

Extranodal Rosai-Dorfman disease involving paranasal sinuses, orbits and anterior cranial fossa

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ABSTRACT

Rosai-Dorfman disease (RDD) is a rare, benign pseudolymphatous condition, predominantly involving lymph nodes. Although several cases of extra-nodal involvement have been reported previously, central nervous system involvement, particularly in the absence of nodal disease is extremely rare. Extranodal large RDD presenting as a single lesion involving sino-orbital and anterior cranial fossa has rarely been described previously. We report a case of incisional biopsy proved RDD in a young lady who presented with nasal obstruction and subsequent proptosis with visual diminution. Radiography of head and paranasal sinus demonstrated a strongly enhanced, diffuse polypoid lesion filling the bilateral sinonasal cavity and orbit with extension to the anterior cranial fossa by way of splaying the bony foramina. Pre-operative low dose steroid therapy had resulted in decreased size of the mass which facilitate gross-total surgical resection. RDD was confirmed by histopathology (emperipolesis) and immuno-histochemistry (S-100 positivity). The follow-up computed tomography 3 months later showed minimal tumor residue in left parasellar region with complete sinonasal decompression.

Key words: Emperipolesis, extranodal, intracranial, Rosai-Dorfman disease, sino-orbital

INTRODUCTION

Sinus histiocytosis with massive lymphadenopathy is a unique benign histiocytic proliferative disorder of unknown etiology, initially described by Azoury and Reed and later described to have a benign course by Rosai and Dorfman in 1969.^[1] Although, it was initially considered to be a lymph node limited disease typically causing bilateral, painless cervical lymphadenopathy, now is recognized as a distinct clinicopathological entity. Isolated extra-nodal form presents in 25% to 40% of cases; most commonly in the skin, head and neck, paranasal sinus (PNS), orbit, upper respiratory tract, bone, testis and rarely central nervous system (CNS).^[2] CNS involvement is uncommon and accounts for less than 5% of cases, with three-fourths in the brain and the other one-fourth in the spine.^[3] Though cases of isolated intracranial Rosai-Dorfman disease (RDD) have been

described in the literature.^[4,5] simultaneous involvement of sinonasal and intracranial cavity is rare with only few cases have been reported in the literature.^[6-10] Three of the reported cases presented as multi-focal involvement of the PNS, intracranial lesion, spinal lesion and skin. Herein, we report a case of RDD having a single lesion involving the sinonasal, orbital and intracranial cavity.

CASE REPORT

The present case report is about a 17-year-old girl presented with 4 year history of persistent nasal obstruction with recent onset of proptosis and visual diminution of 3 month duration. Physical examination showed bilateral proptosis with polypoid subcutaneous mass in the infra-orbital region [Figure 1a]. There was no palpable lymph node or any cutaneous lesion. Computed tomography (CT) scan of the brain and PNS demonstrated contrast enhanced diffuse polypoid hypo to isodense mass filling the nasal, paranasal and orbital cavities extending to anterior intracranial fossa with splaying of osseous foramina [Figures 2 and 3a]. Magnetic resonance imaging (MRI) of the head and neck revealed infiltrative lesions filling the nasal, ethmoid sinus cavities, extending to bilateral orbit and anterior cranial fossa through the osteomeatal complex [Figure 4]. The lesion showed iso to heterogenous signal intensity

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on T1-weighted images, hypo to heterogenous on T2-weighted images and exhibited strong, homogeneous contrast enhancement.

