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Research Article

Isolated Intracranial Rosai-Dorfman Disease: A Diagnostic Challenge

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Abstract

Rosai-Dorfman disease (RDD) is a benign condition characterized by abnormal proliferation of white blood cells called histiocytes and an uncommon biological process called emperipolesis, in which a living cell penetrates inside another living cell. Central nervous system involvement in RDD is extremely rare. We report one such rare case of 47-year- old man who presented with right sided focal seizure from 1 year. MRI Brain showed a dural based lesion. On the basis of clinical presentation and radiological findings initial diagnosis made was of en plaque meningioma. Gross total resection of lesion was performed. Initial histopathological diagnosis was of IgG4 related HPM. Post surgery there was significant improvement in patients clinical condition. Intensity of seizure episodes were reduced. However due to persistence of right sided focal seizures, tissue blocks were reviewed and diagnosis of intracranial Rosai-Dorfman disease was made. Upon which further hematologist opinion was taken and whole body PET CT was done which showed solitary uptake in brain. It was observed that characteristic feature of RDD emperipolesis is sometimes masked by storiform fibrosis and lymphocytic infiltration leading to misdiagnosis. Therefore close follow up is required, in case of persistence of symptoms tissue blocks should be reviewed and possibility of intracranial Rosai-Dorfman disease should be considered while dealing with dural based lesions.

Keywords

Histiocytes, En-plaque, Intracranial, Emperipolesis, Storiform, Infiltration

1. Introduction

Rosai-Dorfman-Destombes disease (RDD) is a rare histiocytic disorder described by Destombes in 1965 and later by Rosai and Dorfman in 1969 as ‘sinus histiocytosis with massive lymphadenopathy’. Classical RDD present with massive bilateral painless cervical lymphadenopathy associated with fever, loss of weight and night sweat. [1] It predominantly affects children and younger adult. [3, 13] Extranodal disease is seen in over 40% of cases and may rarely occur in the absence of nodal disease. CNS can be involved less than 5% of Rosai-dorfman Disease. [6, 9] Common extranodal sites of involvement include the skin (10%), nasal cavity (11%), bone (5%–10%), orbital tissue (11%) central nervous system (5%, predominantly dural): mostly involving the two sides of the dura and mimicking meningioma. [1, 4, 10, 11, 12]

2. Case Summary

A 47 year male presented in Department of Neurosurgery with chief complaints of right sided focal seizure involving

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face and upper limb associated with tingling sensation from 1 year. There was 5 to 6 episodes of focal seizure per day lasting for 40-50 secs was present. Patient consulted many neurologist who started him on multiple antiepileptic drugs but there was no relief. There was no history of trauma, incontinence, altered speech, memory alteration, visual impairment, vertigo and childhood seizure. On examination right hand grip weakness was present. Rest of the examination was unremarkable. Further radiological investigations were done.

