

# Extranodal Rosai-Dorfman Disease of the Nasal Septum: A Case Report

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## ABSTRACT

Rosai-Dorfman Disease (RDD) is a rare, benign histiocytic proliferative disorder mostly involving the lymph nodes. Extranodal disease can occur in the skin and soft tissue including the mucosal surface, central nervous system, gastrointestinal tract and breast. Here, a case of a 17-year-old Indo-Asian girl is presented, who came to the Ear, Nose and Throat (ENT) Outpatient Department (OPD) with a mass in her left nostril for the past two years. A sessile globular mass from the left nasal septum was seen during diagnostic visual endoscopy. A Computerised Tomography (CT) scan of the paranasal sinus revealed a rather large right maxillary sinus with a soft tissue density lesion filling it and obliterating the osteomeatal complex, while blood tests revealed an increased eosinophilic count. The nasal tumour was removed, and its histological analysis revealed characteristics of RDD. This is typically sporadic histiocytosis with emperipolesis in the cells. Clinically, the condition is benign, and all that is required is conservative therapy and periodic monitoring of its progression. Surgical excision is rarely advised until the disease has grown to an unmanageable size and is causing respiratory impairment or a cosmetic deformity. Currently, therapy revolves around surgical and immunosuppressive treatments, but the optimal diagnostic and therapeutic management of RDD remains to be defined.

**Keywords:** Deviated nasal septum, Histiocytosis, Inferior turbinate hypertrophy, Septal mass

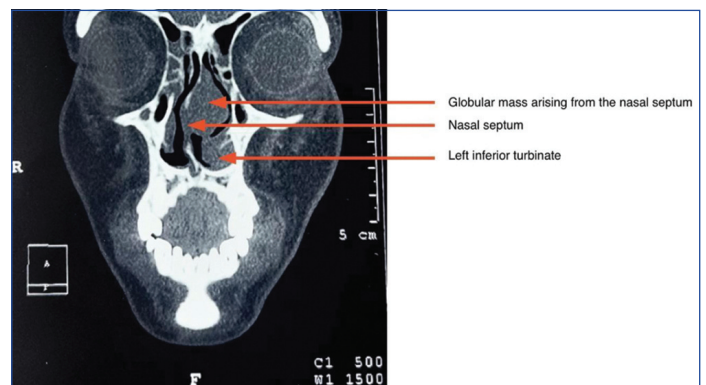
## CASE REPORT

An 17-year-old Indo-Asian girl reported to the ENT OPD with complaints of nasal mass in the left nostril for the past two years. The swelling was gradually progressive in size and painful to the touch; it was associated with a running nose and frequent sneezing. There was also a history of snoring and recurrent allergy to dust in the past two years, which were being treated with topical nasal drops and antihistaminics. The allergies were exacerbated during the winter season. The swelling did not bleed on the touch. There was no history of discharge.

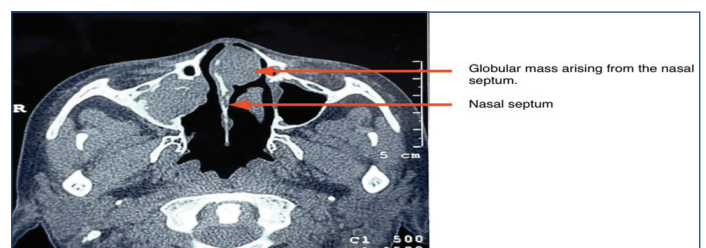
A physical examination of the nose revealed a globular mass arising from the left-side of the nasal septum with the deviated nasal septum towards the right-side and a nasal spur on the right-side. A sessile globular mass from the left nasal septum was visualised on diagnostic nasal endoscopy. Laboratory investigations revealed an elevated eosinophilic count ( $>40 \times 10^3/\mu\text{L}$ ).

A CT scan of the paranasal sinus revealed that the left maxillary sinus was relatively enlarged in size, with a soft tissue density lesion filling it and obliterating the osteomeatal complex. The periosteal reaction was seen circumferentially along the inner margin of the right maxillary sinus. A soft tissue density polypoidal lesion was seen along the left lateral margin of the nasal septum, measuring 18×22×27 mm in craniocaudal×transverse×anteroposterior dimensions. Mild mucosal thickening was seen in the right frontal, bilateral ethmoid, bilateral sphenoid and left maxillary sinuses [Table/Fig-1,2].

