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Case Report

An Unusual Clinical Presentation of Extranodal Rosai-Dorfman Disease – A Rare Case Report

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Abstract

Rosai-Dorfman disease is a rare, idiopathic, benign, histiocytic proliferation, usually seen in children and younger adults. Most common site of involvement is cervical lymph node. However, extra nodal manifestations are seen in significant proportion of cases.This case report discusses the unusual presentation of Rosai-Dorfman disease as mass lesion involving bilateral nasal cavities, paranasal sinuses and orbit along with multiple deposits in dura and cervical lymph node.

Keywords: Rosai-Dorfman disease, extranodal, lymph node, nasal cavity, paranasal sinuses, emperipolesis.

INTRODUCTION

Rosai-Dorfman-Destombes disease is an uncommon histiocytic disorder first described by Destombes in the year 1965. Later Rosai and Dorfman described as 'sinus histiocytosis with massive lymphadenopathy' in 1969. The Working Group of the Histiocyte Society of 1987 classified Rosai-Dorfman disease as Non-Langerhans cell histiocytosis [1].

According to recent classification Rosai-Dorfman disease is classified under 'R group' histiocytosis. This histiocytic disorder present with diverse clinical manifestations and outcomes. Most frequent presentation is cervical lymphadenopathy. Extranodal disease may present in locations including the skin, soft tissue, central nervous system, gastrointestinal tract and breast [2]. Rarely extranodal disease can present as mass lesions in nasal cavities, paranasal sinuses and orbit.

Diagnosis is based on routine histopathology and small panel of immune-histochemical markers. Emperipolesis is considered to be pathognomonic and remains as classical histopathological feature [2].

CASE HISTORY

A female patient aged 44 years presented with bilateral nasal obstruction and difficulty in breathing. Clinical examination revealed friable mass lesions involving bilateral nasal cavities, bilateral sinuses, and nasopharynx. On palpation, enlarged lymph node measuring 2x2cm is identified on right side of neck. MRI shows multiple homogenous soft tissue lesions in left orbit, right medial canthal region, paranasal sinuses and left perizygomatic region along with multiple dural based lesions.

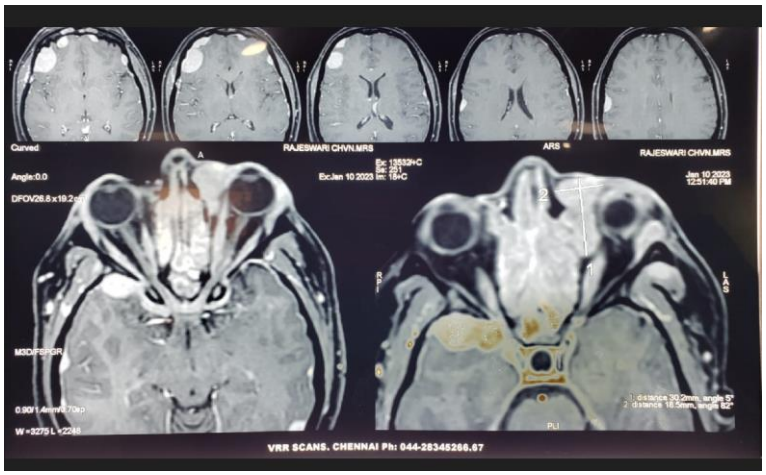

Figure 1: MRI showing homogenous enhancing soft tissue lesions in nasal cavity, paranasal sinuses and dura

Multiple biopsies are taken from bilateral nasal cavities, paranasal sinuses and right cervical lymph node and sent to pathology department for histopathological examination. Formalin fixed, paraffin embedded sections are stained with hematoxylin and eosin stain. Histopathological sections from nasopharyngeal mass reveal respiratory mucosa with extensive areas of squamous metaplasia. Subepithelial stroma shows sheets of large foamy histiocytes admixed with lymphocytes, plasma cells with russel bodies. Few histiocytes exhibit prominent emperipolesis.

Sections from lymphnode reveal expansion of sinuses by sheets of large histiocytes. Emperipolesis is evident in few histiocytes.

