Unmasking the Enigma: A Rare Case report of Paranasal Sinus Rosai-Dorfman Disease

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Abstract

Rosai-Dorfman Disease (RDD) is a benign histiocytic disease of unknown pathology. The diagnosis of such disease is mainly via histochemical examination. The extranodal manifestation of RDD in the paranasal sinus without lymphadenopathy is a rare and has limited reported literature. Herein, we report a case of an adult Asian male, who presented with a progressive history of left sided nasal blockage associated with facial pain, mouth breathing and reduce in smell. Here-in we report a case of a 50 years old, Asian, male, who presented with a progressive nasal blockage with facial pain, hyposmia and mouth breathing. The challenge in facing with this case is the post diagnostic management, as there are various management strategies outline in the available literature, which range from watchful waiting, various medical therapy and surgical excision.

Keywords: Rosai-Dorfman Disease, Extranodal Histiocytosis, Nasal Cavity.

Introduction

Rosai-Dorfman Disease (RDD) represents an etiologically unknown pathology characterized by the non-malignant proliferation of distinct histiocytic cells. First identified by Rosai and Dorfman in 1969, it was initially described in a case series featuring massive cervical lymphadenopathy with specific histopathological features.[1] While RDD typically presents with massive cervical lymphadenopathy and a low-grade fever, it is noteworthy that 43% of RDD cases manifest extranodal symptoms.[2] This disease exhibits diverse clinical manifestations and outcomes, affecting nearly every organ system.

A definitive diagnosis of RDD is established through histopathological examination, which reveals a marked proliferation of sinus histiocytes, accompanied by the phagocytosis of lymphocytes and erythrocytes by histiocytes.[3]

In this report, we detail a case involving an adult male who presented with a nasal polypoidal mass originating from the nasal septum. The diagnosis of extranodal RDD was confirmed through surgical excision and subsequent histopathological examination, aligning with the characteristic features of the disease.

Case Report

A 50-year-old Asian male, with no known medical conditions, presented at our outpatient Ear, Nose, and Throat clinic with a progressive nasal obstruction accompanied by persistent nasal discharge, diminished sense of smell, mouth breathing, and facial pain persisting for a duration of one year. There was no reported history of trauma or

prior surgical procedures. He denied any complaint of blood-stained nasal discharge, headache, or vision problems. The patient is a chronic smoker employed as a cattle breeder, had an unremarkable medical and surgical history, and no other associated otorhinolaryngological or systemic symptoms were identified.

Upon nasal examination, a widened nasal bridge with a mass occupying the left nasal cavity and no observed telechantus. A confirmatory Rigid Nasoendoscopy was conducted, revealing a polypoidal fleshy mass originating from the septal mucosa and occupying the entire left nasal cavity with some soft tissue seen over the right osteomeatal complex. The mass was multilobulated, soft, painless and do not bleed upon manipulation. Additional examinations of the ear, nose, throat, and systemic systems indicated the absence of other pathologies or lymphadenopathy.

A computed tomography (CT) scan of the paranasal sinus revealed an extensive soft tissue lesion over the bilateral nasal cavity, more over the left side, and involvement within the maxillary, ethmoid, and sphenoid sinuses, with extension toward the nasopharyngeal cavity. No evidence of association or extension into the base of the skull was identified [Figures 1 and 2]. A chest radiography shows no significant findings. [Figure 3]

