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A rare presentation of Extra Nodal Rosai Dorfman Disease of Nose and Para nasal Sinuses.

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Abstract

Rosai Dorfman disease or Sinus histiocytosis with massive lymphadenopathy is a rare benign disease of unknown etiology that generally manifests as bilateral cervical lymphadenopathy. It is a rare pathologic entity characterized by fever, lymph node enlargement, neutron philia, high ESR and polyclonal hypergammaglobulinemia. The cause of RDD has yet to be established. Usual presenting complains of nasal blockage with intermittent epistaxis.RDD involves the cervical region in about 90% [7,8] of the patient and every organ system can be affected and in 43%

Blood investigation, diagnostic nasal endoscopy, contrast enhanced CT and Histopathology are the modalities of management. Rosai Dorfman disease is benign in nature and complete excision is the treatment of choice.

Keywords: Rosai Dorfmandisease (RDD), Bilateral Cervical Lymphadenopathy.

Introduction

Rosai Dorfman disease or Sinus histiocytosis with massive lymphadenopathy is a rare benign disease of unknown etiology that generally manifests as bilateral cervical lymphadenopathy. The disease was first described by Destombes in 1965 and later described by Rosai and Dorfman in 1969.

It is a rare pathologic entity characterized by fever, lymph node enlargement, neutrophilia, high ESR and polyclonal hypergammaglobulinemia. Sometimes disease involves extra nodal sites such as skin, upper respiratory tract, bone, and retroorbital tissue.

The cause of RDD has yet to be established, but is thought to be an aberrant response to an unspecified

antigen, possibly an infective agent or an aberrant response of macrophage to cytokine.

Etiological hypothesis include immune regulation disorder as well as infections determined by agents such as varicella zoster, and other herpes viruses, Epstein Barr virus, Cytomegalovirus, Brucella and Kleibsella.

Occurrence of Rosai Dorfman disease in first and second decades of life is more common. Rosai Dorfman disease has a 2:1 male-to-female ratio.

Materials and methods

A 60year female patient presented with chief-complain of left nasal blockage for three months with intermittent epistaxis. Patient had no history of allergy, anosmia, headache, blurring of vision. She did not have cervical adenopathy or any evidence of systemic illness.

On diagnostic nasal endoscopythere was bilateral irregular fleshy mass filling entire nasal cavity more on left than right with irregular surface which bleeds on touch. Bilateral choana cannot be seen.

Contrast enhanced CT of paranasal sinuses showed polypoidal mucosal thickening noted in bilateral ethmoidal sinus which was extending in to bilateral nasal cavity and nasopharynx on left side. It showed heterogeneous enhancement on post contrast study. Blockage of left fronto ethmoid recess, left osteomeatal complexes and both spheno-ethmoidal recesses was seen.

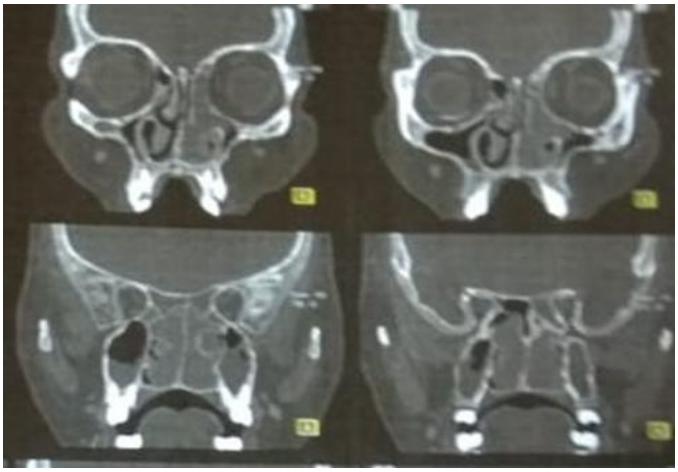

Figure 1:

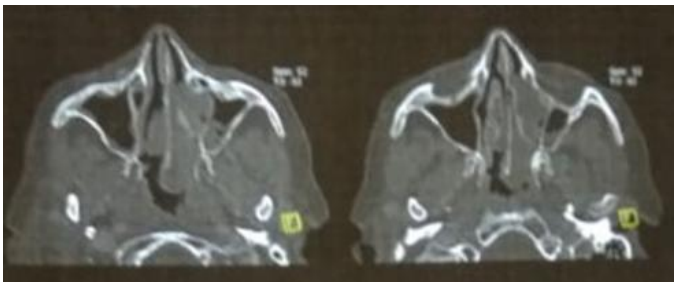

Figure 2:

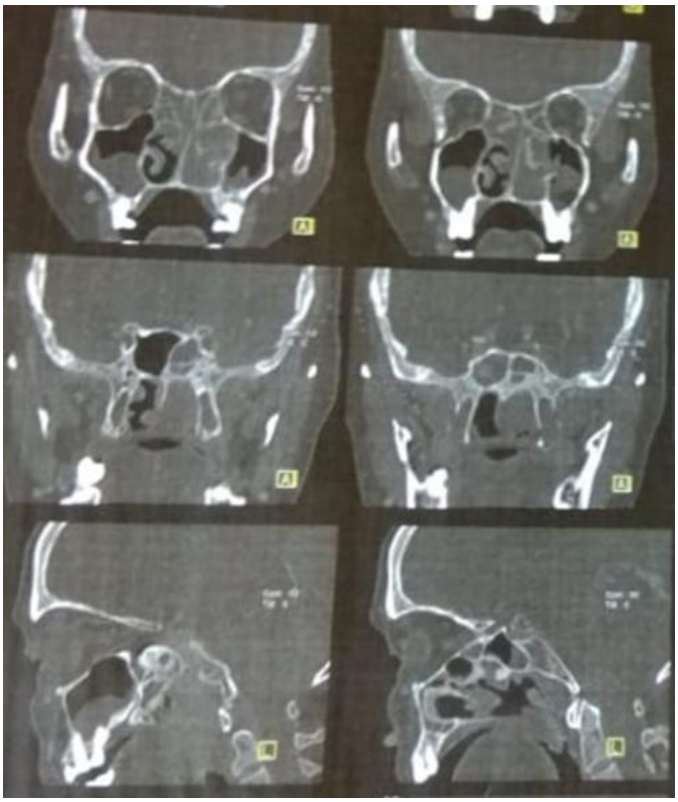

Figure 3:

We did routine blood investigationsand ESR which were within normal limits.

