

CASE REPORT

Sinonasal Manifestation of Rosai-Dorfman Syndrome

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ABSTRACT

Sinus histiocytosis with massive lymphadenopathy (SHML) (also known as Rosai-Dorfman syndrome) is a rare, idiopathic, benign and self-limiting histiocytic proliferative disorder. It most commonly involves the cervical lymph nodes. The disease has a benign course and involvement of the nasal cavity as an extranodal site is exceptional. Here, we report a case of bilateral sinonasal mass with subsequent involvement of cervical lymph nodes in 22-year-old lady. Histological examination of the cervical lymph nodes and nasal mass biopsy demonstrated evidences of Rosai-Dorfman disease (RDD). The clinical presentation, histologic characteristics, radiographic findings and treatment of the disease are discussed.

Keywords: Cervical lymphadenopathy, Rosai-Dorfman syndrome, Sinonasal mass, Sinus histiocytosis.

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INTRODUCTION

Sinus histiocytosis with massive lymphadenopathy (SHML) is a unique disease of unknown etiology, first described by Ajoury and Reed, and later on by Rosai and Dorfman, in 1969¹ is now widely known as Rosai-Dorfman disease (RDD) or syndrome (RDS). The most common clinical finding is massive painless, bilateral cervical lymphadenopathy. Extranodal involvement is present in about 43% of cases, preferably in head and neck region involving skin, nasal cavity, paranasal sinuses, orbit, ear and central nervous system. The diagnosis of Rosai-Dorfman disease is made on the basis of clinical suspicion and confirmed by histopathological examination and immunohistochemical studies. We present a rare case of Rosai-Dorfman syndrome (RDS) involving

sinonasal region with cervical lymphadenopathy in a 22-year-old female and provide clinical, imaging, pathologic findings and differential diagnosis.

CASE REPORT

A 22-year-old female presented to ear, nose and throat outpatient department (ENT OPD) with 18 months history of progressive bilateral nasal obstruction, insidious in onset, occasional episodes of epistaxis, and yellowish mucoid nasal discharge. There was no history of fever, allergy, headache, visual discomfort and weight loss. No self or family history of diabetes, hypertension, asthma, drug allergy, substance abuse or anti-tubercular treatment was present.

Ant rhinoscopy Figure 1 revealed bilateral firm pinkish polypoidal mass with prominent vessels, which bleed severely on manipulation. Posterior rhinoscopy showed fleshy lobulated mass in the both choana. Examination of the ears showed no evidence of secretory otitis media and her hearing was within normal limits. On neck examination, she found to have grossly enlarged painless bilateral, multiple, non-matted, mobile, firm cervical lymphadenopathy (3 × 5 cm in size). No axial or inguinal lymphadenopathy. Rest of the ENT examination and systemic examination revealed no abnormality.

On clinical examination, provisional diagnosis—sinonasal mass with cervical lymphadenopathy was made and patient underwent routine investigations and radiological imaging for confirmation of diagnosis.

Investigations showed a HB-8.7 gm/dl, total white blood cell (WBC) count-8700/mm³, differential WBC count-N 86, L12, E01, M01 and erythrocyte sedimentation rate (ESR) 85 mm 1st/hour. Peripheral blood film showed microcytic, hypochromic red blood cells showing anisocytosis. Chest X-ray and ultrasonography (USG) scan of the whole abdomen was normal. Computed tomography (CT) scan of nose and paranasal sinuses (PNS) showed soft-tissue opacification of the both nasal cavities, bilateral maxillary ethmoidal and frontal sinuses with widened osteomeatal complex (Fig. 2). No bony erosion was seen.

Immunohistochemistry-histocytic positivity for S-100 proteins and CD 68 were present.

Fine-needle aspiration cytology (FNAC) of right and left cervical node showed sheets of sinus histiocytes

