Paranasal Sinus Rosai-Dorfman Disease: Report of a Rare Case

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Abstract

This report presents a rare case of Rosai-Dorfman Disease (RDD), a benign histiocytic disease. The patient, a 50-year-old Asian male, presented with progressive left sided nasal blockage associated with facial pain, mouth breathing, and hyposmia. A diagnosis of RDD was made. The extranodal manifestation of RDD in the paranasal sinus without lymphadenopathy is rare. The post diagnostic management of this case was challenging, as the literature provide a variety of management strategies ranging from watchful waiting, medical therapy, and surgical excision.

Keywords: Rosai-Dorfman Disease; Extranodal Histiocytosis; Nasal Cavity; Malaysia

Introduction

Rosai-Dorfman Disease (RDD), also known as sinus histiocytosis, 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] RDD typically presents with massive cervical lymphadenopathy and a low-grade fever, but nearly half (43%) of the cases manifest extranodal symptoms.[2] This disease exhibits diverse clinical manifestations and outcomes, potentially affecting nearly all organ systems.

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 present 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 Ear, Nose, and Throat (ENT) outpatient clinic with a progressive nasal obstruction accompanied by persistent nasal discharge, diminished sense of smell, mouth breathing, and facial pain persisting for one year. There was no reported history of trauma or prior surgical procedures. He denied any complaint of bloodstained nasal discharge, headache, or vision problems. The patient, who was employed as a cattle breeder, revealed he was a chronic smoker. He had an unremarkable medical and surgical history, with no other otorhinolaryngological or systemic symptoms.

Nasal examination revealed a widened nasal bridge with a mass occupying the left nasal cavity with no observable telecanthus. 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 did not bleed on 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 showed no significant findings. [Figure 3]

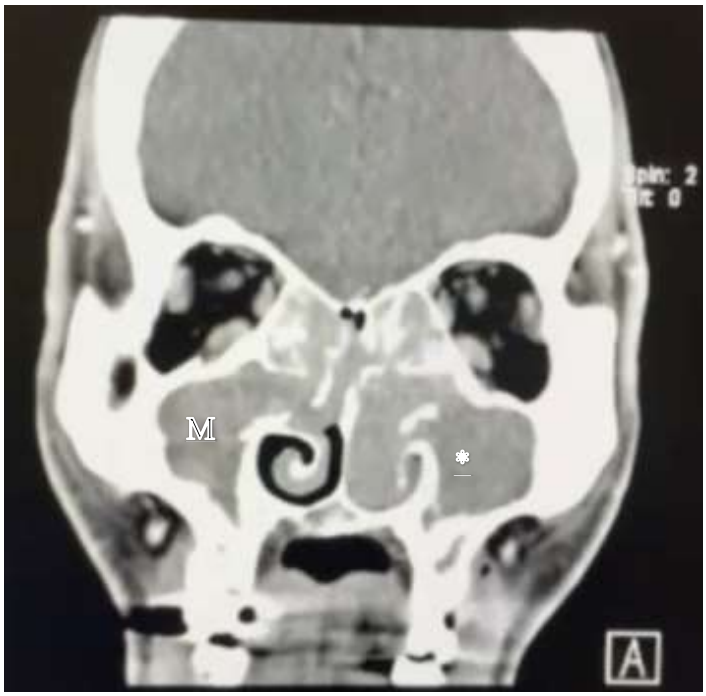

Figure 1: Axial cut of computed tomography paranasal sinus (CT PNS) showing a soft tissue mass occupying bilateral nasal cavity with extension into bilateral maxillary sinuses (M). Opacification is seen more in the left nasal cavity (*).