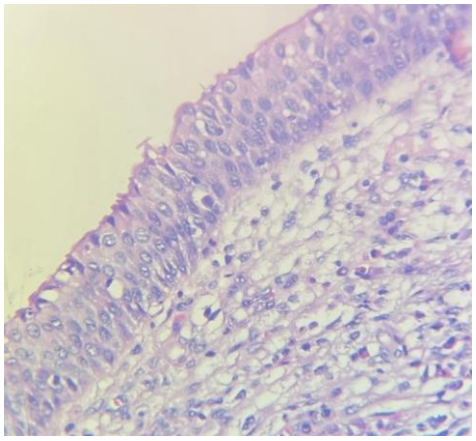

Figure 2: Nasopharyngeal lining epithelium (H&E, X 100)

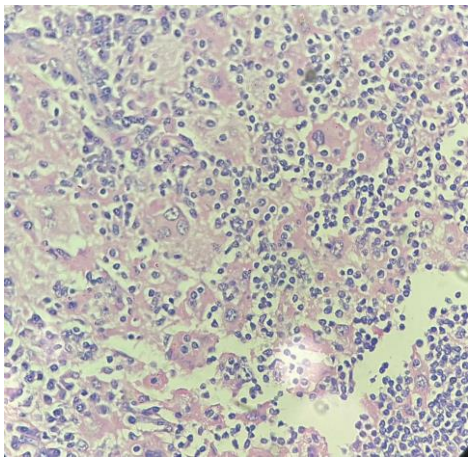

Figure 3: Large histiocytes with evidence of emperipolesis (H&E, X 400)

DISCUSSION

Rosai-Dorfman disease is heterogenous entity that can occur as an isolated disorder or in association with autoimmune, hereditary and malignant diseases [3].

Rosai-Dorfman disease is mostly seen in children and young adults [3]. In our case the age of presentation is 44 years.

Rosai-Dorfman disease commonly present as cervical lymphadenopathy [5]. Extranodal involvement is reported in 43% of cases. Most common extranodal sites are CNS, skin and soft tissues [4].

In the present case, site of presentation of Rosai-Dorfman disease is nasal cavity, paranasal sinuses and orbit, which are relatively uncommon extranodal sites. There is synchronous involvement of cervical lymphnode, perizygomatic region and dura. In the literature, very few cases have been documented in these sites.

In the case study done by Mehotra *et al.,* [6] Rosai-Dorfman disease presented as mass in nasal cavity and paranasalsinuses. Orbital involvement is seen in case series done by Jose G Mantilla *et al.,* [7] In case series done by Jo Ann W Andriko *et al.,* Rosai-Dorfman disease presented as isolated intracranial lesions without nodal involvement [8]. Diagnosis is mainly by histopathological examination of formalin fixed and paraffin embedded tissue sections under light microscopy. Due to rarity and complexity in morphology, multiple biopsies are required for diagnosis [9].

Differential diagnosis of extranodal Rosai-Dorfman disease are chronic inflammation, Rhinoscleroma, Erdheimchester disease, Langerhans cell histiocytosis.

Lesions with chronic inflammation are characterised by infiltration of lymphocytes and plasma cells. But large histiocyte collections with prominent emperipolesis is not seen in chronic inflammation. Rhinoscleroma should be distinguished from Rosai-Dorfman disease in nasal cavity and paranasal sinuses. Vacuolated macrophages, neutrophils and plasma cells are seen under light microscopy in Rhinoscleroma. On Giemsa staining, Bacillus rhinoscleromatis is seen. Emperipolesis is not seen in Rhinoscleroma.

Rosai-Dorfman disease has to be differentiated from other histiocytic disorders with systemic presentation.

Erdheimchester disease is rare non-Langerhans cell histiocytosis, more common in age group above 40 years. Like Rosai-Dorfman disease, it can affect virtually all organs. Microscopic appearance of Erdheim-chester disease shows bland appearing foamy histiocytes, touton giant cells and sparse lymphoplasmacytic infiltrate [10]. Emperipolesis is not seen in Erdheim Chester disease.

The most important differential diagnoses for Rosai-Dorfman disease is Langerhans cell histiocytosis [3]. Langerhans cell histiocytosis primarily affects children and most common sites of involvement are bone, skin, lungs, liver, bone marrow and lymph node [11].

Microscopic features of Langerhans cell histiocytosis are presence of eosinophilic infiltrates, histiocytes containing elongated and grooved nuclei, absence of plasma cell infiltrate. These features differentiate it from Rosai-Dorfman disease.

CONCLUSION

Rosai-Dorfman disease is an uncommon histiocytic disorder most common in children, however can occur in adults. Nodal disease is more common in cervical lymphnode. Extranodal Rosai-Dorfman disease has diverse presentations, nonspecific clinical manifestations and variable outcomes. Diagnosis with specific histopathological features is mandatory in the diagnosis of Rosai-Dorfman disease.

Source of Support: Nil
Conflicts of Interest: Nil

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