within normal limits. Based on clinical features a diagnosis of lymphoma was suggested. Initial clinical diagnosis was lymphoma. Blood examination of the patient revealed haemoglobin 11.5 gm%, packed cell volume 39%, reticulocyte count 1%, total leucocyte

count 9,300/ mm3 with neutrophil 76%, lymphocyte 20%, eosinophil 2% and monocyte 2%, platelet count 240,000/mm3 and erythrocyte sedimentation rate 54 mm/hour. Red blood cell morphology was normal. Routine urine examination and culture were normal. Thyroid function test was within the normal limit. Ultrasonography revealed submandibular lymphadenopathy with hypoechoic attenuated echotexture.

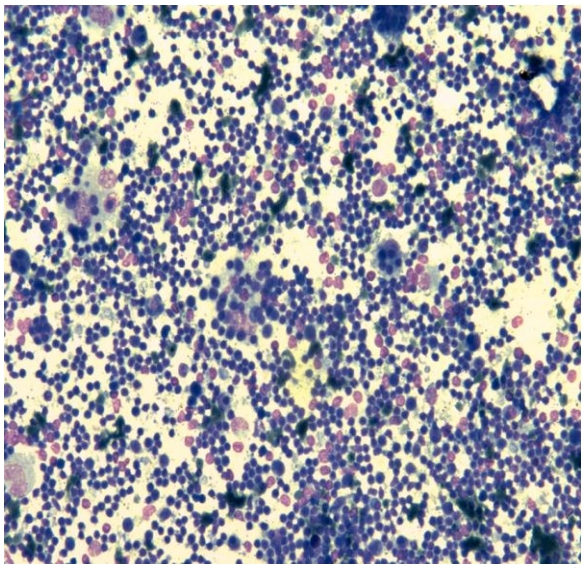

Fig. 1: (200 xs) shows lymphoid cells in various stages of maturation with abundance of macrophages comprising cystic change with marked phagocytic activity admixed with blood.

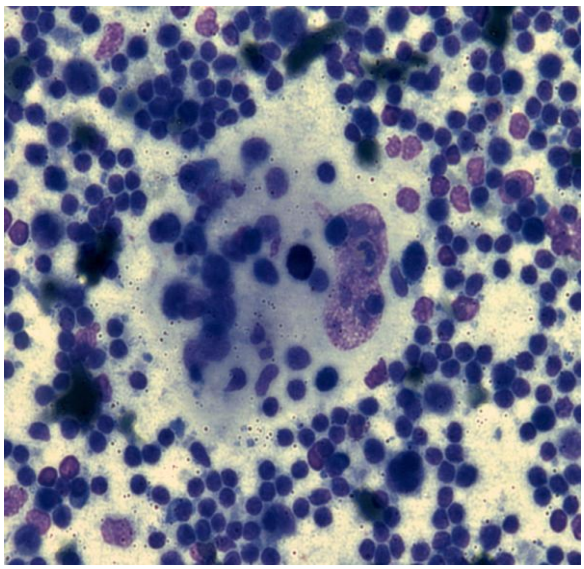

Fig. 2: (400 xs) shows macrophage revealing binucleation and ingested intact lymphocytes and plasma cells (emperipolesis).

FNAC was performed which revealed lymphoid cells in various stages of maturation with abundance of macrophages comprising binucleation, multinucleation and cystic change (Fig. 1) with marked phagocytic activity ingesting lymphocytes and plasma cells (emperipolesis) admixed with blood (Fig. 2). Cytological features were suggestive of Rosai Dorfman Disease (Sinus Histiocytosis with Massive Lymphadenopathy). Later on lymph node biopsy confirmed the diagnosis of RDD.

DISCUSSION

RDD is a rare disease that was initially reported by Juan Rosai and Ronald F Dorfman in 1969. It is a histiocytic proliferative disorder of unknown etiology. Extranodal involvement is seen mainly in the head and neck regions. [2-5] It may involve the skin, orbits, eyelids, salivary glands, upper respiratory tract, peritoneum, bone, kidneys, testis and central nervous system.