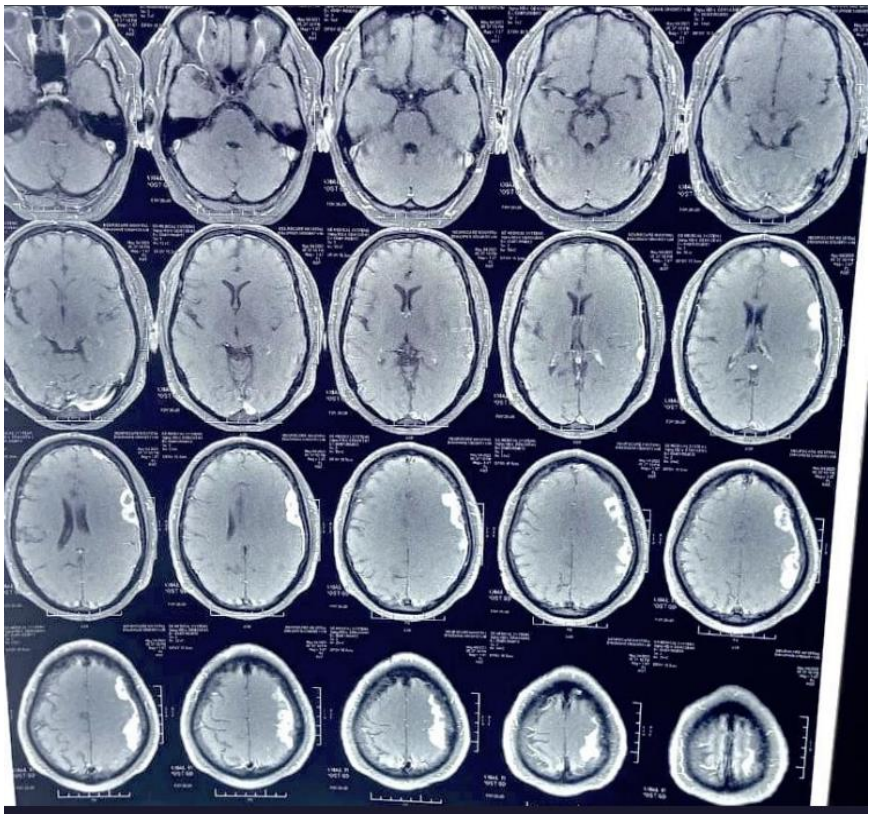

Figure 1. *MRI Brain T1 weighted Post Contrast Image.*

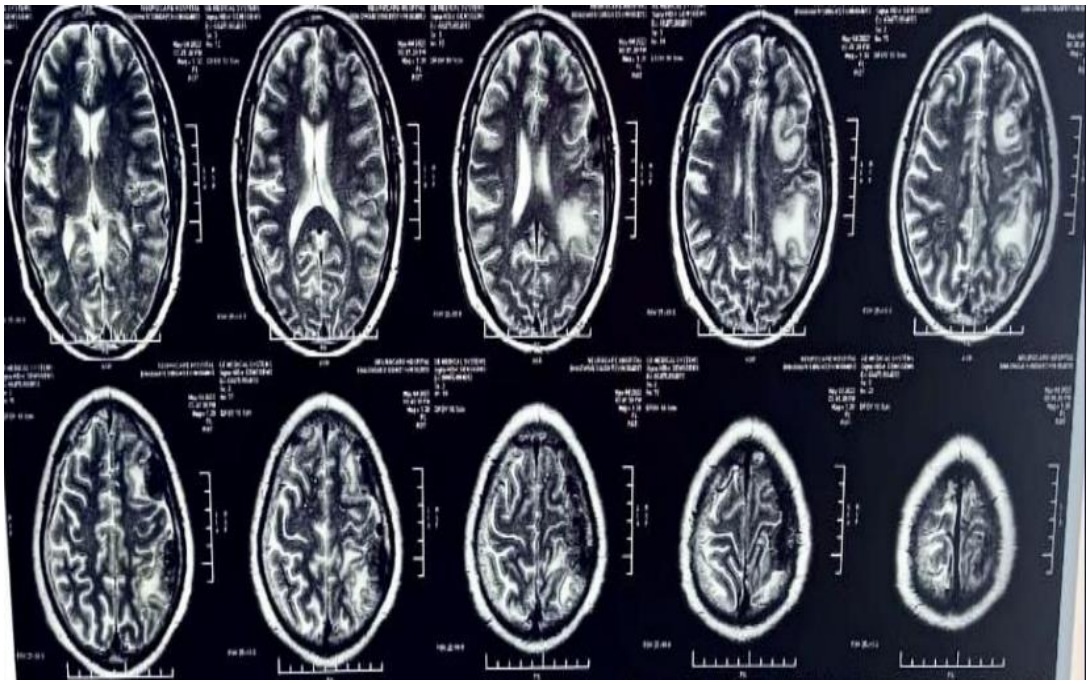

Figure 2. *MRI Brain T2 weighted images showing hypointense dural covexity lesion at left frontal and parietal region with peri lesional edema.*

