

## CASE REPORT

## Rosai-Dorfman Disease

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## ABSTRACT

Rosai-Dorfman disease also known as sinus histiocytosis with massive lymphadenopathy is a rare benign disorder of unknown etiology, which present most commonly as bilateral massive lymphadenopathy of neck associated with fever. These cases are frequently misdiagnosed as lymphoma, and thus it is important to distinguish Rosai-Dorfman disease from other causes of neck swelling because of different treatment modalities.

We report here a case of Rosai-Dorfman disease presenting with massive right cervical lymphadenopathy.

**Keywords:** Massive lymphadenopathy, Rosai-Dorfman disease, Sinus histiocytosis.

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## INTRODUCTION

This fascinating disorder, first described by Rosai and Dorfman in 1969 also known as sinus histiocytosis with massive lymphadenopathy (SHML), is benign, self-limiting disorder that commonly involves the lymph nodes.<sup>1</sup> Although cervical region is by far the most common and most prominent site of involvement but peripheral or central lymph nodes are also affected. In about 25 to 40% of cases, extranodal sites are also affected.<sup>2-4</sup>

The cytological features of SHML are virtually diagnostic and can obviate the need for biopsy in most cases. The S 100 stain is helpful in identifying the histiocytosis in SHML, and there is no specific treatment present for this disease.

## CASE REPORT

A 10-year-old male patient presented with history of swelling in the right side of neck of 25 days duration associated with low-grade fever. On clinical examination, swelling was 10 × 7.5 cm in right anterior triangle of neck (Fig. 1). The swelling was nontender, soft to firm in consistency and mobile in all direction.

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The peripheral blood cell count showed leukocytosis (17,000/mm<sup>3</sup>) with neutrophilia. Erythrocyte sedimentation rate (ESR) was raised to 45 mm after 1st hour and serum gammaglobulins were raised. The fine needle aspiration cytology (FNAC) was taken from the right cervical node, and smears were stained by May-Grünwald-Giemsa stain and smear showed sheets of histiocytes most of them showing intact lymphocytes in their cytoplasm, a feature known as emperipolesis or lymphophagocytosis (Fig. 2). On ultrasonography (USG) abdomen, did not reveal any organomegaly. On immunocytochemistry, the histiocytes were positive for S 100 and CD 68 thus, confirming the diagnosis of Rosai-Dorfman disease.

Based on these characteristic cytomorphology and supported by clinical and laboratory investigations, a diagnosis of Rosai-Dorfman disease was made.

Although this disorder is self-limiting in most cases and undergoes complete spontaneous resolution but it follows a protracted clinical course for years and decades



Fig. 1: Neck swelling

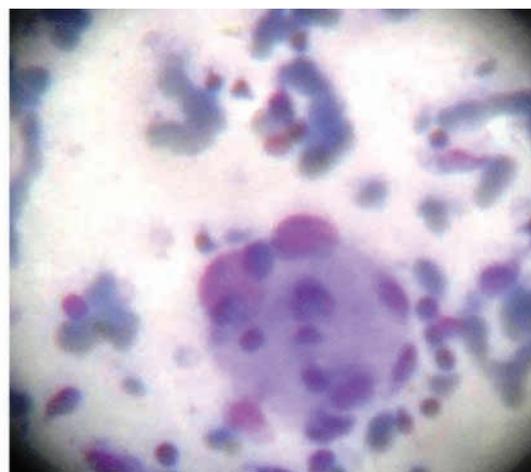


Fig. 2: Smear showing sinus histiocytes with lymphocytphagocytosis (emperipolesis) (MGG, ×400)

in cases with widespread extranodal involvement. This patient was put on oral prednisolone 10 mg three times a day in tapering doses for 21 days and patient showed marked clinical improvement.

## DISCUSSION

Sinus histiocytosis with massive lymphadenopathy or Rosai-Dorfman disease is a rare but well defined histiocytic, proliferative disorder of unknown etiology. The disease is thought to be a disorder of immune regulation or response to a presumed infectious agents (HHV-6/EBV) with its major manifestation in lymph nodes with resultant proliferation of sinusoidal histiocytes. The stimulation of monocytes/macrophages via macrophage-colony-stimulating-factor leads to immunosuppressive macrophages, which is considered as main pathogenesis of Rosai-Dorfman disease (RDD).<sup>5</sup> It may occur at any age but mostly seen in young adults and children with a slight predilection for males (58%) and for individuals of African descent.<sup>6,7</sup> Clinically, patients present as massive, painless, cervical lymphadenopathy associated with low-grade fever, leukocytosis, elevated ESR and hypergammaglobulinemia. Formerly, it was thought to be a process limited to lymph nodes, but in 40% of cases, it also involves extranodal sites, such as eyes and ocular adenexa, head and neck, upper respiratory tract, skin, subcutaneous tissue, bone, skeletal muscles, central nervous system (CNS), gastrointestinal tract (GIT), salivary glands, genitourinary tract, thyroid, breast, kidney, liver, heart and uterine cervix.<sup>2-4</sup> The onset of SHML is typically insidiously; the active phase is prolonged, there may be spontaneous remissions and relapses. Deaths have been reported in very few cases.<sup>8,10</sup>

The histocytological features include a histiocytic proliferation with presence of large histiocytes showing fine vacuoles in the cytoplasm and large vesicular nuclei. The phagocytosis of lymphocytes and plasma cells (emperipolesis) is characteristic feature.<sup>9,10</sup> This disease also exhibits some of the phenotypic features of dendritic cells, such as S 100, cathepsin E, fascin and, at times CD 1a, helping in confirmation of diagnosis.<sup>11</sup>

Radiological and other imaging modalities are useful to investigate extranodal involvement. Sinus histiocytosis with massive lymphadenopathy is generally regarded as benign self-limiting disorder in spite of its propensity to form large masses and to disseminate to both nodal and extranodal sites.

The common differential diagnosis includes infectious lesions, reactive lymphoid hyperplasia with sinus histiocytes (RLHSH), Langerhans cell histiocytosis (LCH), hemophagocytic syndrome and malignant lymphoma.

Although this disorder is relatively resistant to therapy, treatment options can be divided into surgical and radiochemotherapy. Surgery is mainly indicated for life threatening conditions. In chemotherapy, the most effective regimens include a vinca alkaloid combined with an alkylating agent and corticosteroid. Radiation therapy is also indicated for threatened function, such as cord compression and respiratory embarrassment.<sup>8,12,13</sup>

## CONCLUSION

Rosai-Dorfman disease is characterized clinically by massive lymphadenopathy affecting mainly the cervical region. Being a rare entity, clinical suspicion is difficult and it is mostly diagnosed by characteristic cytological features.

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