Incisional biopsy from the infra-orbital subcutaneous

mass showed features of extranodal sinoorbital RDD. She was started on oral a systemic steroids (prednisolone 60 mg/day) which was continued for 3 weeks. Within 2 weeks of treatment, the size of proptosis was reduced clinically [Figure 1b] and she showed marked



Figure 1: Clinical photograph of the patient at the time of presentation (a) After 2 week of steroid therapy (b) Showing decrease in the size of the proptosis. Post-operative photo, at the time of discharge (c) At 3 month follow-up (d) shows no proptosis

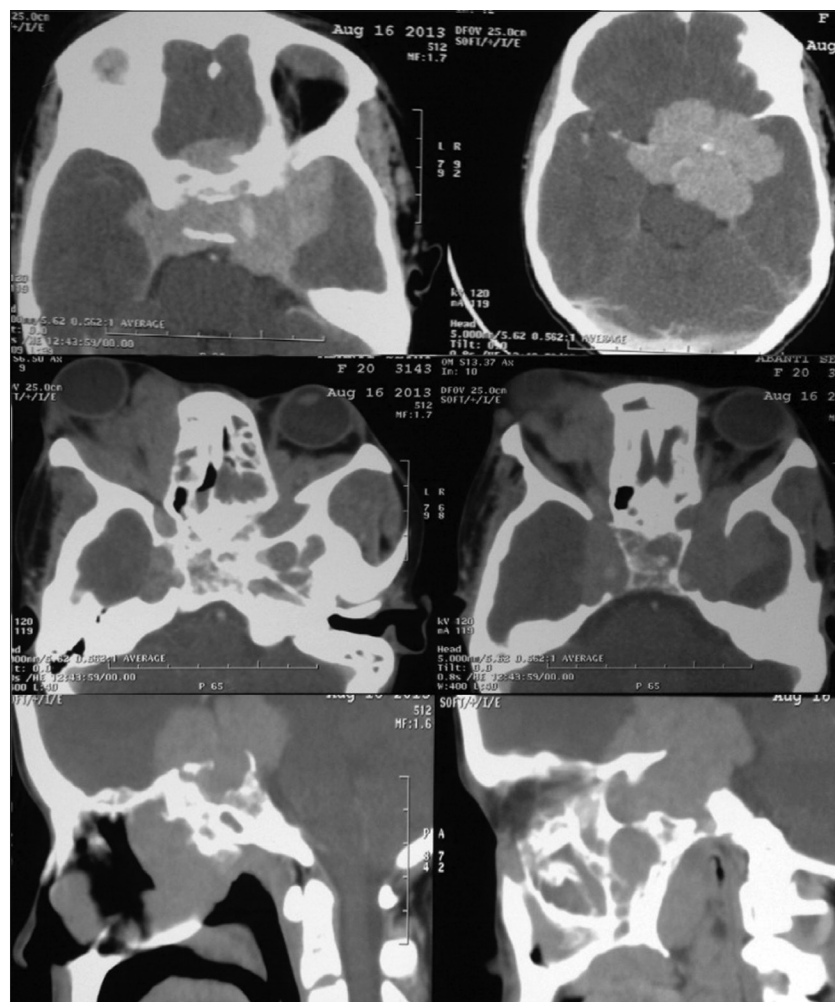


Figure 2: Computed tomography scan of brain and paranasal sinus demonstrates contrast enhanced diffuse polypoid hypo to isodense mass filling the nasal, paranasal and orbital cavities extending to anterior intracranial fossa with splaying of osseous foramina

