



Intracranial and Intraorbital Rosai Dorfman Disease

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ABSTRACT

Rosai Dorfman disease is a rare histiocytic disorder. It is also known as Sinus Histiocytosis. It is with massive lymphadenopathy involves an overproduction of a type of white blood cell. The disease is rarely associated with intracranial and intraorbital involvement. Intracranial Rosai-Dorfman can mimic meningioma. Other pathologies also underline its pathologies. Here, we report a nine-year-old boy with a history of proptosis of the right eye and presenting with multiple skull lesions. Histopathological study revealed Sphenopetroclival lesion, which features that of Rosai Dorfman Disease. His MRI scan of the brain was taken, which showed evidence of right optic nerve meningioma with sella and suprasellar extension, causing severe proptosis. The child underwent right frontotemporal craniotomy with petrosectomy and Transylvanian, subtemporal approach to multicompartamental Rosai-Dorfman's lesion. After four months, the patient had a recurrence of the disease on which chemotherapy and steroids were started, which also did not show much response while taking an MRI scan. A corticosteroid is a useful option in the Central Nervous System Rosai Dorfman disease treatment. But this patient showed a negative outcome to the treatment.



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swelling of the lymph node sinuses ([Rodriguez-Galindo et al., 2004](#)). Mostly the cells get accumulated in the lymph nodes, but also occurs in other parts of the body leading to organ damage ([Ali and Mackay, 2009](#)). Juan Rosai and Ronald Dorfman, two pathologists reported RDD as Sinus Histiocytosis with massive lymphadenopathy in young black male, in 1969 ([Mahzoni et al., 2012](#)). Patients with this non-neoplastic disorder, usually present with weight loss, leukocytosis, fever and nonpainful cervical lymphadenopathy ([Gui et al., 2014](#); [Dalia et al., 2014](#)).

INTRODUCTION

Rosai-Dorfman disease (RDD), a rare non-malignant histiocytic disorder characterised painless massive cervical lymphadenopathy, that is due to the overproduction of Non-Langerhans sinus histiocyte, a type of white blood cell which results in accumulation of lymphocytes and histiocytes thereby

Even though lymph nodes in the neck and head are the most preferred area for the disease, RDD can moreover occur in extranodal sites, and the frequent ones are the soft tissue and skin and the central nervous system (CNS) ([Dalia et al., 2014](#)). RDD in other extranodal places like respiratory tract, visceral organs, bone, gastrointestinal tract, genitourinary tract and orbit also reported but is less common ([Hinduja et al., 2009](#); [McPherson et al., 2006](#)).

CNS involvement of RDD is rare and exhibit as dural-based lesion imitating meningiomas ([Krishnamoorthy et al., 2011](#)).

Case Report

History

A 9-year-old boy came with a history of proptosis of the right eye for three years. According to the account, the child also had a squint at the age of 6 years. Gradually noticed that right eye progressively became prominent. For two years, parents didn't do anything, and later a few months ago, the child developed difficulty in closing the right eye completely. He had no history of headache, nausea, loss of consciousness or any other co-morbid illness.

Examination

On admission, the child was conscious, oriented to time, place and person; normal speech and memory. Proptosis causes a restriction to movements of the right eye in all directions. Pupils 2mm and reacting to light on the left side and 4mm reacting to light on the right eye with RAPD (Relative Afferent Pupillary Defect). VA (Visual Acuity) right eye 3/60, left eye 6/9. VF (Vision Field) normal by confrontation.

MR imaging scan of the brain was taken, which revealed evidence of right optic nerve meningioma with sella and suprasellar extension, causing severe proptosis of the right eye. The lesion is hypointense on T1 and homogeneously contrast-enhancing, with another lesion in the right parasagittal high convexity region. Routine post-operative changes with complete excision of the intracranial component of the mass involving the dorsum sella and clivus and a persistent mass in both orbits and paranasal sinuses were observed in the MRI done later. The MR images of axial, coronal and sagittal sections are shown in [Figures 1, 2, 3 and 4](#).

MRI done on the previous day of surgery showed sub acute infarct at right middle cerebral artery territory and an acute infarct at right posterior cerebral artery territory.

Pathological analysis: Histopathology report revealed Spheno petroclival lesion that shows features of Rosai Dorfman Disease.

Operation: On the diagnosis of the disease, the child underwent Right frontotemporal craniotomy with petrosectomy and transsylvian, subtemporal approach to multicompartmental Rosai-Dorfman's lesion (Suprasellar, subtemporal, retroclival) under GA (general anaesthesia).

