Nasal Manifestation of extranodal Rosai – Dorfman disease – A case report

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ABSTRACT

Rosai – Dorfman disease, i.e. Sinus histiocytosis with massive lymphadenopathy (SHML) is a rare, idiopathic, non-neoplastic histiocytic disorder. The most common presentation is painless lymphadenopathy in a young patient. However in 25% of cases it presents as mass in skin, soft tissue, nasal cavity, eye, bone, or other extranodal sites. We report a case of extra nodal form of Rosai Dorfman disease in a young lady who presented with a nasal mass arising in the left inferior turbinate which clinically mimicked malignancy, and was treated with Endoscopic resection.

KEYWORDS: Rosai – Dorfman disease, Sinus histiocytosis with massive lymphadenopathy (SHML)

INTRODUCTION

Rosai – Dorfman disease, i.e. Sinus histiocytosis with massive lymphadenopathy (SHML) is an idiopathic, nodal based histiocytic proliferative disorder that usually resolves spontaneously [1]. It was first described by Rosai and Dorfman in 1969. Extranodal sinus histiocytosis is a form of pseudotumor involving the extranodal sites, which is independent of the lymph node status. The head and neck region is the common extranodal areas affected by SHML. The other extra nodal sites being, orbit, ear, GIT, meninges and brain [2].

CASE REPORT

A 25yr female presented with history of mass in the nasal cavity. On clinical examination, there was a polypoidal and bosselated mass arising from the left inferior turbinate, completely obstructing the nostril. The neck examination revealed a single discrete cervical lymph node measuring 2x2 cm. Gross examination of the neck mass revealed multiple friable bits of grey brown tissue. Lymph node measured 2x2 cm. Cut section of the node showed grey brown to grey white areas (Figure 2).

Microscopy of the nasal mass revealed, normal stratified nasal epithelium, and the subepithelium showed dense aggregates of chronic inflammatory cells comprising of lymphocytes, plasma cells and histiocytes, which were surrounding the seromucinous glands but not destroying them (Figure 3). The presence of histiocytes gave a mottled appearance. The histiocytes were in aggregates, which had abundant eosinophilic, granular to foamy to clear cytoplasm with round to oval vesicular nucleus and prominent nucleoli. These were SHML cells or Rosai Dorfman Cells (RD Cells) (Inset of figure 3).

Emperiploesis which is characterized by histiocytes with phagocytosed lymphocytes was noted (Figure 4). The lymphocyte and plasma cells infiltrates were also noted. Russell bodies were noted in the cytoplasm of plasma cells. Areas of fibrosis and congested blood vessels also noted. Special stains Periodic Acid Schiff (PAS), Grams stain, were negative (to rule out Rhinoscleroma) and Zeihl Neelsen (ZN) stain for Acid Fast Bacilli (AFB) was also negative (to rule out tuberculosis).

The sections from the lymph node revealed an effaced architecture. Most of the lymphnode was replaced by...
plasma cells, SHML cells. Para-amyloid and few atrophic follicles were seen. Emperipolesis was also noted. A close follow up of patient till now reveals no evidence of any disease or recurrence.

**Figure 1:** CT Scan: showing the mass arising from left inferior turbinate (Arrow).

**Figure 2:** Gross: Cut section of the lymph node showing grey white and grey brown areas.

**DISCUSSION**

SHML was identified and described by Rosai and Dorfman in 1969. It was initially characterized as a massive, bilateral and mostly cervical lymphadenopathy[2]. It is also established that SHML can affect the extra nodal sites in the form of pseudo tumor. The head and neck region represents one of the more common extranodal areas to be affected by SHML representing 25% of cases of extra nodal SHML. In the head and neck, there is predilection for the nasal and paranasal sinuses [3]. The diagnosis of extra nodal Rosai – Dorfman disease can be challenging particularly in areas in which the incidence of pulmonary tuberculosis is high and extra pulmonary tuberculosis is common [3].

The histological hall mark of SHML is the tendency for the infiltrate to recapitulate lymphnode architecture. This is referred to as a “Sinusal” pattern in which the clustering of lymphocytes simulates the appearance of germinal centers with surrounding cellular proliferation and dilated “Sinuses”. Emperipolesis is a standard and diagnostic feature in relation to nodal based SHML. However, emperipolesis has been considered to be or less frequently identified feature when extra nodal sites are affected, as is seen in present case. In an analysis of 14 cases by Weing et al[1], emperipolesis was identified in all the cases, but varied from being prominent and readily identifiable to being less conspicuous and only seen in an occasional histiocytes.
Fibrosis is another important finding in association with SHML in extranodal site. Weing et al [1] demonstrated fibrosis in 9 out of 14 cases. In the present case fibrosis was a prominent feature. However similar case study by Sook-RM et al [4] showed that presence of focal fibrosis made it difficult to differentiate this condition from inflammatory pseudotumor. Rosai – Dorfman disease of extranodal site may be confused with, infectious diseases, midline granulomatous destructive disease (Wegener’s granulomatosis, and midline malignant reticulosis), eosinophilic granulomas, Hodgkins disease and fibro-inflammatory lesions.

Rhinoscleroma is an infectious disease with predilection for the upper aero-digestive tract which is more commonly seen in this region. It is also characterized by diffuse cellular proliferation of polymorphonuclear cells lymphocytes, plasma cells and macrophages called “Mikulicz cells” which have foamy cytoplasm in which the organisms can be identified by gram stain, PAS or warthin starry. More over Rhinoscleroma does not have the sinusoidal pattern nor there evidence of emperipolesis.


In the present case, the gram stain, PAS were negative and there was emperipolesis evidently seen in the nasal mass and lymphnode. RD disease is an indolent and self limiting disease. However disseminated nodal and extra nodal disease has increased morbidity and mortality [1]. Loco-regional infiltration of the nasal cavity by Rosai-Dorfman disease can be effectively treated with endoscopic resection. This makes it important to recognize the entity as early as possible. However, cases with persistence of symptoms can be treated with a combination of surgery, radiotherapy and chemotherapy [3].

**CONCLUSION**

The present case highlights the importance of considering the diagnosis of Rosai-Dorfman disease in differential diagnosis of nasal masses as the clinical and histopathologic features of the disease may be overlooked because of scarcity of cases.

**REFERENCES**


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