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Case Report (Pages: 18984-18987)

Sinus Histiocytosis in a Child: A Rare Case Report

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Abstract

Background: Rosai-Dorfman disease is a self-limiting condition caused by histiocyte proliferation within the sinusoids of lymph nodes and in the extranodal tissue. It is a rare disease, particularly in children, that progresses with extensive lymphadenopathy.

Case Presentation: We report the case of a 5-year-old child who presented with an enlarged cervical lymph node. Fine Needle Aspiration Cytology (FNAC) was done, which was suggestive of 'active lymphoid hyperplasia with sinus histiocytosis'

Conclusion: In this case, after careful analysis of the biopsied specimen, a conservative approach was adopted. The patient was discharged and advised for follow-up. On follow-up, after 2 weeks the size of the lymph node had decreased.

Key Words: Cervical Lymphadenopathy, Non-Langerhans Cells, Rosai-Dorfman Disease, Sinus Histiocytosis.

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1- INTRODUCTION

Sinus histiocytosis with massive lymphadenopathy or Rosai Dorfman disease (RDD) is a benign, uncommon, and self-limiting disease. It is a non-Langerhans cell histiocytosis. It is characterized by non-neoplastic proliferation of histiocytes/phagocytes in the sinusoids of lymph nodes and in extranodal tissues. Although rare in children, RDD can mimic malignant lymphoproliferative disorders (1).

2- CASE REPORT

A 5-year-old male child presented with complaints of swelling over the left side of the neck for 1 month which was associated with pain while eating and during any jaw movement. No history of cough, fever, or significant weight loss. He was diagnosed to have Tubercular lymphadenitis at the age of 6 months and was treated with anti-tubercular drugs for 6 months. His height was 105cm and his weight was 15 kg, with both weight and height having a Z score of - 0 to -2 standard deviations. On examination, his pulse rate was 74/min, respiratory rate was 18/min, Blood pressure - 98/70mmHg, and oxygen saturation was 98%. No hepatosplenomegaly on per abdominal examination was observed. Bilateral air entries were equal on respiratory examination. On cardiac examination, S1 and S2 were normal with no murmur. On local examination: large single cervical lymph node- tender, firm in consistency, well-defined margins, measured 2.5 cm x 2cm. He also had insignificant inguinal lymph nodes on palpation. Fine Needle Aspiration Cytology (FNAC) was done, which was suggestive of ‘active lymphoid hyperplasia with sinus histiocytosis’ (Fig. 1). As there was no other issue, the patient was discharged and advised for follow-up. On follow-up, after 2 weeks the size of the lymph node had decreased.

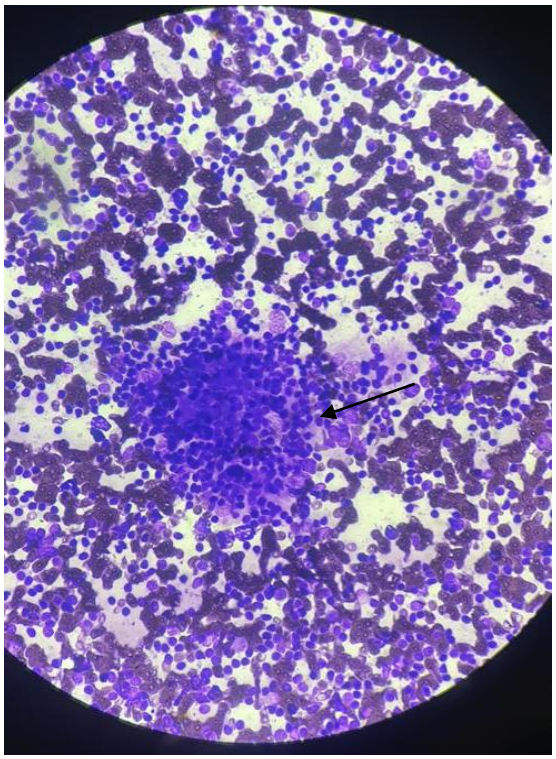

Fig. 1: Active lymphoid hyperplasia with sinus histiocytosis.

3- DISCUSSION

RDD is a rare proliferative histiocytic disease with a benign course. It is found worldwide and affects individuals with an average age of 20.6 years, RDD is slightly more common among men (1.4:1) and is significantly more common among Caucasians and blacks than among Asians (2). In children, it is a rare cause of rapidly progressive lymphadenopathy, which sometimes mimics malignancy. It has a prevalence of around 1:200000 (2-4).

Lima FB et al. (1) reported a case of sinus histiocytosis in a 2-year-old child. Laura LE (5) reported a case of a 4-year-old female patient, who presented with bilateral level V lymphadenopathy of the cervical neck. The disease typically presents with extensive cervical lymphadenopathy, most often bilateral and painless (87%). At first lymph nodes are mobile and discrete, but over time they become adherent and tend to develop into a large multinodular mass (1-3). The axillary (23.7%), inguinal (25.7%), and mediastinal (14.5%) regions may be affected, though not as severely as the cervical region (6). The diagnosis of RDD is based on clinical history and confirmed by histopathological examination. Specimens may be obtained by open surgical biopsy or fine-needle aspiration. The most important histological finding in RDD is histiocytes with a variable number of phagocytosed cells, usually lymphocytes, plasmocytes, or erythrocytes. Some cells, especially lymphocytes, remain viable inside the vacuoles, giving rise to a phenomenon known as lymphophagocytosis or emperipolesis, defined as the presence of intact lymphocytes inside other cells. The most useful marker of histiocytes in RDD is the expression of protein S100 (5-7). Due to its low incidence, no standard treatment has yet been defined for RDD. However, since the condition is self-limiting, it is often unnecessary to intervene, except when the airways are obstructed or vital organs are compressed. Several forms of therapy have been described involving corticosteroids, chemotherapy combined with periwinkle alkaloids, anthracyclines, antimetabolites, and alkylating agents, interferon, antibiotics, radiotherapy, and partial or total surgical resection (7).

4- CONCLUSION

In this case, after careful analysis of the biopsied specimen, a conservative approach was adopted. The patient was discharged and advised for follow-up. On follow-up, after 2 weeks the size of the lymph node had decreased.

5- REFERENCES

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