Operative Findings

1. The tumour was occupying multiple compartments.

2. The tumour was well capsulated with clear margins, greyish firm to soft at places and not much vascular.

3. The tumour was attached to the dorsum, clivus in the midline and right side laterally.

4. The brain was lax at the end of the surgery.

Post-operative: His post-operative MRI showed gross total excision of the multicompartmental lesion. He was hemodynamically stable and moving all four limbs. He was drowsy and had hyponatremia on post-operative day one. He was managed with added salt in diet and fluid restriction and hypertonic saline correction. He became drowsy again, and MRI brain was repeated, which showed the right MCA infarct. He underwent emergency decompressive craniectomy under GA on post-operative day four. Post-operatively he was kept on ventilatory support and extubated on post-operative day one. He had left hemiplegia and left UMN type facial palsy following surgery. Due to chemosis of the conjunctiva, his right eye could not be evaluated. The patient was discharged with Dexamethasone 4mg twice daily for one week, which then tapered to a once-daily dose for one week.

After four months of surgery, the child was again admitted with the recurrent disease for which a Hematology consultation was made. The child received chemotherapy with CHOP regimen + Vinblastine + Prednisolone X 6 cycles. He had an optimal sub response to Prednisolone and Vinblastine. On clinical examination, right eye proptosis caused restriction of the movements in all directions. Vision: left eye- 6/18; right eye- complete proptosis and could not examine.

MR imaging report showed enhanced intraorbital masses in both left and right side, and the size of the lesion slightly increased than the previous scan.

Discussion

RDD is a benign disease characterised by overproduction and accumulation of histiocyte in the lymph nodes, mostly those of the neck (cervical). Abnormal accumulation occurs in other areas of the body (extranodal). In many cases, signs and symptoms go away without treatment but in some cases, various treatment options required.

A well-established treatment protocol is not yet available for the treatment of Rosai Dorfman disease. Conventional therapies given for the condition are surgery, steroids, chemotherapy and radiation. Treatments have improved symptoms in some patients, but in others, it's not been effective ([Pinto et al., 2008](#)).

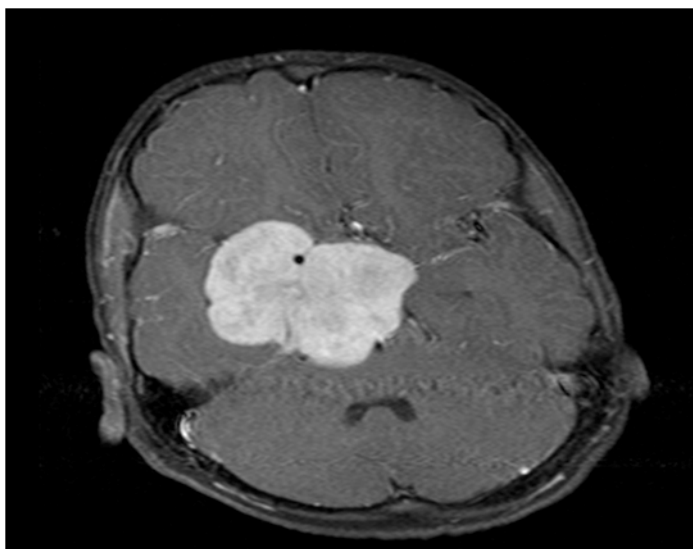


Figure 1: MRI contrast axial section shows suprasellar meningioma extending to right parasellar region abutting medial temporal lobe posteriorly and caudally extending into pre pontine cistern abutting mid brain and pons anteriorly encasing right cavernous internal carotid artery.

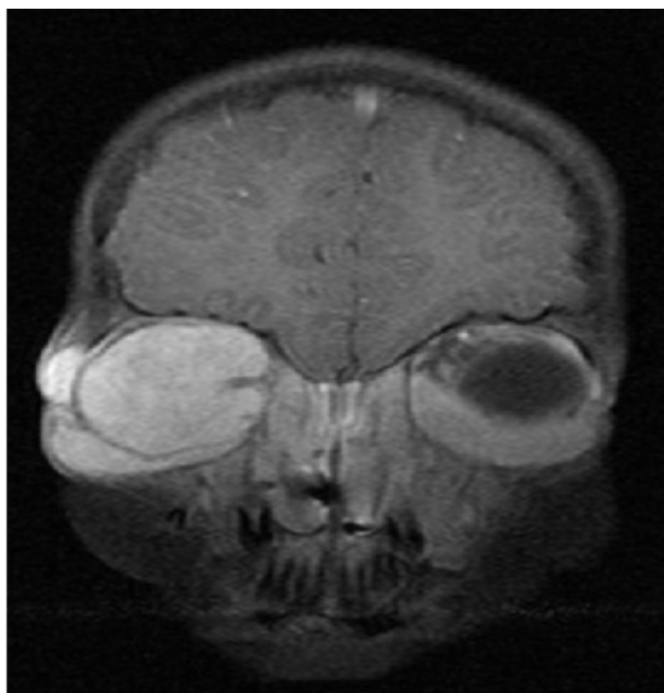


Figure 2: MRI contrast axial section show right optic nerve meningioma with sella and suprasellar extension causing severe proptosis of the right eye.