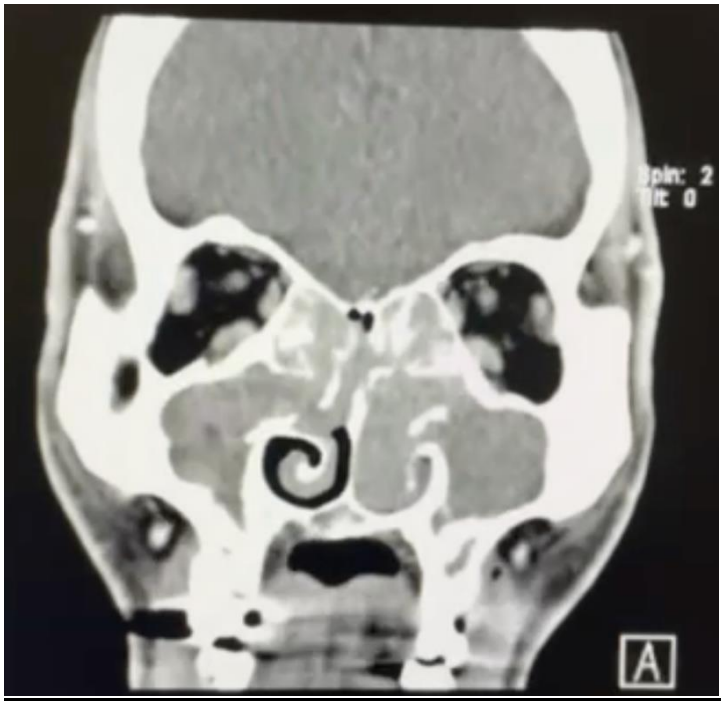

Figure 1: Axial cut of CT PNS showing a soft tissue mass occupying bilateral nasal cavity with extension into bilateral maxillary sinuses (M), opacification is seen more in the left nasal cavity (*).

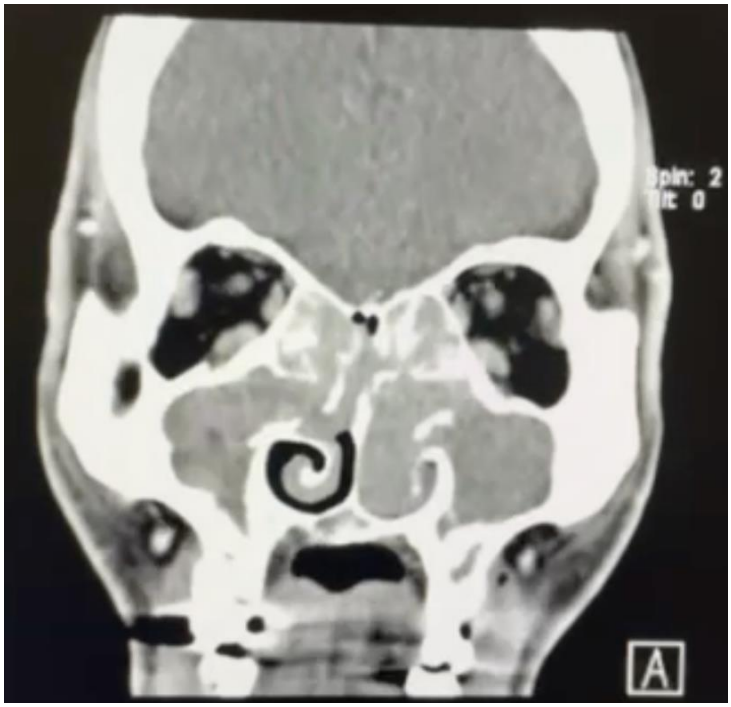

Figure 2: Coronal cut CT PNS showing a soft tissue mass occupying more on the left side with involvement of the nasal septum (*small arrow*).

