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Case Report

Rosai Dorfman disease: a case report with nodal involvement

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ABSTRACT

Rosai Dorfman disease is characterised by sinus histiocytosis with massive lymphadenopathy. Here, we present a case report of 33-year female with low grade fever and multiple swellings of variable size in cervical region. Laboratory examinations showed erythrocyte sedimentation rate to be increased up to 40 mm / hour. Histopathological examination revealed a number of histiocytes containing lymphocytes and few plasma cells in their cytoplasm showing emperipolesis of plasma cells and lymphocytes. Immunohistochemistry showed positivity for CD68 with marked sinus expansion by histiocytic cells and emperipolesis. Treatment with tapering dose of systemic corticosteroids over 1 month gave excellent results.

Keywords: Emperipolesis, Rosai Dorfman, Histiocytosis, CD68, Corticosteroids

INTRODUCTION

Rosai Dorfman disease also known as sinus histiocytosis is a rare, non-neoplastic disorder that is caused by histiocytic proliferation within sinusoids and is usually self-limiting.[1] It typically occurs in the first two decades of life as painless cervical lymphadenopathy and fever. There is extra nodal involvement in 1/3rd of cases. Skin lesions are found in approximately 10 % of cases with predilection for the eyelids and the malar area. It is characterized by abundant histiocytes in the lymph nodes throughout the body. Most patients are young adults, but it has a wide age distribution from birth to 74 years. The disease is slightly more common in males. (58%).[2] On immunohistochemistry, the cells in Rosai Dorfman disease are CD68 and S100 positive.[3]

Emperipolesis is a common histopathological finding. The most affected organs are lymph node and skin, but multiple organs can be affected at the same time. The treatment response to steroids is very diversified and clinical course is very unpredictable.

The pathologic findings that Rosai and Dorfman described became the basis for a new entity called 'sinus histiocytosis with massive lymphadenopathy. These pathologic findings were: emperipolesis -histiocytes with lymphocytes (most common), plasma cell or erythrocytes inside them, marked fibrosis of the lymph node capsule with lymphocyte and plasma cell infiltration, dilation of subcapsular and medullary septate, increased intra-sinusoidal histiocytosis with little atypia and few mitoses.

There were about 1000 case descriptions in the medical literature till now.[4]

CASE REPORT

In the current study, we present a 33-year-old female patient who presented in ENT Outpatient Department of

Sir Sunder Lal Hospital, BHU, Varanasi with a 1-year history of multiple cervical masses and with occasional low-grade fever. On physical examination we found bilateral cervical lymphadenopathy with diffuse swelling in left side of neck.

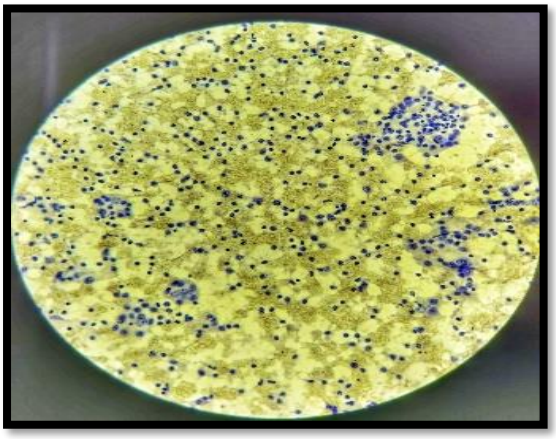

Figure 1: FNAC showed emperipolesis (Giemsa stain ×200 a number of histiocytes containing lymphocytes and few plasma cells in their cytoplasm showing emperipolesis of plasma cells and lymphocytes).

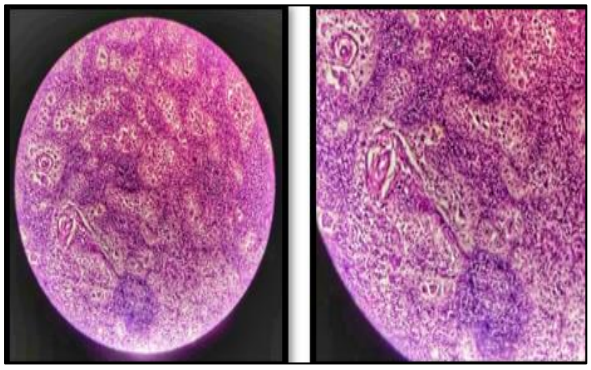

Figure 2: H and E staining shows reactive population of lymphoid cells admixed with many histiocytes showing emperipolesis of plasma cells and lymphocytes.

The swelling on left was diffuse, firm, mobile, tender and there was redness on overlying skin. Other swellings were approx. 2×2 cm, non-tender, mobile present at level 2b and posterior triangle of neck on right side. There was no difficulty in breathing or swallowing.

Laboratory tests showed haemoglobin (Hb)-12.8 gm/dl, erythrocyte sedimentation rate (ESR) -40 mm/hr.

The serum lipid levels and chest X-ray were normal.

As the initial suspected diagnosis was a probable tuberculosis (in Indian scenario) a fine needle aspiration cytology was performed on left cervical lymph node present in posterior triangle after 1 week of proper antibiotic course.

