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Case Report

# Isolated intracranial Rosai Dorfman disease without nodal involvement

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## Abstract

**Background:** Rosai Dorfman disease (RDD) is a benign lymphohistiocytosis that often involves lymph nodes and present as massive painless lymphadenopathy with sinus histiocytosis. Usually systemic involvement and with rare intracranial and extremely rarely intracranial involvement without the involvement of lymphadenopathy.

**Case Presentation:** We presented a case of 60 years old female with seizures and left side weakness and no lymphadenopathy. The magnetic resonance imaging (MRI) revealed contrasting right frontal homogenously enhancing convexity mass.

**Management & Results:** The patient was kept on antiepileptic medications but soon presented with fits and slight expansion in frontal mass. Surgery was performed (right frontal craniotomy), the mass was surgically resected and biopsy indicated RDD.

**Conclusion:** By now only seven of such cases are reported and prognosis of the disease is not poor if surgically treated however other measures including radiotherapy, chemotherapy, steroids are not very effective for treating the disease. And due to the rarity of disease suspicion of such disease should always be kept as a key differential in homogenously enhancing lesions with dural involvement with or without lymphadenopathy.

## Keywords

Rosai Dorfman, Extranodal, Intracranial

## Introduction

RDD also called sinus histiocytosis is one of the rare benign proliferative pathological condition characterized by massive, painless, bilateral, cervical lymphadenopathy associated with fever, leukocytosis, elevated erythrocyte sedimentation rate (ESR), and weight loss massive lymadenopathy[1]. Most of the cases with intracranial involvement include nodal involvement along with extra nodal component of disease including orbit, head and neck region, upper respiratory tract, skin, bone and testis. With prevalence of 43%, isolated intra cranial involvement without nodal involvement is extremely rare[1-3] and by now only 7 cases had been reported[4,5]. The disease usually affects young adults and can have a protracted course lasting from several months to years and commonly results in complete recovery[6]. Central Nervous System (CNS) RDD typically presents during the 4th to 5th decade and is more prevalent among males with the mean age of 39.4 years old. In this study we have presented a case with an extremely rare presentation of intracranial RDD without nodal involvement in a female with 60 years of age.

## Case Presentation

A 60 years old unmarried lady presented to walk-in clinic in February 2017, she previously had hypertensive history, current observation at the time of presentation at the clinic included focal fits with secondary generalization and left side weakness for one day. According to patient and attendant she had developed fits which were focal initially and then transformed into secondary generalization lasting, five mins without aura. She had no previous history of fits, nor any significant family history.

## Management & Results

Patient was rushed to the hospital and got admitted to accident and emergency (A&E), where she was initially treated symptomatically and taken to the walk-in clinic on next day with weakness of left side of body, power was 0/5 in left side. In addition to hypertension, she also had past history of allergic rhinitis therefore, systemic enquiry, clinical and systemic examination was insignificant. Her initial MRI brain scan showed homogenously enhancing right frontal convexity lesion, considered temporarily as convexity meningioma, patient was kept on antiepileptics and follow-up was recommended. But two months later in April-2017, the patient was readmitted due to fits and there was slight expansion in frontal mass. However, a surgical procedure was planned, taking all ethical measures and attaining consent from the family of the patient. Right frontal craniotomy was carried out with excision of lesion completely and irregular intact nodular tissue covered with frontal tissue piece specimen was sent for histopathology. Histologically, lesion revealed dense fibrocollagenous tissue with inflammatory infiltrates, scattered lymphocytes, plasma cells along with histiocytes and extensive areas of emperipolesis. Specimen with cluster of differentiation-68 (CD-68) highlighted histiocytes and immunohistochemical stain S-100 was positive while CD-1a was negative hence indicating "Intracranial RDD".