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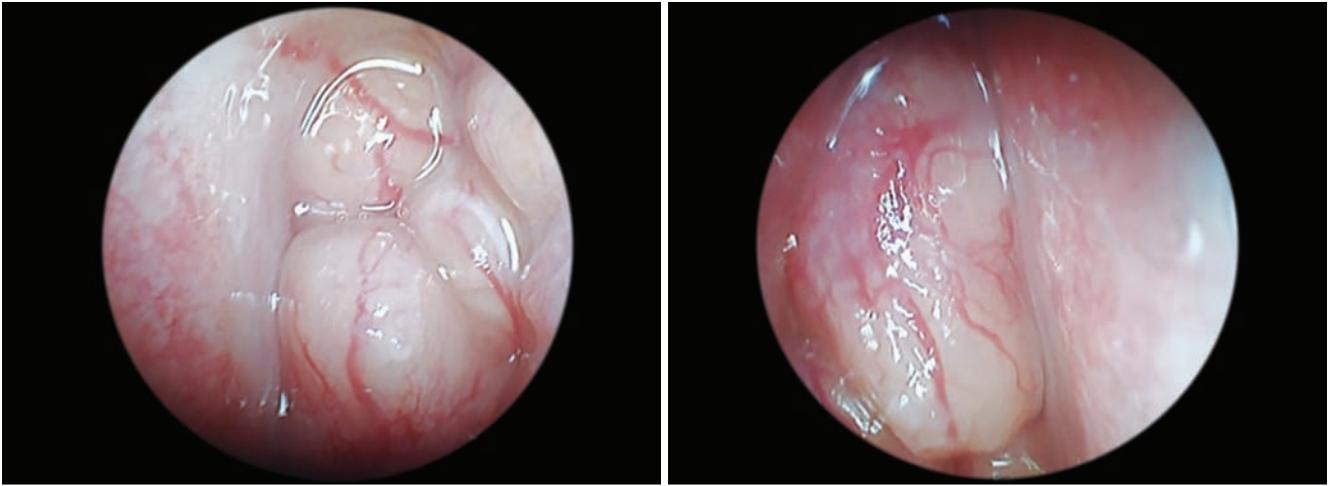


Fig. 1: Bilateral pinkish mass

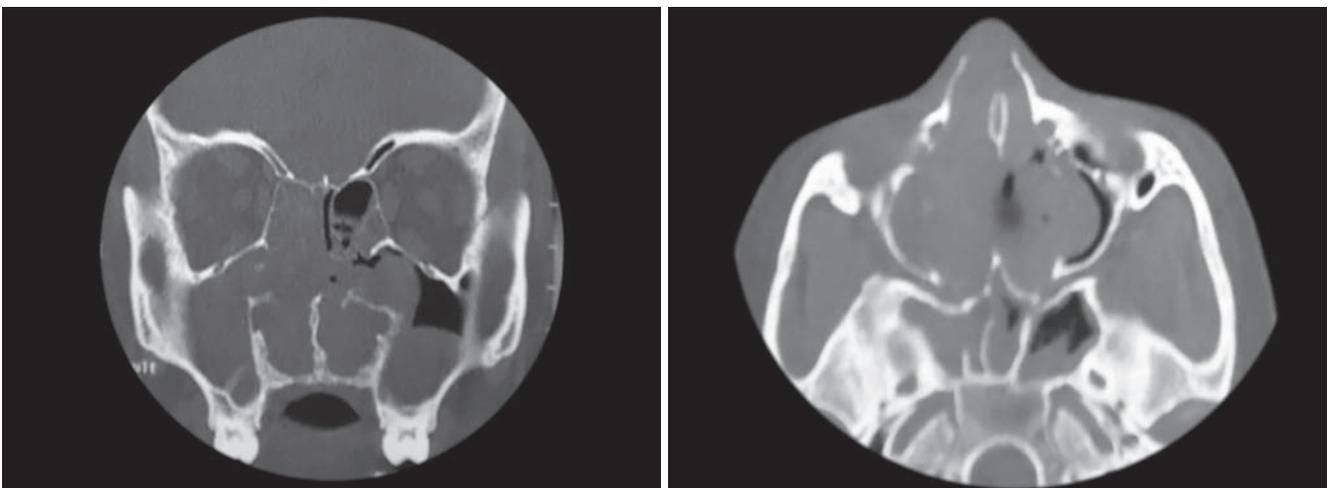


Fig. 2: Soft-tissue opacification on CT

with lymphocytphagocytosis and erythrophagocytosis (emperipolesis). Biopsy tissue from lymph node and nasal mass showed features consistent with RDD (Fig. 3).

She was started on low dose oral steroids. Five mg/kg/day, started showing regression of neck swellings and nasal obstruction after 3 months.

