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ROSAI DORFMAN DISEASE-A RARE DISEASE, ATYPICALLY PRESENTING AS PALINDROMIC RHEUMATISM

Internal Medicine

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ABSTRACT

Sinus histiocytosis with massive lymphadenopathy (SHML)(Rosai Dorfman disease) is a rare disorder of unknown etiology, usually associated with lymph node enlargement in various superficial or deep sites. These cases can often be misdiagnosed as lymphoma. The disease affects predominantly the younger age group. It is a benign proliferative histiocytic disorder morphologically characterized by 'emperipolesis'. Extra-nodal involvement isnot rare (43%).Sites include skin, nasal cavity, para nasal sinuses, eyelids, orbit, bone and central nervous system (1)

KEYWORDS

Lymph nodes,Benign,histiocytosis.

A 30 yr. old male presented with a firm, nontender palpable lymph node in his right cervical area along with joint pain affecting lower extremity joint/bone pain since 1 month . history of intermittent fever non documented, mild , not associated with chills, rigors , rash or bleeding tendencies.

Vitals were stable. There was bilateral multiple firm , non tender massive lymphadenopathy noted in cervical region at levels II, III ,IV.

- No pallor, cyanosis , clubbing.
- No evidence of any joint deformities.
- Rest systemic examination within normal limits.

Investigations

Hb-10.2
TLC – 12400
ESR – raised (40mm)

Biopsy was taken. HPE examination s/o preserved architecture of lymph nodes with sinus filled with histiocytes with relative lymphoid hyperplasia.

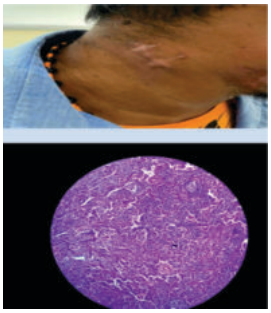
Immunohistochemistry: histiocytes positive for S100,CD-68. Chest Xray- within normal limit.

Rheumatologic work-up was positive for Anti-Nuclear Antibody (ANA), and negative for other autoimmune/inflammatory antibodies including RF, antiCCP.

Diagnosis of palindromic rheumatism was confirmed based on her work-up, and history of recurrent attacks of arthritis/peri-arthritis without joint space loss or erosion.

Patient was started with 1mg/kg of prednisone,responded well to treatment with remission of rheumatic symptoms.

Later gradually tapered off over 3-4 weeks.



Treatment

First line therapy: observation (course of RDD is usually benign, indolent and self limiting). surgery is limited for biopsy and obstructive symptoms.

Patients with progressive disease treated with steroids.

Second line therapy: radiotherapy, cladribine, clofarabine, imatinib, rituximab, azathioprine, methotrexate. Other systemic therapies include combinations such as CVP (cyclophosphamide, vincristine, prednisone) and 6mercaptopurine with prednisone.(5)

Complications

- Complications are mainly due to Extra nodal involvement : most common site being skin. Other includes nasal cavity/paranasal sinuses. Salivary glands,CNS.
- Compressive symptoms due to massive lymphadenopathy

DISCUSSION

- Rosai Dorfman disease is a rare, idiopathic histiocytosis. Characterized by intra sinusoidal proliferation of abnormal histiocytes in sinuses of lymph nodes, lymph vessels of internal organs and in other extra nodal sites.
- The disorder is thought to occur as a result of immune dysregulation or response to a presumed infectious agent. The stimulation of monocytes/macrophages via Macrophage- Colony-Stimulating-Factor leads to immunosuppressive macrophages, which is a main pathogenesis of RDD.
- Clinically mean age of onset is second decade; other features include fever, non- tender bilateral cervical lymphadenopathy, leukocytosis,elevated ESR and polyclonal hypergamma-globulinemia.
- The most common sites for extra nodal disease are the skin, upper respiratory tract and bone, followed by the genitourinary and lower respiratory tracts, oral cavity and soft tissues.
- The most important D/D is RLHSH and LCH; the features favoring SHML are numerous histiocytes with prominent emperipolesis. On IC histiocytes in RLSH and SHML show strong positivity for S-100 and CD- 68; however in LCH the histiocytes also show positivity for CD1a. Further confirmation of LCH can be done by demonstrating Birbeck's granules on electron microscopy.(2)
- An association with immunological diseases, hematological malignancy and post-infection conditions has also been described.

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