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# Sinus Histiocytosis With Hypothyroidism in A Case of Diabetes Mellitus : A Rare Case Report

KEYWORDS: Rosai-Dorfman disease (RDD) or sinus histiocytosis with massive lymphadenopathy (SHML)

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**ABSTRACT** *37 year female patient presented with axillary lymphadenopathy since 3 months, pedal edema and abdominal distension since 2months, known case of diabetes mellitus since 4 years. After investigating and thorough examination a diagnosis of sinus histiocytosis with hypothyroidism in a case of diabetes mellitus was made.*

**Introduction:**

Sinus histiocytosis with massive lymphadenopathy (SHML), which is also known as Rosai-Dorfman disease (RDD), is a rare histiocytic proliferative disorder of unknown etiology initially described by Rosai and Dorfman in 1969. RDD can occur in any age group but is most commonly seen in children and young adults. Usually it presents with massive painless lymph node enlargement at any site, with the cervical lymph nodes being common. Histologically, lymph nodes show pericapsular fibrosis and dilated sinuses, heavily infiltrated with large histiocytes, lymphocytes, and plasma cells.Immunohistochemically, the sinus histiocytes are strongly positive for S-100 protein.[3] BUT OUR CASE PRESENTED WITH AXILLARY LYMPHADENOPATHY.

**Case history:**

35 years old female presented with axillary lymphadenopathy since 3 months,pedal edema and abdominal distension since 2 months, she was a known case of diabetes mellitus since 4 years on treatment. On examination patient had pallor, 2*2 cm right axillary lymphadenopathy, ascitis, pedal edema. On investigating, complete blood counts revealed haemoglobin 8.9 gm%, TLC and platelet counts were within normal limits.Renal function tests were within normal limits. Liver function tests shown bilirubin total 3.5mg/dl ( direct-2.3, indirect 1.2,SGOT 46, SGPT 27,alkaline phosphatase 288), serum albumin 2.8 ,A:G ratio1. On ascitic fluid examination, ascitic fluid was transudative, proteins 0.7gm%,sugar 97mg%,ADA 15.2 U/L,negative for malignant cells, microscopy 2-3 lymphocytes/HPF,TLC 30/cumm , cobweb absent, culture was sterile . USG abdomen & pelvis suggestive of proximal CBD dilated, hepatosplenomegaly, 1.9*1.5cm splenic infarct, gross ascites but with normal liver echotexture. FNAC of axillary lymph node was inconclusive. So we did axillary lymph node biopsy as shown in Figure 1. and on histopathological examnation diagnosis of sinus histiocytosis was made as shown in Figure 2.USG neck suggestive of small colloid nodule in left lobe of thyroid. Thyroid function tests were suggestive of hypothyroidism (TSH 7.12 microIU/ml, T4 3.76 microgm/dl, T3 0.6 ng/ml).After ruling out other causes, we made a diagnosis of sinus histiocytosis with hypothyroidism in a case of diabetes mellitus was made. We did therapeutic ascitic tapping about 2 litres, treated her for deranged LFT,transfused her 20% human albumin, started treatment for hypothyroidism, controlled her blood sugar levels with injection Human mixtard insulin before breakfast and dinner. ENT,surgery evaluation was done. Due to financial constraints CECT abdomen and immunohistochemistry was not done. As her ascitis, pedal edema reduced, blood sugar levels controlled and was feeling better, and she was discharged and asked to return for follow up.