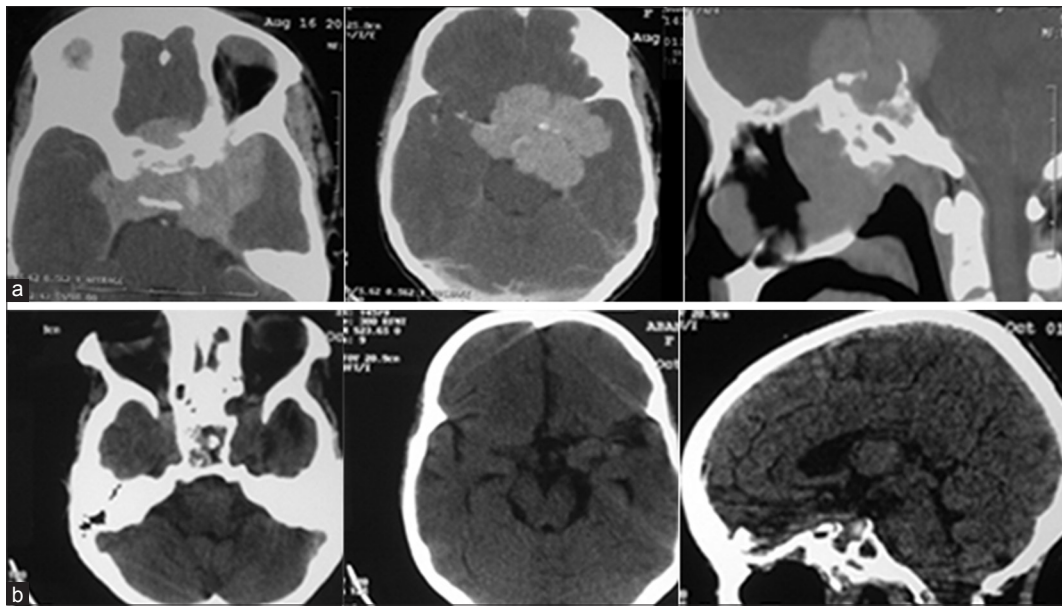


Figure 3: Comparative computed tomography scan at 3 month follow-up showing near complete sino-nasal decompression (b) In comparison to pre-operative tumor status (a)

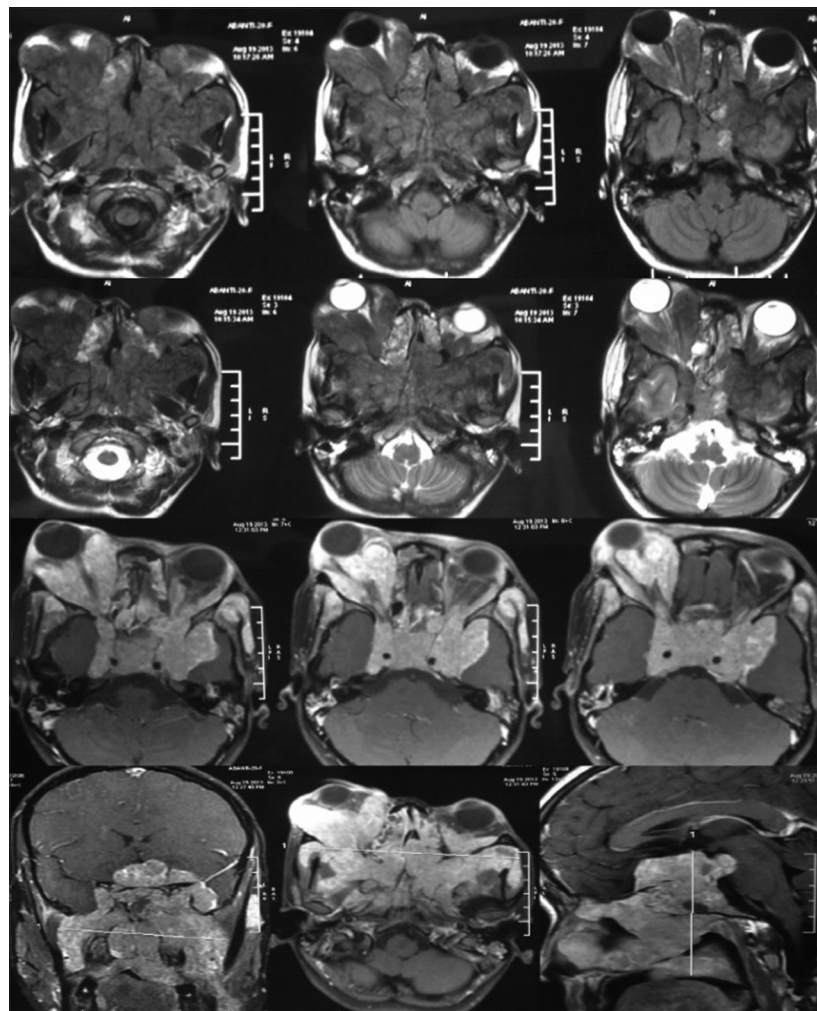


Figure 4: Magnetic resonance imaging of the head and neck shows a T1-weighted iso to heterogenous, T2-weighted hypo to heterogenous intensity infiltrative lesion filling the nasal, ethmoid sinus cavities, extending to bilateral orbit and anterior cranial fossa through the osteometal complex with strong, homogeneous contrast enhancement

