CASE REPORT

# Rosai-Dorfman Disease with Pleural Involvement

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## Abstract

Sinus histocytosis with massive lymphadenopathy (SHLM) disease is considered to be indolent with self limiting pathology. However, severe morbidity and mortality have been attributed to complications of SHLM. Lower respiratory tract involvement, which is often unfavorable, is rarely reported and carries particularly grave prognosis. A case of sinus histocytosis with massive lymphadenopathy (SHLM) is reportedhere. The patient had lower respiratory and pleural involvement.

**Keywords:** Rosai Dorfman; Sinus histocystosis; Respiratory tract

## Introduction

Sinus histiocytosis with massive lymphadenopathy (SHLM) was identified and established as a distinct clinicopathologic entity following the reports of Rosai and Dorfman.[1] It was initially characterized as a nodal-based disease process. Since then, it has been well established that SHLM may affect a variety of extranodal sites. A review of the cases from the registry of SHLM reported that 43% of patients with SHLM had at least one site of extranodal involvement. Extranodal SHLM can occur independently of lymph node involvement. Lower respiratory tract involvement has rarely been reported in the literature. We report a case of SHLM with lower respiratory tract and pleural involvement.

## Case report

A 58 year-old Caucasian man presented in 1995 with a 4-month-history of bilateral cervical adenopathy and a 2-month-history of progressive dyspnea, cough and left chest pain. Physical examination on admission revealed massive lymphadenopathy in the anterior and posterior cervical chains bilaterally and the left inguinal region as well as xanthomatous-like skin lesions. The examination of the lungs was normal. Laboratory investigations showed an erythrocyte sedimentation rate at 105 mm/hour and normal white blood cell count. Chest x-ray showed an alveolar infiltration of the lower right lung field as well as pleural effusion. Tuberculin skin reaction was negative. The sputum showed no acid-fast bacilli on Ziehel-Neelson stain and cultures for any *Mycobacterim spp.* were negative. Human immunodeficiency virus and toxoplasmosis serologies were also negative. A cervical lymph node biopsy was performed showing capsular fibrosis and dilatation of subcapsular and medullary sinusis, filled with active but benign-appearing histocytes containing cellular debris, erythrocytes and numerous lymphocytes expressing CD68 and S100 protein. A bronchial fibroscopy was performed showing numerous granular-appearing polypoid tissues on the wall of the main bronchus. Neither of these tumefactions compromised the airway. Bronchial, transbronchial and skin biopsies showed histological appearances similar to the node biopsy, being compatible with SHLM (Figure 1).

Thoracoscopy followed by pleural biopsy of nodular and inflammatory spaces showed exactly the same histological aspect of the cervical lymph node (Figure 2). Corticosteroids were administrated daily (0.75 mg/kg of prednisone equivalent). At the follow up after presentation, the patient reported a progressive worsening of his symptoms and died four months later with severe obstruction. Postmortem examination was not performed.

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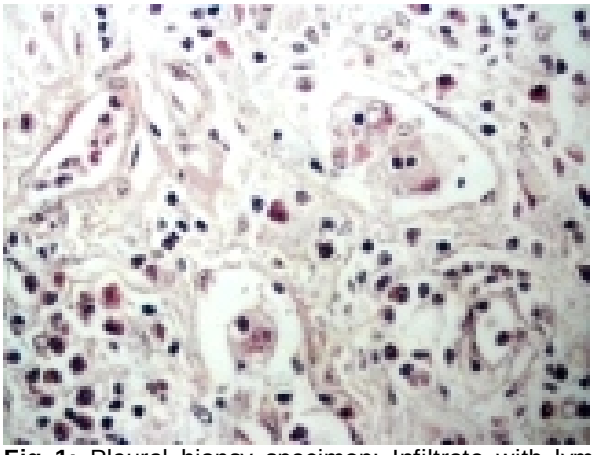

**Fig 1:** Pleural biopsy specimen: Infiltrate with lymphoid cell clusters altering with pale areas representing the histiocytic cell population (H&E, x 40).

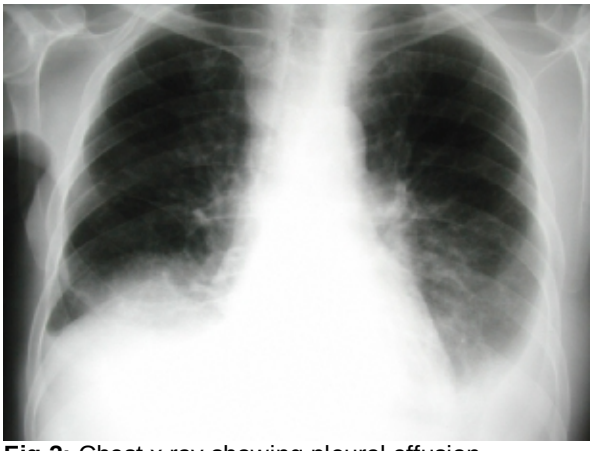

**Fig 2:** Chest x ray showing pleural effusion.

## Discussion

Since the original descriptions of sinus histiocytosis with massive lymphadenopathy, the extent of the extranodal manifestations of this condition has been well recognized. The etiology remains unknown, but it is thought to be a disorder of immune regulation or response to a presumed infection agent.[1]

Irrespective of the site of occurrence, the histopathologic appearance of SHLM is essentially similar. The hallmark is the tendency for the infiltration to recapitulate the lymph node architecture. This has been referred to as a "sinusal" pattern in which the clustering of lymphocytes stimulates the appearance of germinal centers with surrounding cellular proliferation and dilated sinuses. More fibrosis, fewer typical histocytes and a lesser degree of lymphophagocytosis are the minor histological differences between nodal and extranodal SHLM.[2]

Rosai-Dorfman disease involving the thorax is rare and most often manifests as pulmonary disease, accounting for only 2% of Rosai-Dorfman disease cases.[3] The patients who develop pulmonary Rosai-Dorfman disease are young (average age, 14 years) and their lymph nodes and other extranodal sites are involved (nasal cavity and paranasal sinuses). The tracheobronchial tree is infiltrated most commonly, and intraluminal polypoid growth of Rosai-Dorfman disease may produce airway obstruction. Diffuse interstitial or air space involvement occurs less frequently. However, when there is extensive parenchymal involvement, the pleura may be rarely involved.[3] Our case is specific in that the patient is not young and the extensive involvement of the pleural lymphatics by the characteristic histiocytes appeared to be responsible for the pleural effusion without extensive parenchymal involvement.

The natural history of SHLM is variable and typically reported to follow a benign course with spontaneous resolution within a period of several months to several years. It must be stressed that lower respiratory tract involvement in SHLM is unfavorable and carries particularly grave prognosis as in our case.[4] Treatment does not appear to be necessary in the majority of patients with SHLM since the disease is often self-limited and subject to spontaneous regression. The major indication for surgery other than biopsy is life or function threatening obstruction. Patients with progressive symptoms, as in our case, have been treated with chemotherapy such as alkylating agent and corticosteroids. However, these treatments did not lead to effective outcomes.[5]

**Conflict of interest:** None declared.

## References


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