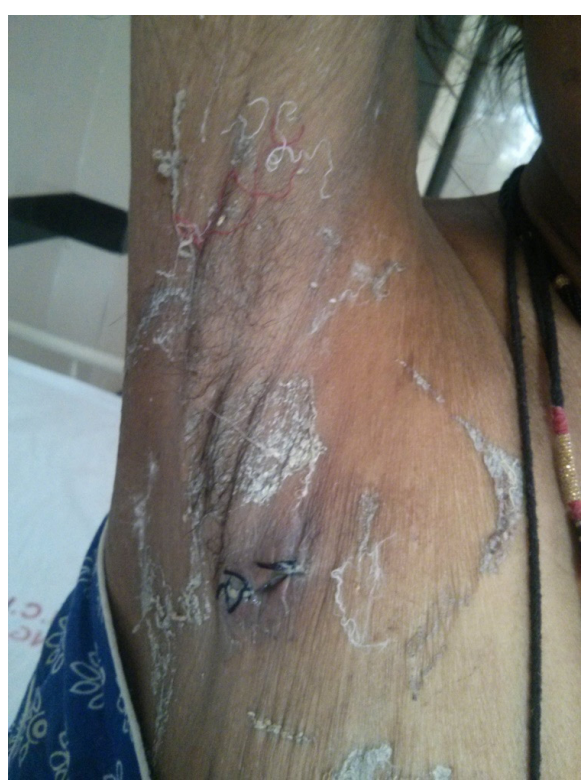

**Figure 1.**

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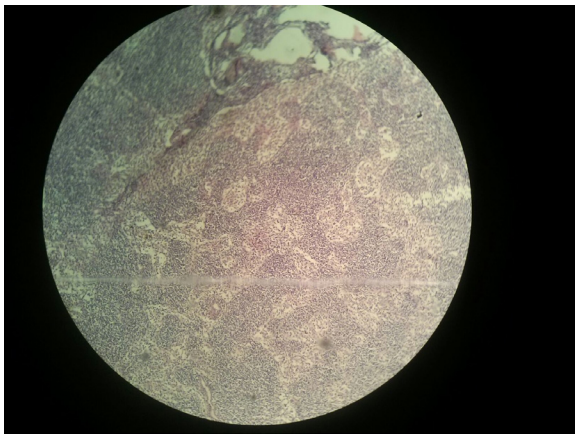

**Figure 2.**

**Conclusion:**
37 year female patient presented with axillary lymphadenopathy since 3 months, pedal edema and abdominal distension since 2 months, known case of diabetes mellitus since 4 years. After investigating and thorough examination a diagnosis of sinus histiocytosis with hypothyroidism in a case of diabetes mellitus was made.

**Discussion:**
Sinus histiocytosis with massive lymphadenopathy (SHML), which is also known as Rosai-Dorfman disease (RDD), is a rare histiocytic proliferative disorder of unknown etiology[3] .The typical age group affected is from second to third decade of life.[1] Usually it presents with massive painless lymph node enlargement at any site, with the cervical nodes being common[3]but in our case axillary lymphadenopathy was involved. But extranodal involvement has also been documented in around 40% of cases, the most common being skin followed by upper respiratory tract and bone. However, RDD also can occur in a variety of other sites, including thyroid as in our case, the genitourinary system, lower respiratory tract, oral cavity, and soft tissues. Other reported specific sites include brain, spine, liver, kidney, breast, parotid gland, orbit, nasal cavity, and lungs.[3]

Etiology of the disease is still unclear and controversial. Some studies have demonstrated that pathogenesis of RDD is related to viral infections, such as human herpes virus (HHV), parvovirus B19, and Epstein–Barr virus (EBV). However, other studies have not confirmed these results. RDD has been reported in patients with immunoglobulin IgG4-related disease, but there is no clear evidence that these disorders have a common etiopathogenesis.[4]

Low grade fever is generally present along with, normocytic normochromic anaemia, elevated ESR, leucocytosis, and hyperglobulinaemia, which are non-specific clinical findings.[2]-Ultrastructurally, histiocytes in RDD lack cytoplasmic Birbeck granules unlike in Langerhans cell histiocytosis. On immunostaining RDD show positivity for S–100 protein, CD11c, CD14, CD33, and CD68 antigens and are CD1a negative.[2]

The characteristic pathologic feature of this disease is proliferation of distinctive histiocytic cells that demonstrate emperipolesis (the engulfment of lymphocytes and erythrocytes by histiocytes) in the background of a mixed inflammatory infiltrate, consisting of moderately abundant plasma cells and lymphocytes.[3] Immunohistochemically, SHML cells express phagocytic markers such as CD68 and S100, but not the markers for Langerhans (CD1a) or dendritic cells (DRC, CD23, and CNA42).[3]

Treatment for the RDD is recommended only in patients who are symptomatic or have vital organ or systemic involvement. Sometimes, in 20% of cases the disease is self-limited and shows spontaneous regression. Complete surgical resection is the best option for treatment of the localized RDD. In symptomatic cases of extensive RDD steroids are the first-line therapeutic option. Radiotherapy can be used in cases with orbital, airway, and central nervous system involvement but till now no guidelines have been established for that management. In cases of disseminated RDD or those refractory to surgery or other modalities, chemotherapy has been used with varying degrees of success [4].

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