improvement in symptoms of nasal stuffiness. This facilitated surgical resection by decreasing tumor size and reducing the amount of intraoperative bleeding. On surgery, right side Weber-Ferguson incision was given with the horizontal arm extending to the opposite side. On opening the skin flap, a firm capsulated avascular tumor was encountered in the bilateral nasal cavity and ethmoid sinus [Figure 5]. The tumor has splayed the infraorbital foramen and extended to right orbital cavity and from there it extended to anterior cranial fossa through the superior orbital foramen. Partial osteotomy of inferior orbital rim was done which improved the exposure. Maximum part of the tumor including the cranial part was excised near totally in a single sitting. A small part of tumor attached to the parasellar region close to the cavernous sinus and chiasm was left in view of possible critical neurovascular injury. Histopathological examination revealed diffuse infiltration of pleomorphic inflammatory cells, including lymphocytes, plasma cells and histiocytes with emperipolesis (lymphophagocytosis) in some affected histiocytes. The histiocytes were found to be immunopositive for S-100 protein, CD68 and negative for CD1a [Figure 6] which confirmed the diagnosis of RDD. The post-operative course was uneventful with decreased nasal obstruction and proptosis [Figure 1c]. She was discharged on oral prednisolone which was tapered-off gradually in next 4 weeks. At 3 months follow-up, she was doing well [Figure 1d] with CT scan showing near complete sino-nasal decompression [Figure 3b].

DISCUSSION

RDD is a rare disease – especially in the Indian subcontinent – and only a few cases have been reported with extranodal involvement as seen in our case with simultaneous sino-nasal-orbital-intracranial fossa involvement. It is a histiocytic lymphoproliferative disorder with unknown etiology and has a distinctive histological appearance characterized by infiltration of lymphoplasmic cells and histiocytes with emperipolesis. The histiocytes in RDD are immunopositive for S-100 protein, CD68 and negative for CD1a.^[11]

RDD usually presents with massive, painless bilateral cervical lymphadenopathy and systemic symptoms of fever and body weight loss. Sinonasal RDD may present as nasal discharge or obstruction, epistaxis, pharyngitis and tonsillitis.^[12] Regarding CNS RDD, the most frequent locations include the cerebral convexities, the parasagittal, suprasellar, cavernous sinuses and the petroclival regions. Cephalalgia, seizure or cranial nerve deficit are the common symptoms of intracranial RDD.^[13]

In our patient with an extranodal large RDD involving bilateral nasal cavities, PNS, orbits and anterior cranial fossa, the clinical presentations were long-standing nasal obstruction, proptosis and subsequent visual diminution.

Sinonasal RDD may present as an infiltrative submucosal lesion or a discrete pseudotumor without bony destruction.^[8] As seen in our cases, the lesion typically appears as soft-tissue masses filling the sinonasal system with mild bony erosion of the contiguous walls on CT. On the other hand, typical intracranial RDD usually shows a well-circumscribed, dura based mass lesion with strong contrast enhancement. Meningioma is the most common differential diagnosis in such cases. Intracranial RDD at various locations mimicking meningioma have been described in the literature.^[13-16] It is slightly hyperintense to the muscle on T1-weighted MR images, with intermediate signals on T2-weighted MR images and strong, homogeneous contrast enhancement on enhanced T1-weighted MR images.^[8,12] MRI is superior to CT in the delineation of the lesion extent and in the discrimination of tumor invasion from the obstructive sinus secretions. All these findings were well demonstrated in the lesion of our case. In our case, it is more likely that the intracranial lesion was due to the extension of same sinonasal mass

