Case Report

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Rosai Dorfman disease: a case report

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ABSTRACT

Rosai Dorfman disease (RDD) is a rare granulomatous disease that typically presents with massive lymphadenopathy. To our knowledge, there have only been 650 cases reported worldwide. It is more common among adults and has characteristic clinical, radiological and histopathological findings. Here, we present a case of RDD in a 51 year-old man with massive submental lymphadenopathy which was not responding to antibiotics. Computerized Tomographic Scan showed ill-defined non-enhancing soft tissue density while fine needle aspiration for cytology displayed emperipolesis which is seen in patients with RDD. The swelling followed a benign course and reduced in size progressively without further intervention.

Keywords: Rosai Dorfman disease, Sinus histiocytosis, Massive lymphadenopathy, Emperipolesis

INTRODUCTION

RDD, also known as sinus histiocytosis, is a rare benign disease that typically presents with massive lymphadenopathy. Histologically, it is characterized by proliferation of histiocytes within the sinus of lymph nodes and the presence of emperipolesis which are hematological cells within the cytoplasm of histiocytes. In the majority of cases, the disease is self-limiting requiring no specific treatment. Surgical excision and other medical treatments are often reserved for cases with vital organ involvement. Here we report a rare case of Rosai Dorfman disease (RDD) involving the submental region.

CASE REPORT

An otherwise healthy 51 year old man presented with an 11-day history of submental swelling which was

associated with fever, cough and sore-throat. On examination, the swelling measured 5×4 cm, was firm with erythema of overlying skin. He had a history of dental extraction of the left lower molar 2 days prior to the onset of swelling and was admitted to the district hospital. Blood investigation showed a slight raise in the total white cell count $(13x10^3/\mu l)$. He was also screened for tuberculosis which was negative. The patient was given a week of antibiotics but despite that, the size of the swelling remained static and there was slight improvement in the total white cell count (from $13x10^3/\mu l$) to $10.7x10^3/\mu l$).

He was then referred to our tertiary center for further management. An ultrasound scan done revealed subcutaneous edema and thickening of the submental region with no definite collection of pus seen. Multiple subcentimeter lymph nodes were seen at the submandibular region. Other structures were normal. The impression was that of submental subcutaneous swelling

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with no definite abscess collection suggestive of cellulitis. The patient was planned for fine needle aspiration for cytology (FNAC) of submental lymph node.

The FNAC results (Figure 2) showed abundant neutrophils with polymorphous population of lymphocytes, erythroid cells, foamy histiocytes and tangible body macrophages. Plentiful emperipolesis displaying numerous large histiocytes with abundant cytoplasm, and occasional nucleolus were detected. The cytoplasm contained vacuoles with engulfed lymphocytes and nuclear debris in various numbers. Lymphoids tangles were also noted. No granuloma or malignant cells seen. These features were suggestive of RDD and excision biopsy was required for proper histopathological assessment.

During the period while awaiting the FNAC results, the swelling increased in size and was painful with minimal pus discharge seen over a small area of the swelling. He was then admitted to ENT ward for intravenous antibiotics and Computed Tomographic (CT) scan neck was arranged. CT scan neck reported an ill-defined nonenhancing soft tissue density at the submental region measuring $2.4 \times 2.5 \times 1.4$ cm (APxWtxCC) with thickening of the overlying skin (Figure 1), associated with surrounding fat streakiness. This lesion obliterated the plane between subcutaneous tissue and the plastysma muscle. Subcentimeter submental nodes were seen deep to this lesion. The CT scan findings together with FNAC results correspond to that of RDD.

The nature of disease was discussed with the patient and he was offered for biopsy of submental swelling for proper histopathological diagnosis of which he refused. The swelling subsequently reduced in size with no further intervention.

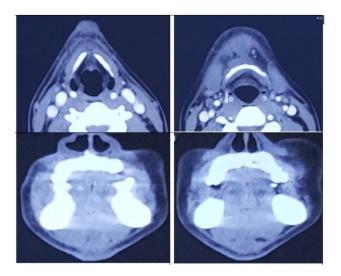


Figure 1: Axial cut of CT scan with contrast shows illdefined non-enhancing soft tissue density at submental region associated with adjacent fat streakiness.

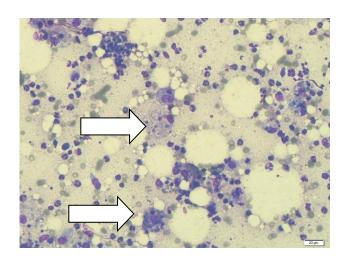


Figure 2: FNAC of submental swelling showing emperipolesis (arrow), seen under microscope at power x40, using May-Grunwald Giemsa (MGG).

DISCUSSION

Sinus histiocytosis with massive lymphadenopathy was first described as a newly recognized benign clinicopathological entity by Dr. Rosai and Dr. Dorfman in 1969 who reported 4 patients with such illness. It is a benign histiocytic disorder of unknown origin that is extremely rare. For the past 48 years, there are about 650 cases reported in medical literature. The disease is more commonly seen in males than females and in individuals of African descent. It often occurs in young adults under the age of 20. Extranodal disease accounts for 43% of cases and the skin is the most common site. We seldom encounter a case of RDD among Asians like that of our patient.

The disease may be explained by a cytokine-mediated migration of monocyte involved in accumulation and activation of histiocytes. This cytokine-mediated activity could be triggered by various stimuli such as autoimmune diseases, hematological malignancies, and post infectious conditions. Viral infection including Herpesvirus-6 and Ebstein-Bar virus has been implicated as causative agent. We suspect that our patient's condition was a result of post infectious stimuli as he presented with prodromal symptoms of viral infection of the upper respiratory tract.

In RDD, histologically, the lymph nodes show pericapsular fibrosis and dilated sinuses, heavily infiltrated with large histiocytes, lymphocytes and plasma cells. Emperipolesis has been considered an important indicator of this disease even though it is not a pathognomonic marker for RDD. This phenomenon can be found in several haematological diseases such as myeloproliferative disorders and lymphoma as well as non-malignant diseases. Our patient FNAC showed histological characteristic of RDD however, further immunostaining is required for confirmation which include S-100 positive and CD1a negative. A further

need for biopsy was discussed with the patient but he refused.

RDD usually has a benign course and treatment is not necessary in most cases unless there is involvement of vital structures.¹ Involvement of the central nervous system and larynx may need targeted therapies which include surgery, radiotherapy, chemotherapy and immunotherapy.³ We have opted to manage our patient conservatively and he is currently progressing well with the submental swelling reducing in size.

CONCLUSION

RDD is a rare cause of cervical lymphadenopathy. One should first rule out more common causes of cervical lymphadenopathy or life threatening conditions prior to coming to a differential of RDD. We hope this case will help clinician to recognize the clinical presentation and histological findings in patient with RDD.

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