Extranodal Manifestation of Rosai Dorfman Disease of the Nasopharynx

T. Ramadass • P. Thulasi Das • N. Geetha • Nithya Narayanan • Gopi Ayyaswamy • S. Swapna

Abstract

The aim of presenting this case is to highlight the fact that extranodal manifestation of Rosai Dorfman Disease (RDD) without lymphadenopathy was seen in a teenaged girl with recurrent episodes of epistaxis, due to a lobulated mass in the nasopharynx, which mimicked malignancy. This case is a rare presentation because the extranodal manifestation of RDD in multisystem organs are reported in literature, but the nasopharyngeal manifestation without lymphadenopathy is unique. The clinical presentation and surgical management of the case by endoscopic sinus surgery, the aetiopathology, differential diagnosis and review of literature are described here.

Keywords Sinus histiocytosis with massive lymphadenopathy [SHML] Rosai-Dorfman disease [R.D.D] Lymphoma Rhinoscleroma

T. Ramadass (✉)
H.O.D. and Senior Consultant
Dept. of ENT and Head and Neck Surgery
Apollo Hospital
21 Greaves Lane, Off Greaves Road, Chennai - 600 006.
Email: ramadassthot@yahoo.com

P. Thulasi Das
Consultant ENT and Plastic Surgeon
Dept. of ENT and Head and Neck Surgery

N. Geetha
Consultant Pathologist
Dept. of Pathology

Nithya Narayanan
Associate Consultant
Dept. of ENT and Head and Neck Surgery

Gopi Ayyaswamy
Ex-Senior Resident
Dept. of ENT and Head and Neck Surgery

S. Swapna
Resident
Dept. of ENT and Head and Neck Surgery
Apollo Hospitals, Chennai.

Introduction

Rosai Dorfman Disease or Sinus Histiocytosis with massive lymphadenopathy (SHML) is a rare histiocytic proliferative disorder defined currently by its unique histopathological features. While nodal involvement is the typical presentation, extranodal manifestations have frequently been described with or without lymphadenopathy. The recognition of extranodal involvement without concomitant nodal disease is rare, therefore poses a diagnostic problem. The most common extranodal site involved is skin. Involvement of the upper aerodigestive tract, especially nasal cavity, larynx and nasopharynx is uncommon. The importance of recognition of this process, which masquerades as a lymphoreticular neoplasm prompted this case report.

Case Report

A seventeen year old girl presented to us with complaints of headache of seven months duration and five episodes of epistaxis from the left nostril over two months, two of which were profuse requiring anterior nasal packing in her hometown. She also complained of nose block for one month, before admission at Apollo Hospitals, Chennai. General examination did not reveal any significant cervical lymph nodes. On anterior rhinoscopy a lobulated fleshy mass was seen in the left nostril posteriorly, the site of origin of which could not be made out and it did not bleed on touch. Flexible nasopharyngoscopy performed under local anaesthesia showed a fleshy lobulated congested mass in the left nostril, completely occluding the posterior choana and partially occluding the choana on the opposite side.

The bony septum was found to be widened and the lesion extended to the fossa of Rosenmuller on either side. The eustachian tubes were patent. Examination of the ears showed no evidence of secretory otitis media and her hearing was within normal limits. There was no proptosis and the palate seemed to be free of disease. Biopsy from the mass was deferred in view of the bleeding episodes.

Fig. 1 CT Scan of the paranasal sinuses (coronal section) showing the mass obliterating the nasopharynx and eroding the floor of the sphenoid sinus.

Fig. 2 MRI – T1 weighted image (sagittal section) showing superior extension of the Mass into the sphenoid sinus eroding the clivus and basisphenoid.

The CT scan of paranasal sinuses done elsewhere showed a well defined homogenous, solid, uniformly enhancing mass whose epicenter was in the roof of the nasopharynx, eroding the clivus and the floor of the sphenoid sinus, involving the sinus cavity superiorly and extending to the fossa of Rosenmuller on both sides laterally, abutting the medial boundary of parapharyngeal space, measuring 6 x 4.6 x 2.2 cm. (Fig 1).

MRI done at our hospital showed a large midline bulky enhancing soft tissue mass lesion in the nasopharynx with obliteration of nasopharyngeal air column, superiorly eroding the basisphenoid and part of the clivus and extending into sphenoid sinus, anteriorly encroaching into the posterior choana on either side of nasal cavity and the posterior portion of bony nasal septum (Fig 2). Chest X ray, haematological and biochemical parameters were within normal limits except for erythrocyte sedimentation rate, which was raised. Her blood group was O positive.

Patient was taken up for endoscopic examination, biopsy of the lesion and debulking of the tumour mass. Two units of blood were reserved. Under general anaesthesia through orotracheal intubation, the extent of the growth was studied under endoscopic TV monitoring. Biopsy from the nasal mass was done and sent for frozen section. Surprisingly, while doing the biopsy there was not much bleeding, hence we proceeded to debulk the tumour mass. The frozen section report of the specimen came as RDD.

