Case Report

Extranodal Sinonasal Rosai-Dorfman Disease in a Nigerian Woman: A Case Report and Review of Literature

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Abstract

Rosai Dorfman disease (RDD) also referred to as sinus histiocytosis with massive lymphadenopathy is a rare non-neoplastic lymphoproliferative disease with unknown etiology and pathogenesis first described by Rosai and Dorfman in 1969. It is characterized by histiocytic proliferation of lymphatic sinuses, which primarily involves the lymph nodes, but has an extranodal tendency. Frequently encountered extranodal sites in the head-and-neck region is paranasal sinuses and nasal cavity, orbit, parotid gland, and middle ear. We present a case of RDD occurring in the nasal cavity of a 28-year-old female from Nigeria. RDD should be considered a differential diagnosis in any patient with benign sinonasal mass especially arising from the nasal septum with massive cervical lymphadenopathy.

Keywords: Extranodal, Rosai-Dorfman disease, sinonasal, unusual disease

Received on: 18-06-20 **Review completed on:** 08-08-20 **Accepted on:** 27-08-20 **Published on:** ****

Introduction

Rosai Dorfman disease (RDD) also referred to as sinus histiocytosis with massive lymphadenopathy is a rare nonneoplastic lymphoproliferative disease with unknown etiology and pathogenesis first described by Rosai and Dorfman in 1969.[1] It is characterized by histiocytic proliferation of lymphatic sinuses, which primarily involves the lymph nodes, but has an extranodal tendency.[2] Sites of extranodal involvement include skin and soft tissue, upper respiratory system, genitourinary tract, eye, orbit, kidney, thyroid, breast, and bone.[3] RDD has been reported in black populations severally in Nigeria and parts of sub-Saharan Africa with involvements of other body regions.[4-9] However, to the author's best knowledge, this is the first reported case of sinonasal RDD in our locality.

Case Report

A 28-year-old female presented with a 1-year history of progressive bilateral nasal obstruction, nasal discharge, recurrent nosebleeds associated with facial disfigurement, and progressive painless multiple neck swellings. Physical examination revealed a young lady with a distorted nasal pyramid, obstructed nasal cavities, deviated nasal septum to the right, fleshy mass in the nasal cavities, mucopurulent discharge, and multiple bilateral huge nontender cervical lymphadenopathy involving levels 1a, 1b, 2, and 3 groups of lymph nodes on the left and group 1b lymph nodes on the right [Figure 1]. She had a similar presentation 7 years back, had excision of the nasal mass through lateral rhinotomy and histology showed undifferentiated squamous cell carcinoma with no further treatment undertaken but recurred 6 years later. Initial histology from an intranasal punch biopsy showed no evidence of malignancy. Blood investigations revealed normocytic normochromic anemia, raised erythrocyte sedimentation rate, leukocytosis, and neutrophilia. Computed tomography scan of the paranasal sinuses and brain showed a mass of homogenous medium density arising from the nasal

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Access this article online

Quick Response Code:

Website: www.atpjournal.org

DOI: 10.4103/atp.atp_37_20

How to cite this article: Ajiya A, Kolo ES, Shuaibu IY. Extranodal sinonasal Rosai-Dorfman disease in a Nigerian woman: A case report and review of literature. Ann Trop Pathol 2020;XX:XX-XX.

cavities (more on the left). It showed significant contrast enhancement and exophytic extension into the maxillary area of the face, posteriorly into the ethmoid, sphenoidal, and frontal sinuses. However, the sphenoid and maxillary sinuses are not directly involved but are compressed by the pressure effect of the mass [Figure 2]. The tumor in the nasal cavities was excised through a left lateral rhinotomy and a bilateral supraomohyoid selective neck dissection was done. Intraoperative findings were that of a fleshy mass filling the left nasal cavity attached to the septum and extending to the right nasal cavity deviating the septum to the right [Figures 3 and 4]. Excision left behind a wide anterior septal perforation. Bony structures appeared intact. The lymph nodes appeared well circumscribed, some matted involving the submental, submandibular, and upper deep cervical on the left and the submandibular on the right [Figure 5]. Other lymph nodes were not palpably enlarged. The patient had both pre- and post-operative blood transfusion. Histology of the excised nasal mass revealed a fibro collagenous tissue exhibiting infiltrate of lymphoplasma cells and numerous histiocytes exhibiting emperipolesis. Excised lymph nodes showed a thick fibrous capsule and effaced native architecture [Figure 6]. The node exhibits sheets of histiocytes exhibiting prominent emperipolesis admixed with lymphoplasmacytic infiltrates [Figure 7]. After discharge from the hospital, the patient was started on oral prednisolone 60 mg daily in divided doses and tapered over 3 months. She is presently still on follow-up.

Figure 1: 28-year-old woman with nasal mass and cervical lymph nodes

Figure 3: Large intra-operative septal perforation after excision of nasal mass

Discussion

RDD is a self-limiting benign lymphoproliferative disease of unknown origin often presenting with massive lymphadenopathy and in some cases involving extranodal sites. RDD has a worldwide distribution and is more common among Africans and Caribbean; it affects all ages, though more common among adults with male predominance. By 2012, only 126 cases of RDD have been reported, while 38 cases of extranodal RDD have been reported in the literature in 2019.[10,11] Most cases of extranodal RDD present in the head and neck. The most common extranodal sites in the head and neck with or without lymphadenopathy include the skin, nasal cavity, paranasal sinuses, eyelid, orbit, bone, salivary glands, and very rarely central nervous system.[12] In the nasal cavity, previous reports have shown the nasal