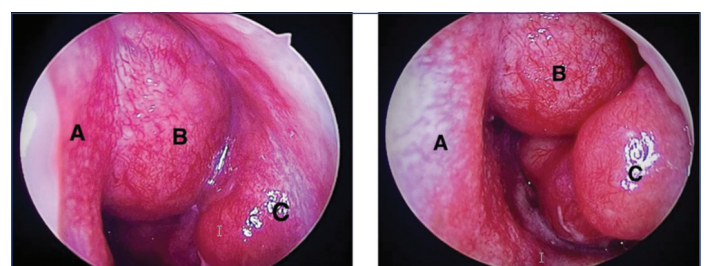
The nasal septum deviated to the right-side with hypertrophy of the left inferior turbinate. The left osteomeatal complex was normal. Bilateral orbital margins were intact, and the overlying soft tissue was normal. All preoperative investigations, including complete blood count (excluding eosinophilic count), serum electrolytes, viral markers including Human Immunodeficiency Virus (HIV), Liver Function Tests (LFTs), Kidney Function Tests (KFTs), Prothrombin Time (PT), Activated Partial Thromboplastin Time (APTT), chest X-ray were within normal limits. Under general anaesthesia, the growth was surgically removed, and the left inferior turbinoplasty was performed using a debrider. The pre-operative view of the left nostril showing the smooth swelling arising from the nasal septum on the left-side can be seen in [Table/Fig-3,4]. The intraoperative view of the septal mass removed in toto



**[Table/Fig-1]:** On the coronal section of CT face, nasal septum is seen to be deviated to the right-side. A globular mass can be seen arising from the left-side of the nasal septum with hypertrophy of the left inferior turbinate hypertrophy (red arrows).

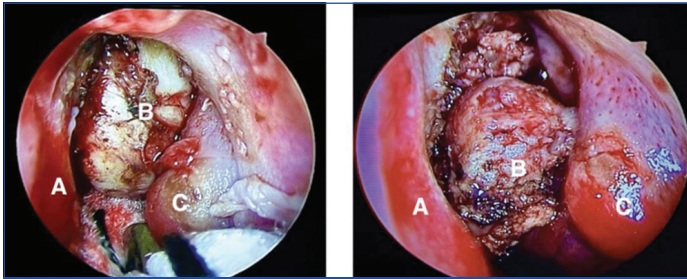


**[Table/Fig-2]:** On axial section of CT face nasal septum is seen to be deviated to the right-side. A globular mass can be seen arising from the left-side of the nasal septum. Mucosal thickening can be appreciated in the right maxillary sinus (red arrows).

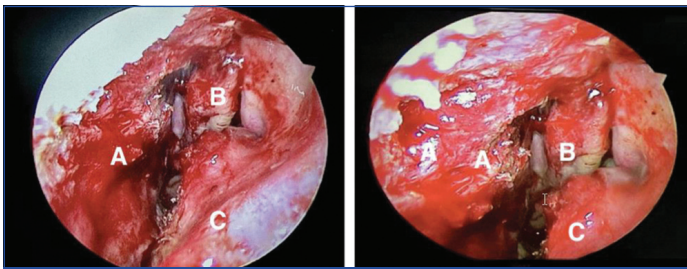


**[Table/Fig-3,4]:** Preoperative view of the left nostril showing the smooth swelling arising from the nasal septum on the left side. (Images from left to right) A: Nasal Septum; B: Globular Mass; C: Inferior Turbinate

can be seen in [Table/Fig-5,6]. Postoperative view of the raw area of nasal septum after removal of septal mass can be seen in [Table/Fig-7,8]. With 'A' being the Nasal Septum, 'B' being the Inferior Turbinate, and 'C' being the globular mass in each figure.

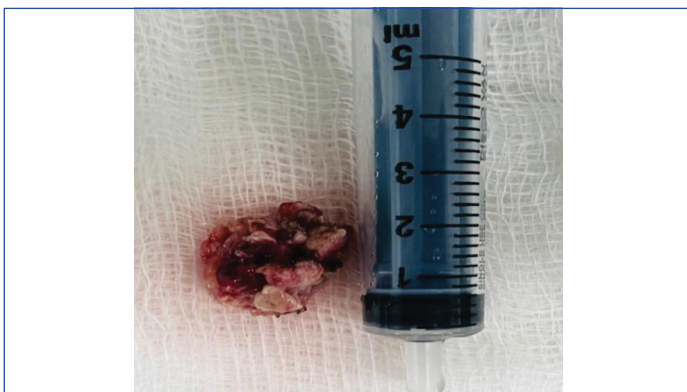


[Table/Fig-5,6]: Intraoperative view of septal mass removed in toto. (Images from left to right)  
A: Nasal Septum; B: Globular Mass; C: Inferior Turbinate