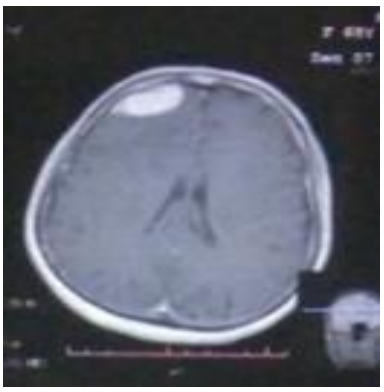

Figure 1: First Scan

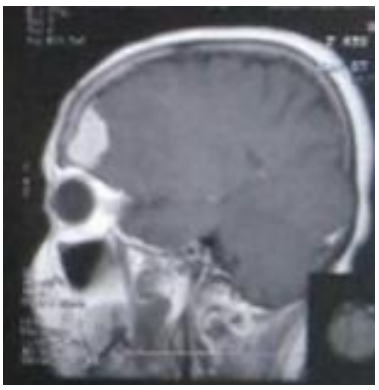

Figure 2: First Sagittal Section Scan

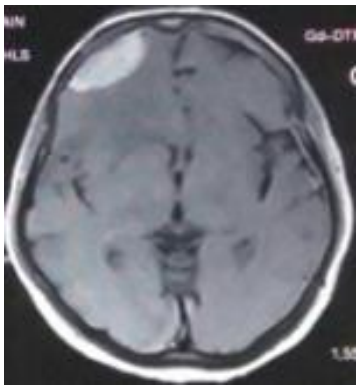

Figure 3: Repeat Scan after 2 months

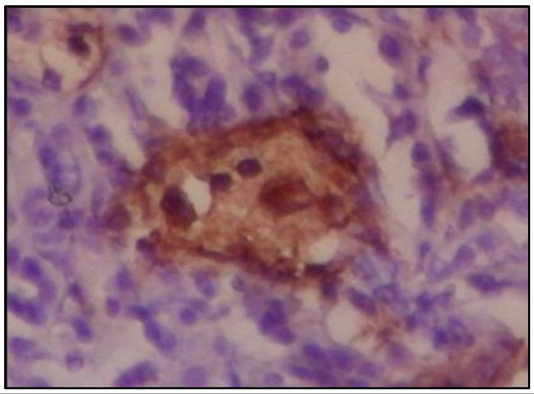

Figure 3: Histiocytic cells with emperipolesis are immunoreactive for S-100 protein and negative for epithelial membrane antigen (EMA)

## Discussion

RDD is a histioproliferative disorder that mainly involves cervical lymphadenopathy[7], the disease was first described in 1965[1], predominantly affecting children and young adults and characterizing massive painless bilateral cervical lymphadenopathy with fever, raised white blood count (WBC) count, high ESR, and polyclonal hypergammaglobulinemia[8].

Microscopically, lymph nodes show dilated sinuses containing foamy histiocytes and plasma cells. Many of these histiocytes contain intact hematopoietic cells, mostly lymphocytes within their cytoplasm, a phenomenon known as emperipolesis[2,3,9]. These histiocytes are positive for S-100 protein and CD-68 and negative for CD1a based on immunohistochemical examination[2,7,10]. Literature describes there may be association of episten bar virus or herpes simplex virus (HSP)-VI associated with this condition[2].

Our case was initially radiologically diagnosed as meningioma based on pathology but histology revealed RDD. Histologically, Langerhans cell histiocytosis is similar to RDD occurrence except the fact that Langerhans cells have folded nucleus with longitudinal grooves which is absent in RDD histiocytes and Langerhans histiocytes are also positive for CD-1a[11], on contrary RDD is negative for CD-1a[11]. In our case patient had a convexity lesion over right frontal region causing seizures and left sided weakness while there wasn't any

systemic or nodal involvement, which made the case extremely rare. As most of the cases usually involve some kind of systemic or nodal involvement. Most of the patients with disease underwent total or subtotal surgical excision. However, radiochemotherapy and steroids role in treatment is still doubtful.

## Conclusion

Isolated intracranial RDD without nodal involvement is an exceptional rare disease and the diagnosis for the lesion is still challenging. The histological displays and immunohistochemical analysis are still the only reliable basis for diagnosis of the disease. Surgical resection demonstrated an effective treatment while other treatment strategies are still controversial for managing the disease.

## Conflicts of Interest

None.

## Acknowledgement

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## Funding

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