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Original Research Paper

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A RARE CASE OF ROSAI-DORFMAN DISEASE IN HYPOTHYROIDISM PATIENT A DIAGNOSTIC COEXISTENCE

Dr. Gridhati Srinivas* Resident, Department of General Medicine, Mahatma Gandhi Medical college and Research Institute, Puducherry, India. *Corresponding Author

Dr. Tarun Kumar Dutta Professor, Department of General Medicine, Mahatma Gandhi Medical college and Research Institute, Puducherry, India.

ABSTRACT Rosai-Dorfman disease (RDD) is a rare, benign, and predominantly nodal disease that most commonly presents as bilateral, painless cervical lymphadenopathy; although inguinal, axillary, mediastinal, and hilar lymphadenopathy has also been reported. Apart from nodal involvement, RDD has extra nodal manifestations involving bone, soft tissue, and liver as well as constitutional symptoms of fever, night sweats, and weight loss, which make it reasonable to rule out the infectious, autoimmune, and malignant conditions as the differential diagnosis.

KEYWORDS : Lymphadenopathy, Emperipolesis, Langerhan's cell histocytosis, Malignant lymphoma, Sarcoidosis.

CASE REPORT-

A 22 year old female known case of hypothyroidism for 2 years not on treatment presented with symptoms of multiple swellings over her neck which are not painful and gradually progressed in size for the past 1 year. She is having other complaints of easy fatiguability, malaise. There is no difficulty in swallowing .There is history of loss of weight and appetite. No history of swellings anywhere in the body.

On physical examination, she has mild anaemia and multiple small swellings in her cervical region as bilateral cervical lymphadenopathy, no thyroid swelling. Rest of the physical findings were unremarkable. The laboratory investigations were as follows:Hb-12.3gm/dl,PCV-38%,TLC-16700 cells/mm3, plateletcount-2,46,000 cell/mm3,creatinine-0.6mg/dl, ESR- 38 mm/1st hour, normal LFT's, deranged TFT. The cervical node histopathological examination showed multiple histiocytes and plasma cells which is suggestive of Rosai-Dorfman disease.

DISCUSSION-

Although Rosai-Dorfman disease rarely presents a diagnostic challenge, uncharacteristic presentations can cause confusion. Originally described by Juan Rosai and Ronald Dorfman in 1969, RDD was observed as a triad of massive cervical lymphadenopathy, expanded lymph node sinuses, and characteristic emperipolesis within histiocytes. With a slight male predominance and a mean onset of 20 years, RDD often presents as prominent bilateral cervical lymphadenopathy accompanied by nonspecific clinical symptoms, such as fever, tenderness, malaise, night sweats, and weight loss.

When RDD is suspected, it is important to exclude other histiocytic disorders, especially Langerhans cell histiocytosis (LCH), a potentially fatal disease. The histiocytes in LCH are characterized by irregular nuclear contours (nuclear grooves and "coffee-bean"–shaped nuclei) that label diffusely for CD1a and contain Birbeck granules on electron microscopy. The associated inflammatory infiltrate is dominated by eosinophils and lymphocytes, with emperipolesis characteristically lacking.

Another differential diagnosis considered was the xanthogranulomatous family of diseases. This collection of cutaneous histiocytic disorders can display a wide variety of morphologic characteristics, including mononuclear cells that may be vacuolated, xanthomatous, spindle-shaped (all characteristically found in juvenile xanthogranulomas), and display oncocytic changes (more commonly seen in adult xanthogranulomas).Although multinucleated giant cells are often found and help classify the disorder (Touton giant cells in juvenile xanthogranuloma and Langhans giant cells in adult xanthogranuloma), they are, by no means, specific. Multinucleated giant cells, as well as foamy histiocytes, can be seen in RDD; therefore, the presence or lack of emperipolesis is probably the single most-important histologic feature that can help distinguish RDD and xanthogranulomatous diseases. Immunohistochemistry can also help because xanthogranulomatous diseases are usually positive for factor XIIIa, CD68, CD163, fascin, and CD14 and negative for CD1a and S100. Notable exceptions are solitary and multicentric reticulohistiocytosis, which are usually negative for factor XIIIa.

RDD has a variable prognosis with most reports indicating indolent and relatively good prognosis as the disease shows gradual resolution of lymphadenopathy in about 50% of the cases. In contrast, some fatal cases have also been reported. Extranodal disease especially of the liver and kidney, immunological abnormalities, and younger age render a poor prognosis. The disease can also be characterized by remission and exacerbation over months to years, hence long-term follow-up is necessary. Treatment if employed consists of corticosteroids that usually show good response, whereas, chemotherapy and radiotherapy are also used in some cases. Surgical resection is only indicated for massive and life or function-threatening disease.

Involvement of thyroid gland in RDD is very rare only six cases have been reported previously. RDD with thyroid involvement is misdiagnosed frequently as malignancy of thyroid gland with lymph node metastasis which may lead to overtreatment complications. In this case there was no involvement of thyroid gland, RDD in hypothyroidism is diagnosis of coexistence here.

CONCLUSION-

Although RDD is a rare and self-limiting disease, it is an important entity as it can imitate other fatal diseases. Consequently, it should be a consideration as an aetiology of mediastinal and hilar lymphadenopathy, particularly in the picture of benign clinical course. And as such it should be differentiated from conditions like malignant lymphoma, occult metastatic disease, and sarcoidosis by relevant examinations and tests and avoiding other unnecessary investigations.

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