The whole mass was completely removed and sent for histopathology and immunohistochemistry for S-100 and CD 68 for confirmation. The bleeding, which was minimal, was controlled by suction cautery and the nasopharynx packed with merocel, used as a conventional postnasal pack to prevent postoperative haemorrhage. The patient had uneventful postoperative period. The postnasal pack was

removed after forty eight hours, there was no bleeding and she was able to breathe normally. She was discharged on the third postoperative day and advised to come for follow up after three months or report immediately in case of remission of symptoms.

The patient reported back to us after three months, for review. She was relieved of nasal block and had no further episodes of epistaxis. There was no evidence of recurrence of the growth on endoscopic evaluation hence no medical treatment was offered. She was asked to come back for review after three months.

Histopathology report

Section revealed the nasopharyngeal subepithelium to be expanded by a hypocellular infiltrate (Fig. 3) of large histiocytic cells with abundant cytoplasm and vesicular nuclei exhibiting phagocytosis of leucocytes including lymphocytes (emperipolesis) (Fig. 4) against a background of plasma cells, lymphocytes and some neutrophils. These histiocytic cells were positive for S-100 protein (Fig. 5) and CD 68, confirming the diagnosis of SHML.

Discussion

SHML also known as RDD is an idiopathic benign histiocytic proliferation affecting the lymph nodes and extranodal sites in all age groups. Although extranodal involvement with lymphadenopathy has been reported in different head and neck sites, namely nasopharynx, ears, orbit, lacrimal gland and major salivary glands, thyroid, larynx, trachea, nose and paranasal sinuses and central nervous system, pure extranodal involvement of the nasopharynx is unusual^{1,3,6,8,10,12,13,14,16}. The disease shows prolonged clinical course with exacerbations and remissions. The disease was first described in 1969 by Rosai J. and Dorfman R.F. in the

lymph nodes, and reporting of similar cases established the disease as a clinicopathological entity.

The characteristic histopathological features of the disease is a proliferation of distinctive histiocytic cells that demonstrate emperipolesis (engulfment of lymphocytes and plasma cells by the histiocytes) against a background of a mixed inflammatory infiltrate, consisting of abundant plasma cells (Russel bodies) and lymphocytes^{9,15,18}. The exact aetiology and pathogenesis of the multicentric disease is unknown although some infections and/or immune mechanisms have been proposed.

Clinically and morphologically SHML should be differentiated from Non Hodgkin's and Hodgkin's lymphomas, infective disorders and Langerhans cell histiocytosis. Hodgkins disease is characterised by the presence of malignant atypical lymphocytic cells but emperipolesis is not seen. Infective disorders like Rhinoscleroma and Histoplasmosis by clinical presentation allow the differential diagnosis. Langerhans cell histiocytosis is characterised by the presence of histiocytes with nuclear grooves associated with prominent eosinophilic infiltrates, though S-100 positive^{15,17}.

Major indications for surgery are life or function threatening obstruction. In this case, frequent epistaxis and nasal obstruction forced us to operate. Most frequently used drug regimen is cytotoxic drugs with corticosteroids though the response rate is poor^{2,7}. In the presence of vital organ compression and/or extranodal localization, as happened in this case, surgical debulking may be necessary. Radiotherapy has limited efficacy, while chemotherapy is in general ineffective as mentioned.

The important points to note are:

- The incidence of nasopharyngeal RDD with extranodal manifestation in the nasopharynx is low, 20% cases exhibit the symptoms and mimics lymphoma and rhinoscleroma.
- By endoscopic sinus surgery the tumour could be removed in entirety which could not have been possible by any other method.
- The frozen section report given by the pathologists helped us to remove the tumour in entirety by endoscopic surgery.
- We did not offer any further treatment as most of these lesions regress spontaneously, but a careful watch is necessary for remission².

Conclusion

We report a case of RDD with extranodal manifestation of the nasopharynx with destructive features, suggestive of nasopharyngeal malignancy and/ or JNA because of episodic epistaxis. The endoscopic approach for obtaining a biopsy for frozen section and complete removal of the tumour was possible in treating this patient.

Fig. 3, 4, 5 Microscopic section revealing the nasopharyngeal subepithelium to be expanded by a hypocellular infiltrate of large histiocytic cells with abundant cytoplasm and vesicular nuclei exhibiting the phagocytosis of leucocytes including lymphocytes (emperipolesis) against a background of plasma cells, lymphocytes and some neutrophils. These histiocytic cells were positive for S-100 protein and CD 68, confirming the diagnosis.

Fig. 3 H&E x10; **Fig. 4** H&Ex40; **Fig. 5** S-100 x40.

In conclusion, clinical observation without treatment is advisable when possible, but life-threatening complications would warrant a surgical intervention⁷.

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