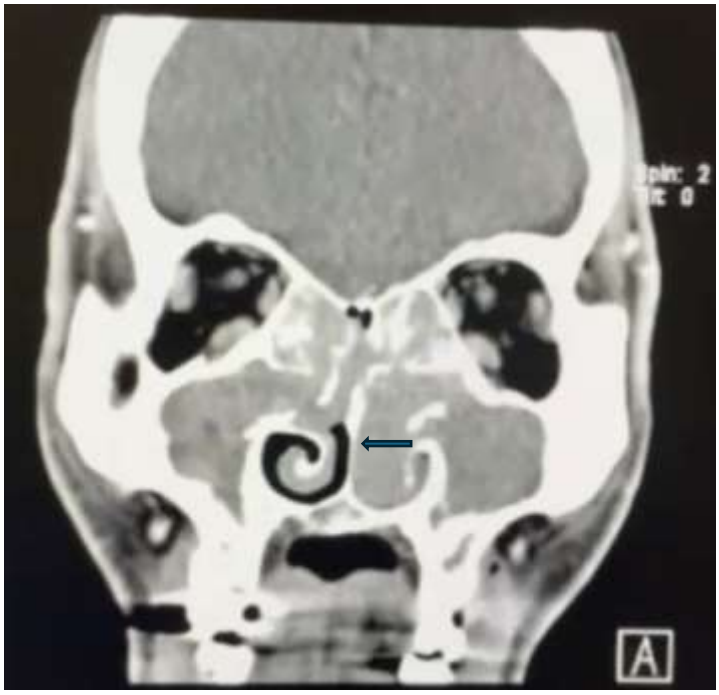

Figure 2: Coronal cut computed tomography paranasal sinus (CT PNS) showing a soft tissue mass occupying more on the left side with involvement of the nasal septum (*small arrow*).

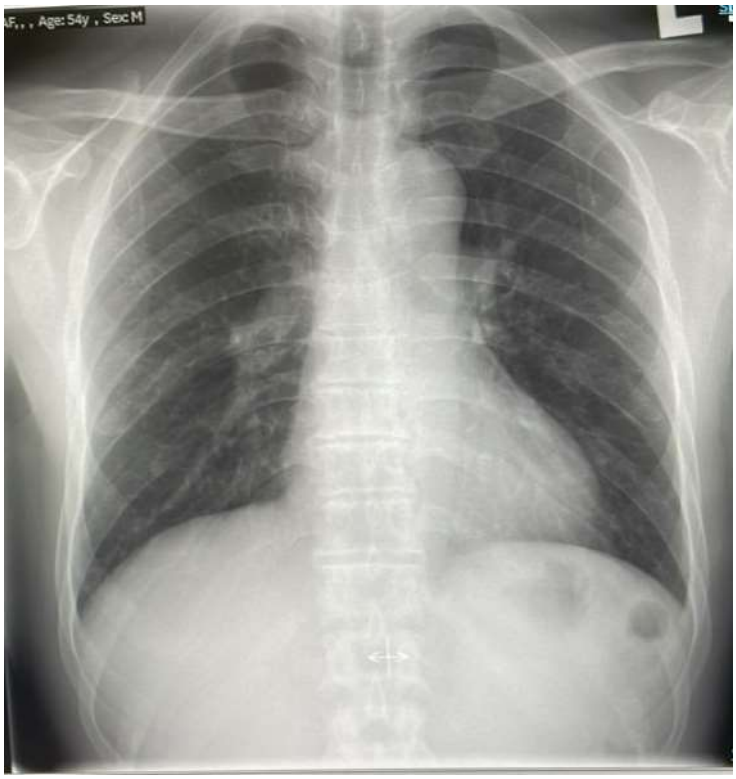

Figure 3: Chest radiograph, which shows no significant finding.

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.

The patient was discharged on Day 4 post surgery. Subsequent follow-up visits at one week and 3 months post-operation revealed no residual or recurrent symptoms.

Discussion

Rosai-Dorfman disease (RDD) 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. None of these hypotheses has been conclusively proven.[3] The clinicopathological aspect of RDD involves an abnormal proliferation of histiocytes with diverse clinical presentations 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, mostly the skin and soft tissues.[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 populations, with a mean onset 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. Head and neck involvement and nasal cavity participation has been reported with about 11% prevalence, often presenting with symptoms such as nasal blockage, epistaxis, and nasal dorsum disfigurement.

Histopathological examination remains the primary method for diagnosing RDD, typically 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 the 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 considered benign and self-limiting. Management strategies range from watchful waiting to surgical excision, steroid use, radiation, sirolimus, chemotherapy, immunotherapy, and targeted therapy with imatinib.[7] In the current case, surgical excision of the mass was performed because the mass was causing nasal obstruction and significantly impacting the patient's quality of life.

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.

Disclosure

The authors declare no conflicts of interest. Informed consent was obtained from the patient.

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