Blood profile- Hb- 11gm%

DLC-88/28/01/01,

PLATELETS- 3.1 lacs/cumm,

RBS -102,

SGPT 22 IU,

S. CREATININE-0.9,

HIV and HBsag nonreactive,

BT1 min30 sec, CT 5 min 45 sec,

PT-14.18, INR-1.01, APTT-29.54,

ESR 48 mm/hr.

Patient underwent Nasal Endoscopy with biopsy, and no unusual bleeding occurred from the lesion during surgery, tissue sent for Histopathological examination to confirm the diagnosis.

Histopathology was suggestive of tissue lined by respiratory epithelium and a lesion composed of sheets of histiocytes admixed with plasma cell, occasional histiocytes show intracytoplasmic lymphocytes and plasma cell, features consistent with sinus histiocytosis with massive lymphadenopathy (Rosai Dorfman disease) On confirmation of benign nature of disease mass was excised en bloc.

Discussion

Rosai Dorfman is the name given to the sinus histiocytosis with massive lymphadenopathy. Most commonly involves cervical neck nodes, but other group of lymph nodes can also be involved.[1]

RDD involves the cervical region in about 90% [7,8] of the patient and every organ system can be affected and in 43%[9] of the cases, the patients have at least one site of extra nodal involvement, and 75% of them occurred within the head and neck region.[9] Within the head and neck, nasal cavity(i.e. NC) (50%) and paranasal sinuses (i.e. PSs) (18.7%) are the most affected sites followed by salivary gland, oral cavity, pharynx and tonsils.[10]

Most common extra-nodal sites involved are skin, soft tissue, upper respiratory tract, bone, eye, retro-orbital tissue. Other clinical feature includes- fever, nasal discharge, weight loss and tonsillitis.

However, in my patient there was no significant lymphadenopathy Laboratory findings in this condition are non-specific. Differential diagnosis is infectious granulomatous diseases, midline destructive diseases, eosinophilic granuloma, Hodgkin diseases, fibroinflammatory diseases.

CT scan with contrast shows paranasal sinuses having polypoidal mucosal thickening noted in bilateral ethmoidal sinus which is extending in to bilateral nasal cavity and nasopharynx on left side. It shows heterogeneous enhancement on post contrast study. Blockage of left fronto ethmoid recess, left osteomeatal complexes and both spheno-ethmoidal recesses is seen.

We did biopsy of mass under nasal endoscopic guidance which shows tissue lined by respiratory epithelium and a lesion composed of sheets of histiocytes admixed with plasma cell, occasional histiocytes show intracytoplasmic lymphocytes and plasma cell, features consistent with sinus histiocytosis with massive lymphadenopathy (Rosai Dorfman disease)

The histiocytes showing emperipolesis (H and E, ×400)

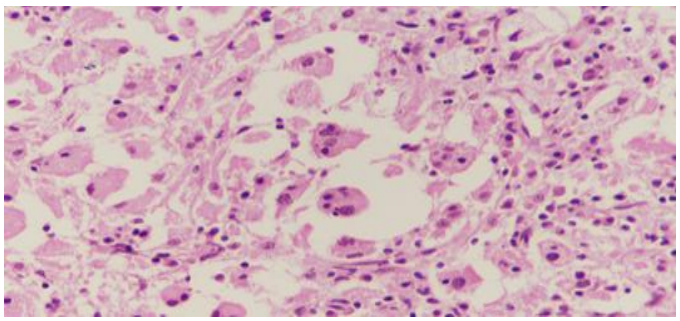

Figure 4:

In RDD, histiocytes are strongly positive for S-100 protein, negative for CD 1a, variably positive for CD 68.

In most of the cases the disease has benign course and self-limiting [2].

No treatment protocol has been decided yet for RDD as it is uncommon. There are various treatment modalities including surgery, chemotherapy, and radiotherapy.

Surgical treatment of resectable lesion gives complete recovery, which can be done [4, 5] endoscopically or externally (cranio facial resection) [6]. Debulking is indicated in lesion causing airway compromise and involving vital organs [3]

Drug therapy consist of corticosteroids, cytotoxic agents,or combination of both.

Beneficial clinical practice would be regular long-term follow-up to detect recurrence and other site involvement and its appropriate management.

On confirming benign nature of mass, we excised the mass en bloc under endoscopic guidance.

Result

With help of histopathology, we confirm the diagnosis of Rosai Dorfman Disease Postoperatively patient recovered well and is on regular follow-up, there are no signs of recurrence.

Conclusion

With reference of other article, the Sino nasal presentation of Rosai Dorfman disease is very rare. Histopathological report is investigation of choice.

Treatment is necessary only in those cases in which lymph node or extra nodal tissue enlargement induce important clinical signs such as upper airway obstruction or another vital organ compression. Rosai Dorfman disease is benign in nature and complete excision is the treatment of choice.

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