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Generalized Lymphadenopathy: A Case Report of Rosai-Dorfman Disease

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ABSTRACT

Sinus histiocytosis with massive lymphadenopathy (SHML), Rosai-Dorfman Disease, is a rare histiocytic syndrome first described by Rosai and Dorfman, seen predominantly in childhood and early adulthood. Even though it is considered a benign disease, fatalities may occur due to cellular infiltrates of SHML.

We report a 16–year–old boy with signs of polydypsia, polyuria, weight loss and generalized lymphadenopathy. He had been receiving corticosteroid following the diagnosis of histiocytosis X.

Due to hyperglycemia, the patient was admitted with the primary diagnosis of diabetic ketoacidosis and medications were initiated. All paraclinical and immunologic examinations were negative. Axillary lymph node biopsy revealed the diagnosis of Rosai–Dorfman disease. ***(Tanaffos 2007; 6(3): 65-67)***

Key words*: Rosai–Dorfman disease, Diabetes mellitus, histiocytosis X, Children.*

INTRODUCTION

Rosai-Dorfman disease (RDD), or sinus histiocytosis with massive lymphadenopathy (SHML), is an unusual clinical entity (1). SHML was first delineated as an entity by Rosai and Dorfman in 1969 and is characterized by benign pseudolymphomatous proliferation with significant histiocytic infiltration. Since its original description, Rosai-Dorfman disease has become a well-established clinico-pathological entity whose registry now numbers more than 750 cases (2). The mean age of presentation is 20.6 years, and males (58%) outnumber females (42%). A few cases have affected two members of the same family. Although the disease has a widespread geographic distribution and most of the reported cases are from the US and western Europe, there is a disproportionally high number of cases from Africa and the Caribbean region (3, 4).

Sinus histiocytosis with massive lymphadenopathy is mainly an idiopathic histioproliferative disease in children affecting the lymph nodes. Extranodal involvement has also been recognized, but central nervous system

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manifestations are extremely rare (5).

Most often the RDD takes a benign course and treatment is not necessary (5,6).

CASE REPORT

A 16-year–old boy was referred to our hospital for polydypsia, polyuria and weight loss initiated 5 months before admission and had gradually progressed. The patient had been suffering from bilateral sensory–neural hearing loss since his birth, which was diagnosed when he was 7 months old. Seven years ago, he was admitted to another hospital because of generalized adenopathy and persistent respiratory infections and was diagnosed with histiocytosis X via lymph node biopsy. He was on steroid therapy for seven years. The weight and height percentile of the patient were 5% and 25%. In his family history the only positive finding was deafness of his older brother.

On physical examination, he had generalized lymphadenopathy. Bilateral, posterior and anterior cervical lymphadenopathy as well as submandibular lymphadenopathy were more significant. The existing lymphadenopathies were painless and non-tender. The examination of cranial nerves revealed impaired sensory-neural hearing function. All cerebellar and coordination tests were normal as well as his mental status. The other organs were normal.

Laboratory test showed the blood sugar level to be 750 mg/dl, ESR level reported to be 40 cm/hr and peripheral blood cells count showed mild leukocytosis. Keton bodies were detected in the urinalysis.

Tuberculin skin test induration diameter was 13mm. Smear and culture of the sputum were negative for tuberculosis. NBT test was 100%, Flowcytometery and immunoglobulin serum levels were normal.

Chest x-ray was normal. Chest, abdominal and brain CT-scans were reported to be normal. For definite diagnosis, lymph node biopsy was performed and sections showed pronounced dilation of lymph node sinuses, resulting in partial architectural effacement. The sinuses were occupied by lymphocytes, plasma cells with notably numerous nuclei and abundant clear cytoplasm. Many of these histiocytes have numerous intact lymphocytes in their cytoplasm (Emperipolesi), these histiocytes were strongly positive for S100 (Figure 1).

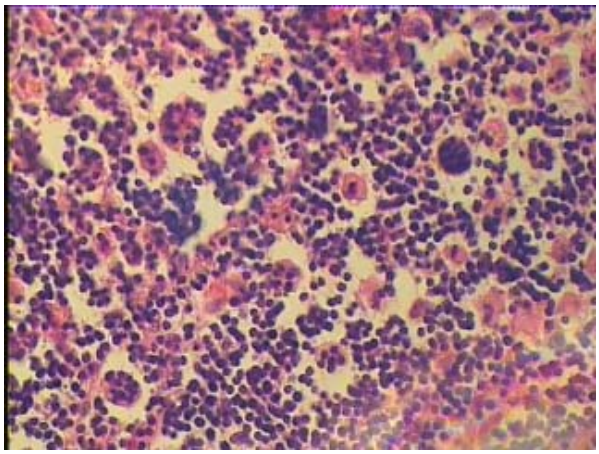

Figure 1. Histiocytes with intact lymphocyte within the cytoplasm.

Considering the clinical and pathological findings, the patient was diagnosed as having sinus histiocytosis with massive lymphadenopathy (SHML).

Due to the high blood sugar level, the diagnosis of diabetic ketoacidosis was made and Insulin therapy was started. Within two weeks, the blood sugar level reached normal. Oral corticosteroid was tapered and the patient was discharged after 2 weeks.

DISCUSSION

In 1969, Rosai and Dorfman described a newly recognized benign histioproliferative disease characterized clinically by massive cervical lymphadenopathy, fever, and leukocytosis and characterized pathologically by enlarged lymph node sinuses containing large histiocytes with

phagocytosed lymphocytes. In our patient the most significant clinical finding was lymphadenopathy especially in cervical nodes (1, 3).

Common presentation of RDD is bilateral painless cervical lymphadenopathy. This phenomenon has also been described as “bull neck”. Cervical nodes are most commonly involved. Involvement of axillary, mediastinal, and inguinal nodes is also found in association with RDD (5, 7).

Due to the diagnosis of histiocytosis X in our patient, he has been taking corticosteroids for 7 years which has led to diabetes mellitus.

To distinguish disease processes and make a definite diagnosis, the histologic features of the lymph node help lead to diagnosis. The histiocytes that should be differentiated include Langerhans cell, interdigitating reticulum cell, and the common phagocytic macrophages or histiocytes. It has been noted that the disease is a reactive rather than a neoplastic disesase. These characteristics are in contrast to Langerhans cell histiocytosis (histiocytosis X) in which Langerhans cell proliferation predominates. The two molecules detected in both SHML and histiocytosis X are the cell adhesion molecule CD31 and the protein S100. Despite the presence of CD31 and S100 in both SHML and histiocytosis X, differentiation of the diseases can be done with careful attention to morphologic features (8, 9, 10).

The final diagnosis of SHML is made based on clinical and histological findings and histiocytes positive for S100 and CD68.

In children with massive lymphadenopathy, SHML should be considered in the differential diagnosis.

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