Rosai Dorfman Disease Presenting as Isolated Cervical Lymphadenopathy

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PRESENTATION OF CASE

A 40-year-old male presented in our surgery OPD with a history of left sided neck swelling since two months. There was no associated history of pain, fever or cough. General physical and systemic examination did not reveal any abnormality. Local examination revealed multiple cervical lymph nodes along the jugular chain, largest measuring 3 cms. His complete blood count and chest X-ray did not reveal any abnormality. FNAC was suggestive of sinus histiocytosis. Patient underwent excision of the lymph nodes under general anaesthesia. Since the submandibular gland also appeared involved on table, decision was taken to excise it as well. Histopathology report revealed multiple foci of dilatation of sinusoids with histiocytes showing engulfment of lymphocytes and plasma cells, thus consistent with Rosai Dorfman disease. The Submandibular Gland was normal. The Patient was managed conservatively. The postoperative recovery was uneventful and subsequent follow ups did not reveal any recurrence.

Isolated painless cervical lymphadenopathy is a common presentation in a surgical OPD accounting for almost 12 - 15 % of all cases¹. Most common differentials in these cases include infections, tuberculosis, lymphoma, and metastatic malignancies. We report a case of Rosai Dorfman disease presenting as isolated unilateral cervical lymphadenopathy in a young male. The diagnosis was made on cytology and later confirmed on histopathology of excision biopsy. The patient was managed conservatively and had an uneventful course.

Rosai Dorfman disease (sinus histiocytosis with massive lymphadenopathy) is a rare benign proliferative disorder of histiocytes, characterised in majority of cases by painless ipsilateral cervical lymph node enlargement and rarely as an extranodal disease^{2,3}. In the setting of other conspicuous differentials, it often presents as a histopathological surprise. This disease often has a benign course and mostly resolves spontaneously. Our case emphasizes on the need for unbiased general approach in the management of cervical lymphadenopathy with the awareness of Rosai Dorfman as a differential diagnosis.

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DIFFERENTIAL DIAGNOSIS

Tuberculosis, lymphoma and metastatic malignancies.¹



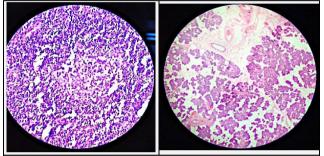


Figure 2. Microscopy: Lymph Node Histiocytosis and Normal Salivary Gland



DISCUSSION

No definitive treatment protocols exist in view of uncertain aetiology and rare occurrence⁴. Most symptomatic cases resolve over time without any treatment⁴. Symptomatic treatment may be required in a few cases.

Rarely, extensive type of Rosai Dorfman may present with generalised painful lymphadenopathy which would require immunosuppressive or systemic chemotherapy⁵.

CONCLUSIONS

Rosai Dorfman Disease is a rare, benign cause of cervical lymphadenopathy. Biopsy is diagnostic in most of the cases. Majority of the cases resolve spontaneously and require just regular follow up. Symptomatic treatment may be required in a few cases, especially with extranodal involvement⁶.

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