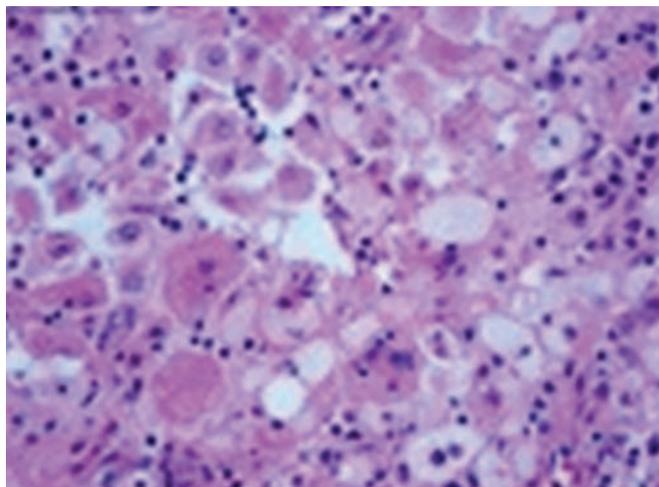


Fig. 3: Sinus histiocytosis with lymphocytphagocytosis or emperipolesis

DISCUSSION

Sinus histiocytosis with massive lymphadenopathy (also known as Rosai-Dorfman syndrome) is a rare, benign and self-limiting histiocytic proliferative disorder. The exact etiology of the disease is unclear, but it is presumed to be due to an immune regulation disorder, as well as infections caused by agents like herpes viruses, epstein-barr virus, cytomegalovirus, brucella and klebsiella. However, no laboratory evidence pointing to an etiologic agent and no other clinicopathological signs suggesting a deficient humoral or cellular immunity or phagocytosis disorder have been reported. It presents during, first or second decades of life and have relatively male sex predominance (male:female ratio = 2:1) and rarely seen in Asian population.

Bilateral painless massive cervical lymphadenopathy is seen in most of the cases (87.3%), axillary (23.7%), inguinal (25.7%), and mediastinal (14.5%) regions can also be affected. Initially nodes are isolated, mobile and small, but becomes adherent with disease progression, forming a voluminous multinodular mass involvement.

Additional features include: fever, weight loss, anemia, leukocytosis, neutrophilia, increased ESR and hypergammaglobulinemia are other common associations.⁵

Extranodal manifestation of the disease is observed in 28 to 43% of cases, with preference for head and neck region.² Skin is the most commonly affected extranodal site, other sites are nasal cavity, paranasal sinuses, orbit, ear and central nervous system.

In our case, bilateral cervical group of lymph nodes, nasal cavity and paranasal sinuses were involved, which leads to differential diagnosis of lymphoma, tuberculosis, sarcoidosis, rhinoscleroma, syphilis and sinonasal carcinoma.

The diagnosis of RDD is made on the basis of clinical suspicion and confirmed by histopathological examination along with immunohistochemical studies.

The characteristic histopathological features of the SHML is distinctive histiocyte/phagocytic cells against a background of a mixed inflammatory infiltrate, consisting of abundant plasma cells (Russel bodies) and lymphocytes within dilated lymph node sinuses and lymphatics in extranodal sites. The cytoplasm of many of the histiocytes contains well preserved lymphocytes and plasma cell, a phenomenon referred to as lymphocytophagocytosis or emperipolesis. On Biopsy the classical finding of emperipolesis differentiates it from other diseases.

The histiocytes of SHML are markedly immunoreactive to the S-100 protein and constantly express monocyte-macrophage-associated antigens CD 11c, CD 14 and CD 68.³ These cell markers can be used to differentiate SHML from various disorders including lymphoma and langerhan cell histiocytes (LCH).

Differential diagnosis (D/D) of sinonasal polyposis with enlarged cervical nodes of separate etiology.

- Infective disorders like rhinoscleroma, lymphoma (Non-Hodgkins) extramedullary plasmacytoma, sinonasal/nasopharyngeal malignancy, sinus histiocytosis with massive lymphadenopathy, post-transplant lymphoproliferative disorder, granulomatous disease of nose-TB, syphilis, sarcoidosis, wegeners granulomatosis.

Its course is usually characterized by insidious onset, protracted duration of active disease state and eventual spontaneous remission, but occasional recurrences are also reported.

The outcome is usually good, and the disease is often self-limited. In approximately 50% of patients, the disease resolves without any sequelae, one-third have residual asymptomatic adenopathy and 17% have persistent sympyomatology after 5 to 10 years.⁴ Treatment is warranted only in few cases, where massive nodal or extranodal enlargement interferes with organ function or threatens life.

The most common treatments for RDD include corticosteroid therapy, surgery, radiation therapy, chemotherapy (vinca alkaloid in conjunction with an alkylating agent) and newer agents, such as interferons.⁵ The reported mortality rate is about 7%; many patients, who have a fatal outcome, suffer from concomitant immune dysfunction (circulating autoantibodies to arthritis). We had treated our patient with corticosteroid therapy for, which the response is favourable.

CONCLUSION

Here, in our case bilateral nasal polyposis with painless cervical lymphadenopathy, of 18 months duration confirmed as RDS by histopathology and immune chemistry in 22-year-old female. Rosai-Dorfman syndrome is a rare cause of lymphadenopathy but a well-defined clinicopathologic entity. Massive cervical lymphadenopathy is the hallmark of the disease and head-neck region is the preferred site of the extranodal involvement. Indolent course, presence of histiocytes with emperipolesis in inflammatory exudates and S-100 positive immunohistochemical staining clinch the diagnosis in favor of RDD or sinus histiocytosis with massive lymphadenopathy.

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