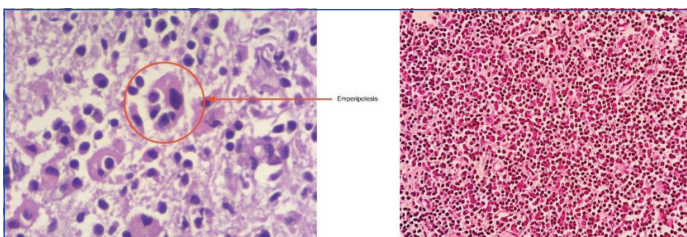


[Table/Fig-7,8]: Postoperative view of raw area of nasal septum after removal of septal mass. (Images from left to right)  
A: Raw area of Nasal Septum; B: Middle Turbinate; C: Inferior Turbinate

The mass was excised as a whole and sent for histopathological examination [Table/Fig-9]. It revealed fragmented tissue bits focally lined by respiratory epithelium showing mild reactive atypia. Stroma showed dense and diffused chronic inflammatory cells with sheets of small non cleaved lymphocytes, many plasma cells with rare cells showing binucleation, scattered histiocytes and occasional histiocytes showing emperipolesis of intact lymphocytes and rare occasional eosinophils. In places, stroma showed oedema. No evidence of atypical cells and bizarre cells, atypical mitosis, granuloma, or fungal elements in the section. The finding was consistent with RDD [Table/Fig-10-13]. After two weeks of the surgery, the patient was followed-up and maintained optimum health.

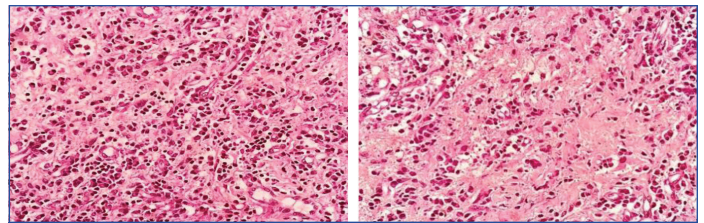


[Table/Fig-9]: Excised mass of size 1.8x1.5 cm.



[Table/Fig-10]: Cells showing emperipolesis (Haematoxylin and Eosin stain, 40x magnification).

[Table/Fig-11]: Lymphoplasmacytic infiltration and aggregation (Haematoxylin and Eosin stain, 20x magnification). (Images from left to right)



[Table/Fig-12]: Sheets of lymphocytes (Haematoxylin and Eosin stain, 20x magnification).

[Table/Fig-13]: Dense areas of Fibrosis (Haematoxylin and Eosin stain, 20x magnification). (Images from left to right)

## DISCUSSION

The RDD is a rare condition with a prevalence of 1:200000 and an estimated 100 new cases yearly in the United States [1]. RDD is rare in India, only about a dozen cases have been reported, mostly in children and very few in adults [2]. It is a malignant, non Langerhans cell histiocytosis of unknown cause characterised by an accumulation of activated histiocytes within the affected tissue. RDD is more prevalent in men and people of African origin, with the cutaneous variant being more prevalent in Asian women [3]. Constitutional symptoms and laboratory signs of inflammation may exist but are not mandatory [4]. Although, it is widely documented in lymph nodes and other organs, it is frequently not recognised in soft tissues. RDD exists in two main varieties: solely Cutaneous RDD (CRDD) and one that affects the lymph nodes and, in some cases, extranodal organs [5,6]. The classic presentation of RDD consists of bilateral, massive and painless cervical lymphadenopathy with or without intermittent fevers, night sweats and weight loss [7]. The extranodal illness, which most frequently affects the skin, nasal cavity and orbit, develops in about 43% of individuals [8]. The testes, spleen and salivary glands may be affected [9,10]. Although, highly unusual, cutaneous illness without lymphadenopathy has been seen [11,12].

Nasal obstruction and epistaxis are two frequent symptoms brought on by the extranodal invasion of the sinuses and nasal cavity. Rarely is the nasal septum invaded. Leukocytosis and an accelerated Erythrocyte Sedimentation Rate (ESR) typically describe laboratory findings. Due to the lack of pathognomonic clinical characteristics, nasal RDD is frequently misdiagnosed in clinical practice [13]. The absence of typical RDD symptoms at the presentation made it difficult to diagnose present case. Despite laboratory results indicating eosinophilia, her body temperature, ESR and other organ involvement were all within normal range. Raje P and Vyas P, reported a case in India of a 58-year-old female patient with extranodal RDD of nasal septal mucosa without lymphadenopathy who had presented with complaints of difficulty in breathing and frequent episodes of epistaxis for three months [2].

