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A rare presentation of Rosai–Dorfman disease involving nose and paranasal sinuses in adult: case report

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Abstract: *A rare case of a 45 years old male, presenting with the history of bilateral nasal obstruction and recurrent episodes of epistaxis for 6 months, was reported. Endoscopic surgery was performed and specimen send for histopathological examination. The report revealed the surprising diagnosis of Rosai-Dorfman Disease. The aim of presenting this case is to highlight the fact that extranodal manifestation of Rosai Dorfman Disease (RDD) without lymphadenopathy and unique clinical features. Pertinent literature is reviewed.*
Keywords: *Case report, nose and paranasal sinuses, Rosai-Dorfman Disease,*

I. Introduction

Rosai-Dorfman disease (RDD) was first reported by Rosai and Dorfman in 1969.[1] It is a rare disorder characterized by sinus histiocytosis with massive lymphadenopathy (SHML) [1]. It is mostly found in children but rare in adults. While nodal involvement is the typical presentation, extranodal manifestation has frequently been described with or without lymphadenopathy.[2] 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 nose and paranasal sinuses is very rare. The goal of this article is to describe previously identified RDD with a rare clinical presentation. Exempt status was granted by the Yangzhou University Health System Instutional Review Board.

II. Case Report

A 45- year old Chinese male presented with a history of bilateral nasal obstruction and recurrent episodes of epistaxis for 6 month duration. He also complains of nasal congestion with non purulent discharge and hyposmia. General examination reveals no any significant cervical lymph nodes enlargement. On anterior rhinoscopy nasal septum mucosa on the left surface was rough, bilateral inferior turbinate visualized and hypertrophy, left nasal cavity – chronic hypermia with mass visualized, nasal septum devation to right. Other examinations were normal.Computer tomography reveled soft tissue lesions in left nasal cavity,ethmoidal sinuses and right maxillary sinus, bilateral inferior turbinate hypertrophy, nasal septum deviated to right side.[Figures 1]. Further investigation, The ESR and plasma globulin levels were high.Routine examination like chest X-ray PA view, ultrasonography of abdomen, renal and liver function test were normal.

Functional endoscopic sinus surgery was performed under general anesthesia. Biopsy was done and sent for frozen section. Surprisingly the report came as RDD. Partial resection of left nasal septum and middle turbinate, bilateral maxillary antrostomy along with clearance of both ethmoid sinuses was done.The mass was sent for histopathological examination. The postoperative period was uneventful. Histopathology of the specimen showed areas of inflamed nasal mucosa with massive infiltration of inflammatory cells predominantly lymphocytes, plasma cells and histiocytes [Figure 2]. Immunohistochemistry reveled S-100(+) and RDD was diagnosed. The patient was found to be symptomatically well on second and sixth month after surgery in follow-up visit. Diagnostic nasal endoscopy was done on follow-up and did not reveal any polyps or residual disease.

III. Discussion

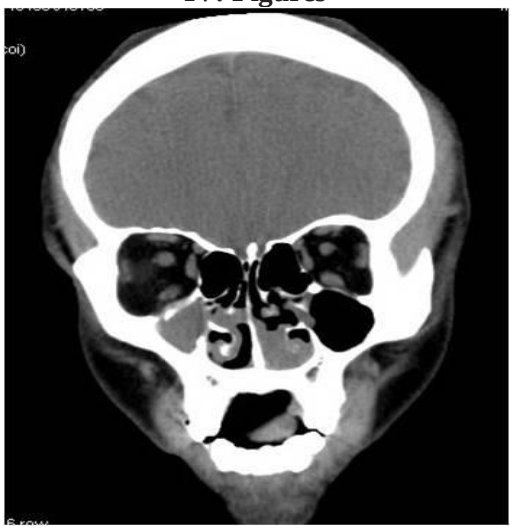
RDD in paranasal sinuses is very rare. Suster et al described a case of SHML involving only soft tissue, based on characteristic S-100 positive histiocytes exhibiting lymphophagocytosis.[3] After that Gregor et al described a case of SHML involving the paranasal sinuses and Leighton and Gallimore reported a case of extranodal SHML affecting the subglottis and trachea without any lymph node involvement.[4, 5]

It is difficult to make correct diagnosis in case of extranodal involvement in which it can only made on the basis of Characteristic histopathological features massive infiltration of inflammatory cells predominantly lymphocytes, plasma cells and histiocytes; confirmatory S -100 test can be done through

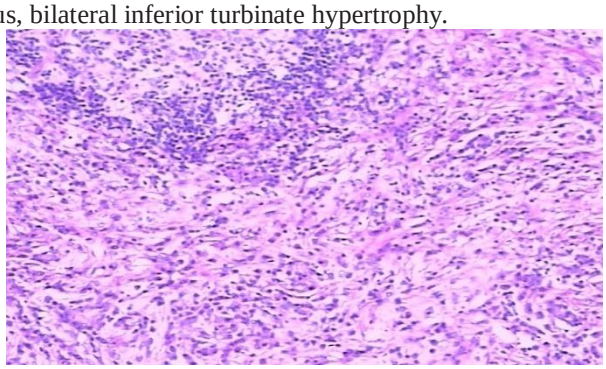
immunohistochemistery. Our case was very rare clinical entity in which there was no any nodal or extra nodal involvement except paranasal sinuses.

According to the previous literatures, different modalities of treatment have been used for the disease including antibiotics, steroids, chemotherapy, antitubercular agent, radiotherapy, surgery. However, none of these modalities has been reported to achieve a sustained response. Surgery can be done for biopsy and relief of airway obstruction or local/regional control of disease. It is self limiting disease, so most of the cases no specific treatment is needed. But long-term follow-up of the patient is beneficial in detecting recurrence and/or any other site involvement and its appropriate management.

IV. Figures



1) Figure 1. Coronal CT-scan of paranasal sinuses showing soft tissue lesions in left nasal cavity, and right maxillary sinus, bilateral inferior turbinate hypertrophy.



2) Figure 2. section showing H&E stain under 100x magnification: massive infiltration of inflammatory cells predominantly lymphocytes, plasma cells and histiocytes

V. Conclusion

Rosai -Dorfman Disese with extranodal manifestation of the paranasal sinuses was diagnosed through the HE report and confirmatory immunohistochemistery (S -100 +) test. No any specific treatment is needed but long term follow-up with close monitoring is required for disease progress.

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