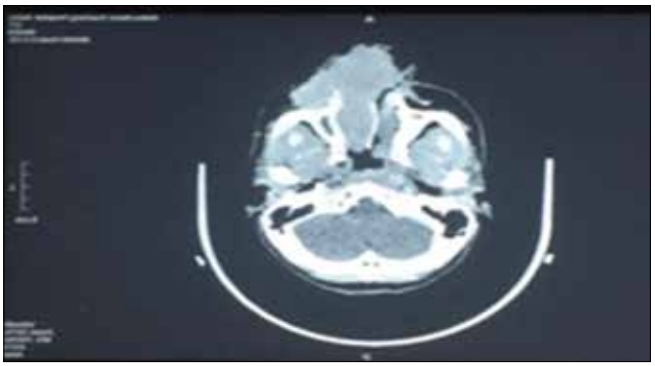

Figure 2: Axial view of the computed tomography scan showing the left nasal mass

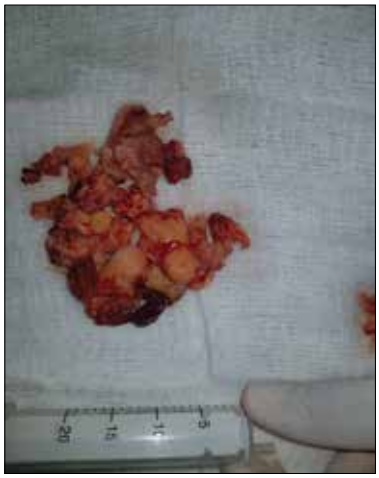

Figure 4: Specimen of excised nasal mass

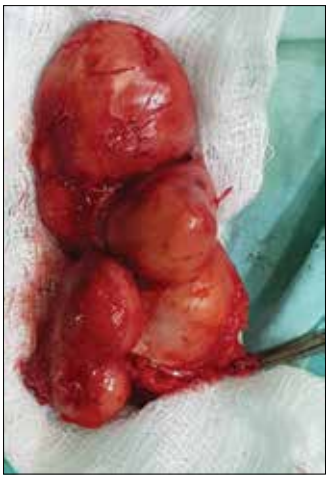

Figure 5: Specimen of excised left cervical lymph nodes

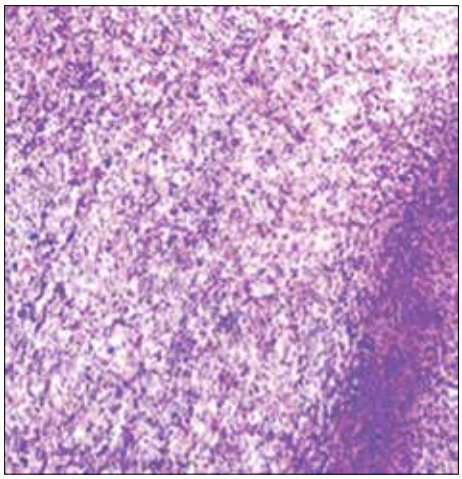

Figure 6: Rosai-Dofman disease showing dilated sinuses, with sheets of histiocytes and large macrophages, some of which contain lymphocytes within their cytoplasm (emperipolesis) H and E, ×10 (nasal mass)

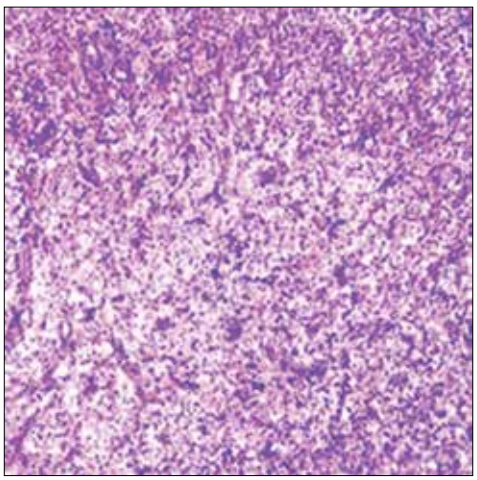

Figure 7: Rosai-Dofman disease showing dilated sinuses, with sheets of histiocytes and large macrophages, some of which contain lymphocytes within their cytoplasm (emperipolesis) H and E, ×10 (cervical lymph nodes)

septum as a commonly affected site consistent with the current case.[1]

Majority of patients with sinonasal RDD present with nasal obstruction, epistaxis, nasal discharge, and hyposmia/anosmia.[12] Our patient had a similar presentation. Blood investigations in these patients commonly show normochromic normocytic anemia, raised erythrocyte sedimentation rate, leukocytosis, neutrophilia, and hypergammaglobulinemia.[12] The current case had a similar blood picture except for hypergammaglobulinemia, which was not requested.

The cornerstone of the diagnosis of RDD is a histopathological examination with immunohistochemistry in challenging cases. The important histopathological finding of the disease is the detection of phagocytosed lymphocytes and occasionally, other cells, like plasma cells and erythrocytes, with undamaged structures in the cytoplasm of the histiocytes, commonly referred to as emperipolesis. Immunohistochemistry shows positive staining with S100 and CD68.[1,12] However, immunohistochemistry was not done in the current case due to its unavailability in the center.

RDD, though, a self-limiting disease is often recurrent and can affect vital human structures, potentially could be fatal or pose significant cosmetic concern requiring intervention as is the case in our patient. There is no generally accepted treatment protocol for RDD. However, corticosteroids, cytotoxic chemotherapy, antivirals, surgery, and radiotherapy have been variously adopted. Spontaneous remission commonly occurs; however, prolonged follow-up is imperative.[12].

Conclusion

Sinonasal RDD with massive cervical lymphadenopathy is rare, especially in sub-Saharan Africa. Diagnostic difficulty and recurrence are attributes associated with the disease. RDD should be considered a differential diagnosis in patients with benign sinonasal mass, especially arising from the nasal septum with massive cervical lymphadenopathy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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