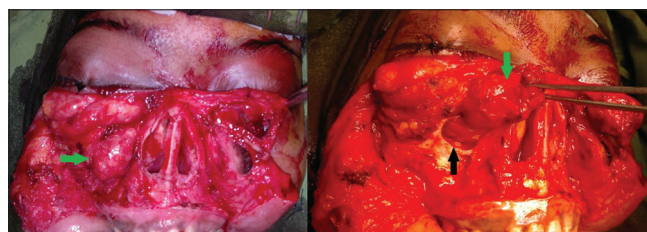


Figure 5: Intra-operative photograph showing the tumor (green arrow) and its extension to orbit through widened infraorbital foramen (black arrow)

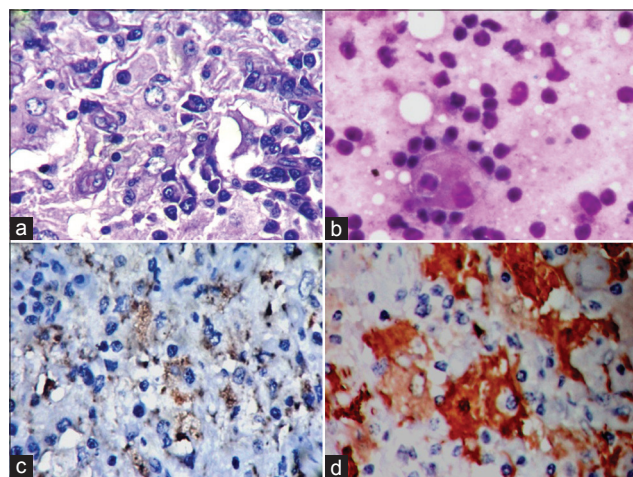


Figure 6: Photomicrograph revealing diffuse infiltration of pleomorphic inflammatory cells, including lymphocytes, plasma cells and histiocytes (a) With emperipolesis (b) In some affected histiocytes. The histiocytes are immunopositive for CD68 (c) and S-100 protein (d)

through the widened infra and supra orbital foramen rather than co-incidental occurrence of different disease entities.

Extranodal involvement, as evident by the presence of sinonasal mass in this case, has to be differentiated from diseases such as orbital pseudotumor, lymphoma, tuberculosis, sarcoidosis, nasopharyngeal carcinoma, nasal carcinoma and olfactory neuroblastoma. Incisional biopsy is the best step in such cases to confirm the diagnosis and hence that optimal treatment could be instituted. The classical finding of emperipolesis (active penetration by a smaller cell into or through a larger cell) differentiates it from other diseases but may not be present in all cases of RDD.^[17]

Specific therapy to RDD is still inconclusive. In the majority of cases, RDD has a benign course and treatment is not necessary. Therapy is required, however, for patients with extranodal RDD having vital organ involvement or those with nodal disease causing life-threatening complications.^[18] Most patients have an indolent course that is characterized by exacerbations and remissions. Extra-nodal RDD tends to be chronic and relapsing^[19] and two thirds of patients with sinonasal RDD have persistent or progressive disease.^[2] Although a variety of treatment modalities had been used, including steroid, radio and chemotherapy, surgical resection appeared to be the most appropriate approach.^[14,20] In our case, oral steroid and surgical resection of the sino-orbital-nasal and intracranial RDD in a single sitting resulted near total decompression leaving only a small part of tumor placed close to the cavernous sinus and chiasm to avoid possible critical neurovascular injury.

In summary, RDD indeed is a rare finding, especially in the Indian subcontinent. The present case of RDD having a single large extranodal lesion involving the sino-naso-orbital system and intracranial area is extremely rare. A high degree of suspicion is needed in such patients. Pre-operative incisional biopsy helps to confirm the diagnosis so as to institute the optimal treatment. Low dose steroid therapy along with radical surgical resection can be used to treat such kind of disease.

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