The etiology of RDD remains unknown; the two most likely possibilities are being infection by a virus or some other microorganism and the manifestation of a subtle undefined immunologic defect. Suggestion has been made that stimulation of monocytes-macrophage via M-CSF leading to immune suppressive macrophages may be the main pathogenic mechanism of RDD. Release of cytokines like TNF-α from these cells is responsible for the genesis of fever and other systemic symptoms. [11] It has also been suggested that it may be due to an unusual response to Klebsiella infection. [6]

Differential diagnosis includes nonspecific sinus hyperplasia; Langerhans cell histiocytosis, hemophagocytic syndrome, tuberculosis, lymphoma and metastatic malignant melanoma. Massive cervical lymphadenopathy is the hallmark of RDD. Clinically patients may be mistaken for lymphoma and other infectious disorders like tuberculosis which is more prevalent in developing countries like in India.

FNAC is a useful and reliable tool for the diagnosis of RDD and the biopsy can be avoided in these patients. [7] Cytology usually reveals numerous large histiocytes

with abundant pale cytoplasm and phagocytosed lymphocytes (emperipolesis). In emperipolesis, the lymphocytes are not attacked by enzymes and appear intact within the histiocytes (in contrast to phagocytosis). The phenomenon of emperipolesis is highly useful for the diagnosis of RDD using FNAC. Clinicians and pathologists require a high degree of suspicion to diagnose this rare, benign and self limiting disease.

RDD is considered to be benign and self-limiting. In approximately 50% of the patients, the disease resolves without appreciable sequelae, one third have residual asymptomatic adenopathy and 17% have persistent symptomatology even after few years. Regression is usually heralded by diminution of extranodal disease. Surgical debulking may be necessary in the presence of vital organ compression. [8,9] Chemotherapy is in general ineffective, while radiotherapy has shown limited efficacy. [10] Others have also advocated the use of long term prednisolone. [9]

REFERENCES

1. Das DK, Gulati A, Bhatt NC, Sethi RG. Sinus histiocytosis with massive lymphadenopathy (Rosai Dorfman Disease): Report of two cases with fine needle aspiration cytology. Diagn Cytopathol 2001; 24: 42-45.
2. Drosos AA, Georgiadis AN, Metafratzi ZM, et al. Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease) in a patient with primary Sjogren's syndrome. Scand J Rheumatol 2004; 33:119-22.
3. Perry BP, Gregg CM, Myers S, et al. Rosai-Dorfman disease (extranodal sinus histiocytosis) in a patient with HIV. Ear Nose Throat J 1998; 77:855-8.
4. Goodnight JW, Wang MB, Sercarz JA, Fu YS. Extranodal Rosai- Dorfman disease of the head and neck. Laryngoscope 1996; 106:253-6.
5. Pelletier CR, Jordan DR, Jabi M. Bilateral eyelid edema: an uncommon presentation of Rosai-Dorfman disease. Ophthal Plast Reconstr Surg 1999; 15:52-5.
6. Lampert F. and Lennert K: Sinus histiocytosis with massive lymphadenopathy. Fifeteen new cases. Cancer, 1976; 37:783-789.
7. Deshpande AH, Nayak S, Munshi MM. Cytology of sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease) by fine needle aspiration cytology. Diagn Cytopathol 2000; 22:181-5.
8. Dodson KM, Powers CN, Reiter ER, Rosai Dorfman disease presenting as synchronous nasal and intracranial mass Am J Otolaryngol 2003Nov-Dec; 24 (6),USA.
9. Raveenathiran V, Dhanalakashmi M, Hayavadana Rao PV, Viswanathan P. Rosai Dorfman Disease: report of a 3 year old girl with critical review of treatment options. Eur J Pediatr Surg 2003; 13(5):350-4.
10. Kushwaha R, Ahluwalia C, Sipayya V. Diagnosis of Sinus Histiocytosis with massive lymphadenopathy (Rosai-Dorfman Disease) by fine needle aspiration cytology. J Cytol 2009; 26 (2): 83-5.
11. Iyer V K, Handa K K, Sharma M C. Variable extent of emperipolesis in the evolution of rosai dorfman disease. Diagnostic and pathogenetic implication. J Cytol 2009; 26:111-6.

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