As seen in the present case, RDD occurred in the extranodal site, causing intracranial multicompartamental and orbital lesions. CNS involvement usually resembles that of meningiomas on MR imaging. Many instances of extranodal have been reported early, central nervous system involvement without nodal disease is particularly unusual (Pinto *et al.*, 2008). Emperipolesis exhibited by RDD cells is the nondestructive phagocytosis of erythrocytes or lymphocytes. Emperipolesis is the characteristics of the

disease and is necessary for the diagnosis. RDD has an unknown aetiology and is regarded as an idiopathic histiocytosis (Dalia *et al.*, 2014; Kattner *et al.*, 2000). RDD involving CNS is uncommon, and mostly it occurs with dura-based, extra-axial involvement of the cranium. In contrast, rare cases of involvement of spinal cord and intracerebral areas are noted.

Neurological symptoms are typically based on the site of the lesion and usually reported ones are

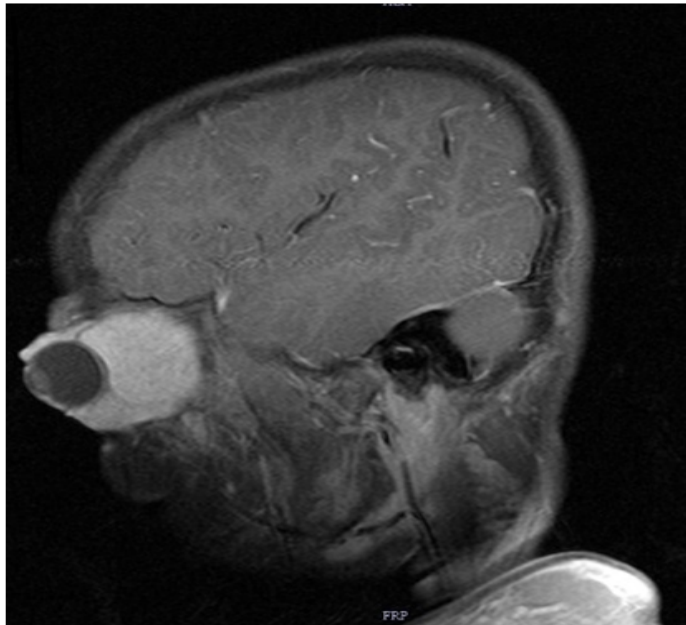


Figure 3: MRI contrast coronal section show right optic nerve meningioma with sella and suprasellar extension causing severe proptosis of the right eye.



Figure 4: MRI contrast sagittal section show right optic nerve meningioma with sella and suprasellar extension causing severe proptosis of the right eye.

headache and seizures. In contrast, constitutional symptoms are generally absent (Dalia *et al.*, 2014). Since RDD is a non-malignant condition, treatment is instructed if they are symptomatic or their vital system or organ is involved (CNS).

In those patients who require treatment, surgery is an apt choice for the disease that can be ablated, consisting of single nodal areas or primary CNS involvement. In CNS-only condition, many cases have been reported of subsidence with surgery alone. For patients requiring systemic treatment, steroids are the drug of choice as that gives responses in classical RDD as well as in extranodal disease even though, the dependability and longevity of these responses are unreliable. For symptomatic disease, radiation is a palliative option.

Chemotherapy can be used for patients with disseminated RDD or refractory to surgery or other modalities (e.g., radiotherapy, steroids).

Using radiotherapy, we can conserve vital organs and system functions like in case of the orbital, airway, and CNS involvement (Dalia *et al.*, 2014). Recurrence after surgery is very rare. Still, this patient had a recurrent episode which was treated with steroids and chemotherapy.

CONCLUSIONS

Rosai Dorfman disease is a rare condition of which its exact number of cases reported is not known. No more extensive studies have been conducted for the treatment options yet for the disease. However, this is not a life-threatening disease. In many cases, without any treatment, signs and symptoms of Rosai Dorfman disease go away within months or a few years. But some individuals may need surgical removal of histiocytic lesions. In more severe cases, other treatment options like therapy with some drugs like steroids, chemotherapy or even radiation therapy are considered. These treatments have improved the symptoms in some individuals, but in some, they don't show much effect. Here, for the above-discussed patient, the existing therapies showed a negative outcome.

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Conflict of Interest

The authors declare that they have no conflict of interest for this study.

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