Wang J et al., presented a case of a 55-year-old woman who presented to the hospital with nasal dorsum collapse while wearing her glasses with occasional epistaxis. Wang J et al., reported a similar observation in which the patient had no clinical signs of RDD and the laboratory results only revealed leukocytosis [13]. A case by Akyigit A et al., reports a similar finding: 15-year-old male patient: the mass completely filled the left nasal meatus and caused difficulty in breathing through both nasal meatus while deviating the septum to the right [14]. The present case is of an Asian Indian girl with RDD with nasal obstruction without any lymphadenopathy, making it very rare.

The traits of RDD in bone and soft tissue are ambiguous and frequently deceptive. Erdheim-Chester disease, lymphoma, plasma cell myeloma and metastatic disease are among the differential diagnosis [15]. Several differential diagnosis (nasal polyp, inferior turbinate hypertrophy and nasal tumours) were ruled out in the present instance. As the mass is arising from the septum and it is firm too hard in consistency, the possibility of the nasal polyp was ruled out clinically. As the mass was attached to the septum and on probe test there was no lateral attachment, and inferior turbinate hypertrophy was ruled out. The mass did not bleed on touch which



is contrary to the findings reported by Raje P and Vyas P and Wang J et al., [2,13] and there was no bone erosion as noted on the CT-PNS scan which was contrary to the case reported by Wang J et al., the possibility of nasal tumours was ruled out [13].

The RDD is characterised histologically by an attenuated dermal infiltration of pale histiocytes and lymphoplasma cells of varying sizes [16]. These histiocytes have single prominent nucleoli and round or oval vesicular nuclei with well-defined, delicate nuclear membranes. Neutrophils and eosinophils are occasionally seen. RDD is incredibly uncommon, the lesions are clinically varied, and it can mimic various conditions, including other histiocytoses, lymphomas, sarcoidosis, viral diseases and other histiocytoses [17]. The cytoplasm of the histiocytes frequently contains phagocytised lymphocytes and plasma cells, a finding known as emperipolesis in the histology of RDD [18]. Since, extranodal RDD can be imperceptible and extranodal lesions typically have greater fibrosis, fewer RDD histiocytes, and less emperipolesis, emperipolesis is not necessary for the diagnosis [19]. Histological analysis of the mass removed in this case revealed cells with emperipolesis combined with thick fibrotic regions, lymphoplasmacytic infiltration and aggregation, supporting RDD as the origin of the mass which is similar to findings of the cases reported by Raje P and Vyas P; Wang J et al., and Akyigit A et al., [2,13,14]. Since, RDD's course typically self-limits, close observation is the advised management strategy. Surgical excision is the most effective course of action when management is necessary, such as in lesions that are physically compressing surrounding tissue or in lesions that are affecting internal organs. Oral steroids should be started in cases of fever >38°C without a known illness and unexpected lymph node enlargement [17]. Although, anecdotal reports of the use of corticosteroids, thalidomide, radiation and alkylating drugs exist, their efficacy has not been established [20]. Various treatment approaches have been suggested for individuals based on prior case reports, including cryotherapy, surgical excision, irradiation, oral corticosteroids, dapsone, thalidomide and isotretinoin [21,22].

In the case reported by Raje P and Vyas P, the mass was endoscopically excised [2], in the case presented by Akyigit A et al., the mass was excised surgically [14], and in the case presented by Wang J et al., most of the diseased tissue was excised for pathological examination [13]. However, no further treatment was performed, for the patient's personal reasons. Moreover, in the second postoperative year, the tumour grew back which was then surgically excised following which there was no recurrence. Similarly, the tumour in the patient's case necessitated removal because it was blocking the patient's airway and causing the nasal septum to deviate to the opposite side.

## CONCLUSION(S)

The RDD is a rare, benign proliferative disease mainly affecting the lymphatic tissues, however, the disease may manifest in extranodal sites, including the skin and the mucosal site. The present case discusses an Indo-asian patient with RDD of the nose without

lymphadenopathy. Eventhough, most cases resolve on their own surgical excision is recommended for a larger tumour involving the airway which causes airway compromise just like in the present case. The appropriate management of RDD must be established by well-designed studies to draw firm conclusions.

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