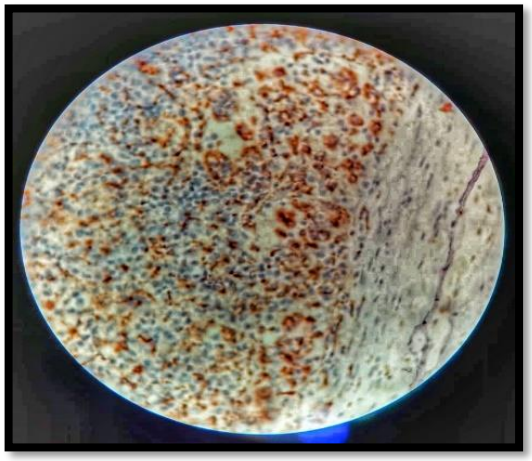

Figure 3: IHC for CD68 is positive in histiocytic cells. Section shows marked sinus expansion by histiocytic cells with pale eosinophilic cytoplasm. Emperipolesis is seen in histiocytes.

Figure 4: Clinical photograph at presentation.

Figure 5: After 2 weeks of antibiotic and steroid therapy.

Lymph node biopsy was performed to confirm the diagnosis and start appropriate treatment, histology

showed lymph nodes with thickened capsule, paracortical hyperplasia and vascular proliferation.

Figure 6: After 4 weeks of steroid therapy.

The histiocytes have round to oval nuclei, and in places small eosinophilic nucleoli. The cells have abundant, pale cytoplasm and many contain intact plasma cells and lymphocytes (emperipolesis). Emperipolesis is the presence of an intact cell within the cytoplasm of another cell. It is an uncommon biological process, and can be both physiological or pathological.[5]

Emperipolesis is different from phagocytosis where the engulfed cell is killed by lysosomal enzymes of the macrophage. Here, the engulfed cell remains viable, and can exit at any time without any damage in either cell.

Emperipolesis has been classified into two categories:5engulfment of haemopoietic cells by megakaryocytes such as in haematolymphoid disorders (Hodgkin's disease, leukaemia, acute and chronic myeloid leukaemia non-Hodgkin's lymphoma, myeloproliferative disorders, myelodysplastic syndrome), engulfment of inflammatory cells by histiocytes, classic of rosai Dorfman disease.

Immunohistochemistry showed positivity for CD68, which is characteristic for RDD.

Accordingly, we started the patient on corticosteroid therapy and response was good. Patient became asymptomatic in 1 month of corticosteroid therapy with tapering dose.

DISCUSSION

RDD is a histiocytic disorder described in 1965 by Destombes for the first time and later in 1969 by Rosai and Dorfman as 'sinus histiocytosis with massive lymphadenopathy'.[6] In our case, the patient was a 33-year-old female, whereas rosai Dorfman disease has a slight male preponderance. (Male to female ratio of 1.4).[7] Singh et al in 2009 reported Rosai-Dorfman disease in an Indian woman with generalized lymphadenopathy and nasal obstruction.[8] Most of the patients present with mobile, non-tender bilateral cervical lymphadenopathy. The lymph nodes become matted and prominent due to pericapsular fibrosis (caused by infiltration of lymphocytes and plasma cells). The clinical features consist of low-grade fever along with, elevated ESR, anaemia, hyperglobulinaemia and leucocytosis.[9] In our case, there was low grade fever with bilateral cervical lymphadenopathy. No associated anaemia, leucocytosis, or hyperglobulinaemia was seen. The erythrocyte sedimentation rate was high in our case. Histologically, RDD shows small lymphocytes, erythrocytes and plasma cells are engulfed by histiocytes without phagocytosis – termed 'emperipolesis'. This can be identified on routine haematoxylin and eosin stains.[10] The literature describes that 70 % of RDD patients are in stable condition, 20 % are self-limiting and only 10 % results in aggressive local or systemic disease.[11] In very small amount of patients RDD can prove to be fatal, when massive enlarged lymph nodes, with combination of mass formation and cellular infiltration start compressing on vital structures.[12] Surgery is indicated in symptomatic patients or in those with involvement of critical structures.[13] It is often unnecessary to intervene until and unless airways are obstructed or some major organ is compressed.[14] Due to its rare incidence, no standard treatment has yet been defined for RDD. However, the condition is mostly self-limiting.[15] Several forms of therapy have been described involving corticosteroids, chemotherapy combined with alkaloids, anthracyclines, antimetabolic and alkylating agents, interferon, antibiotics, radiotherapy and partial or total surgical resection.[16] In some studies it is seen that 82% of untreated patient have complete disease regression.[18] In our case, after careful analysis of the biopsy specimen, a conservative approach was adopted with progressive reduction of the lymphadenopathy and other symptoms. The course of RDD is unpredictable. Episodes of remission and exacerbation may occur for several years.[19]

Differential diagnosis of RDD includes infections caused by mycobacterium and histoplasma involving lymph nodes, classical Hodgkin lymphoma, melanoma, histiocytosis of Langerhans cells, histiocytic sarcoma, lysosomal storage diseases, metastatic carcinoma. Immunohistochemical staining for S-100 and CD68 is helpful in distinguishing RDD from diseases mentioned above.[20]

CONCLUSION

RDD is a rare disease that has a relatively benign clinical course, it often mimics infectious disease and it can easily be mis diagnosed. Definitive diagnosis is made by histology which shows emperipolesis, sinus dilation and histiocytic cell infiltration. Immunohistochemistry shows positivity for CD68 and S100. Due to its low incidence, no standard treatment has yet been defined for RDD. Because the disease generally resolves spontaneously observation is currently advocated approach however for systemic symptoms steroids and chemotherapy has also

been used for its treatment. There is a prevailing need of better treatment strategy for RDD.

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