INTRODUCTION:
Rosai-Dorfman disease (RDD) is a rare histiocytic disorder initially described as a separate entity in 1969 by Rosai and Dorfman under the term sinus histiocytosis with massive lymphadenopathy (SHML) (18). The pathogenesis of RDD are not fully understood and treatment strategies can be different according to severity or vital organ involvement.

CASE HISTORY:
19 year old male patient presented with history of swelling in bilateral submandibular region for last 6 months associated with pain, anorexia and weakness. CBC and ESR were normal. CXR was normal. We diagnosed case by FNAC and confirmatory diagnosis could be put forwarded by histopathological examination and immunohistochemistry.

DISCUSSION:
Although RDD may occur in any age group, it is most frequently seen in children and young adults (11). Patients presenting with isolated intracranial disease tend to be older (5). The disease is more common in males and in individuals of African descent (14). RDD has been reported following bone marrow transplant with multiple relapses and remissions for years. In these patients, non-tender, movable, firm, discrete and free from superficial and deep structures were present. FNAC was done which showed numerous large histiocytes with abundant pale cytoplasm and phagocytosed lymphocytes (Emperipolesis) (FIGURE-1) in background of lymphocytes and plasma cells, multinucleated histiocytic giant cells and eosinophils. Biopsy was done and sent for histopathological examination. Grossly specimen was greyish white firm tissue measuring 3.2 x 1. cm. On cut section it was homogenous white in appearance. Histopathology showed dilatation of lymph sinuses with effacement of follicles. There was proliferation of sinus histiocytes, lymphocytes and plasma cells within cytoplasm (Emperipolesis) (FIGURE-2). Biopsy was sent for immunohistochemistry and it was positive for S-100 (FIGURE-3). Patient responded well to steroid treatment.

Rosai-Dorfman Disease

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ABSTRACT
19 year old male patient presented with complaint of bilateral swelling in submandibular region for last 6 months associated with pain, anorexia and weakness. CBC and ESR were normal. CXR was normal. We diagnosed case by FNAC and confirmatory diagnosis could be put forwarded by histopathological examination and immunohistochemistry.

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70% to 80% of patients have spontaneous improvement of symptoms without treatment, although they may have alternating episodes of worsening and relieving of symptoms for a long period of time. Some patients with severe or persistent disease or cases when organ function is threatened (such as breathing obstruction or kidney failure) may require treatment with surgery, steroids, and/or chemotherapy. Rarely radiation therapy may be used. Chemotherapy may include vinblastine, 6-MP, methotrexate, thalidomide, or Gleevec. The ultimate goal of an overall treatment plan, of course, is to use as little treatment as possible to keep the disease under control and preserve quality of life. Rosai-Dorfman does not usually threaten life or organ function. It is believed that 5% to 10% of patients have progressive disease that may damage tissue. However, for most patients, the disease is self-limited, and the outcome is good.

FIGURE-3 S 100 POSITIVITY

FIGURE-1 FNAC

FIGURE-2 HISTOPATHOLOGY

REFERENCE