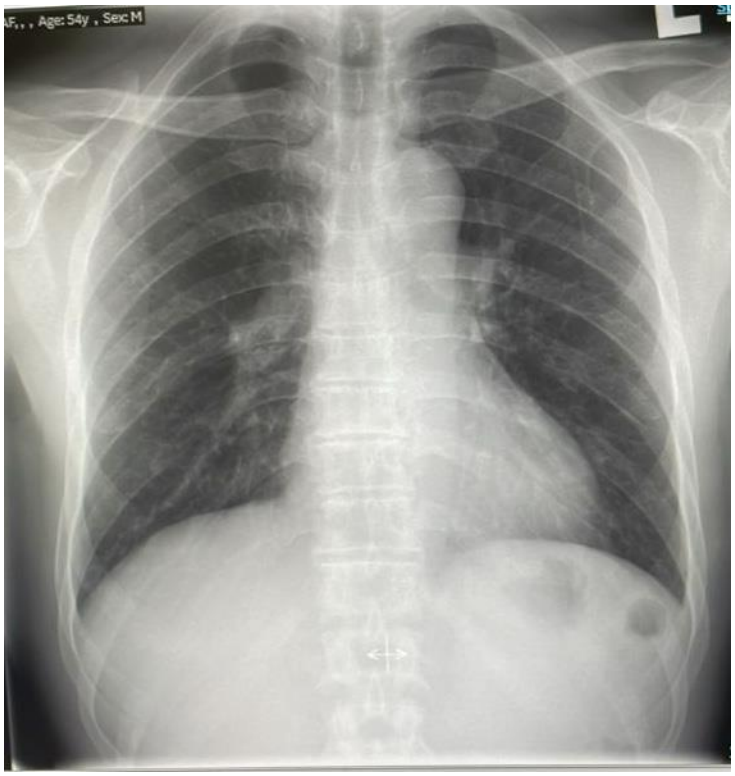

Figure 3: Chest Radiography showing no significant findings.

The patient underwent Functional Endoscopic Sinus Surgery (FESS) for the nasal mass, and histopathological examination revealed an abundance of large histiocytes with numerous plasmocytes. Immunohistochemistry staining was positive for S100 and CD163. These features are consistent with a diagnosis of Rosai-Dorfman Disease.

Subsequent follow-up visits at 1 week and 3 months post-operation showed no signs of residual or recurrent symptoms.

Discussion

Rosai-Dorfman disease (RDD), also known as Sinus histiocytosis with massive lymphadenopathy (SMHL), is a rare pathology of unknown etiology. Various hypotheses regarding its cause include potential involvement of the Epstein-Barr virus (EBV), probable gene mutations, and genetic predisposition; however, none of these hypotheses has been conclusively proven as the definitive etiology.[3] The clinicopathological aspect of RDD involves the abnormal proliferation of histiocytes, presenting with diverse clinical pictures either in isolation or in conjunction with other diseases. This necessitates an integrated diagnostic approach encompassing clinical, radiological, pathological, and molecular perspectives. While lymph nodes are commonly affected, approximately 43% of cases involve extranodal organs, with the skin and soft tissues being the most commonly affected sites.[4] Age predisposition for RDD is contingent upon the subtype, with classical nodal involvement or extranodal involvement exhibiting distinct patterns.

Extranodal involvement is more prevalent in the Asian and Caucasian population, with a mean age of 50 years and a male predominance. In contrast, classical nodal RDD is commonly reported in children and younger adults of African American ethnicity.[5] The presentation of patients plays a crucial role in determining the RDD subtype; classical RDD often presents with isolated cervical lymphadenopathy, whereas extranodal RDD can manifest with a diverse array of symptoms depending on the involved organ. In cases of head and neck involvement, nasal cavity participation accounts for 11% of the study population, often presenting with symptoms such as nasal blockage, epistaxis, and nasal dorsum disfigurement.

Histopathological examination remains the primary means of RDD diagnosis, revealing large histiocytes with round-to-oval nuclei, dispersed chromatin, prominent nucleoli, and abundant clear-to-foamy or vacuolated cytoplasm. S100 expression is a key diagnostic feature, aiding in the visualization of engulfed lymphocytes. Other positive markers in RDD may include Fascin, CD68, CD163, CD4, and CD14.[6]

The treatment and management of RDD remain unclear, as the disease is generally considered benign and self-limiting. Various management strategies exist, ranging from watchful waiting to surgical excision, steroid use, radiation, sirolimus, chemotherapy, immunotherapy, and targeted therapy with imatinib.[7] In the case reported, surgical excision of the mass was performed due to the nasal cavity mass, causing nasal obstruction and significantly impacting the patient's daily activities.

Conclusion

Extranodal RDD without lymphadenopathy of the paranasal sinus is a rare occurrence. RDD is primarily diagnosed through clinicopathological testing. Though it is extremely uncommon, extranodal RDD should be considered in cases with malignancies in the nasal cavity. There is no defined optimum approach for treating this condition; nevertheless, if the lesion creates a functional impairment, it would be prudent to surgically remove it.

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