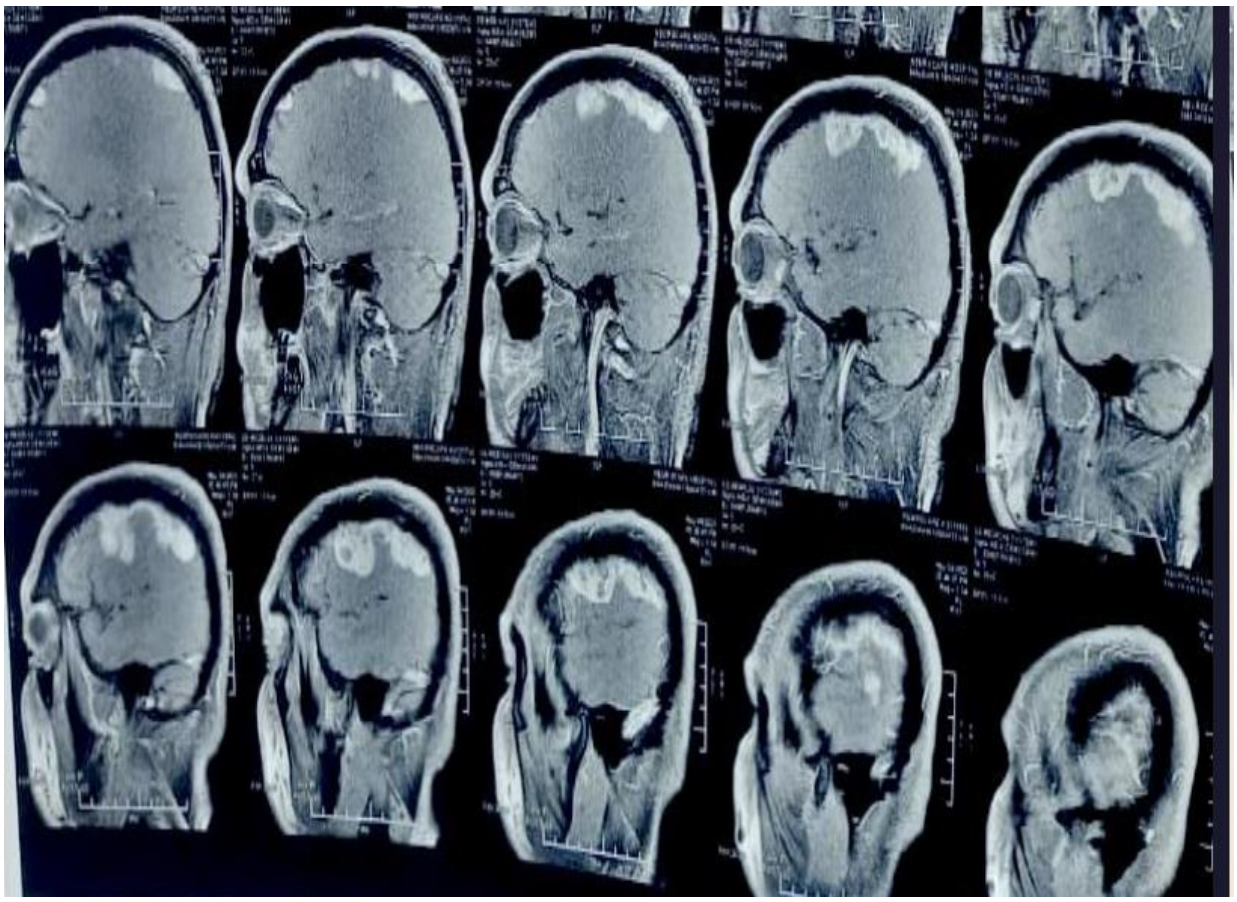

Figure 3. *MRI Brain Sagital Section.*

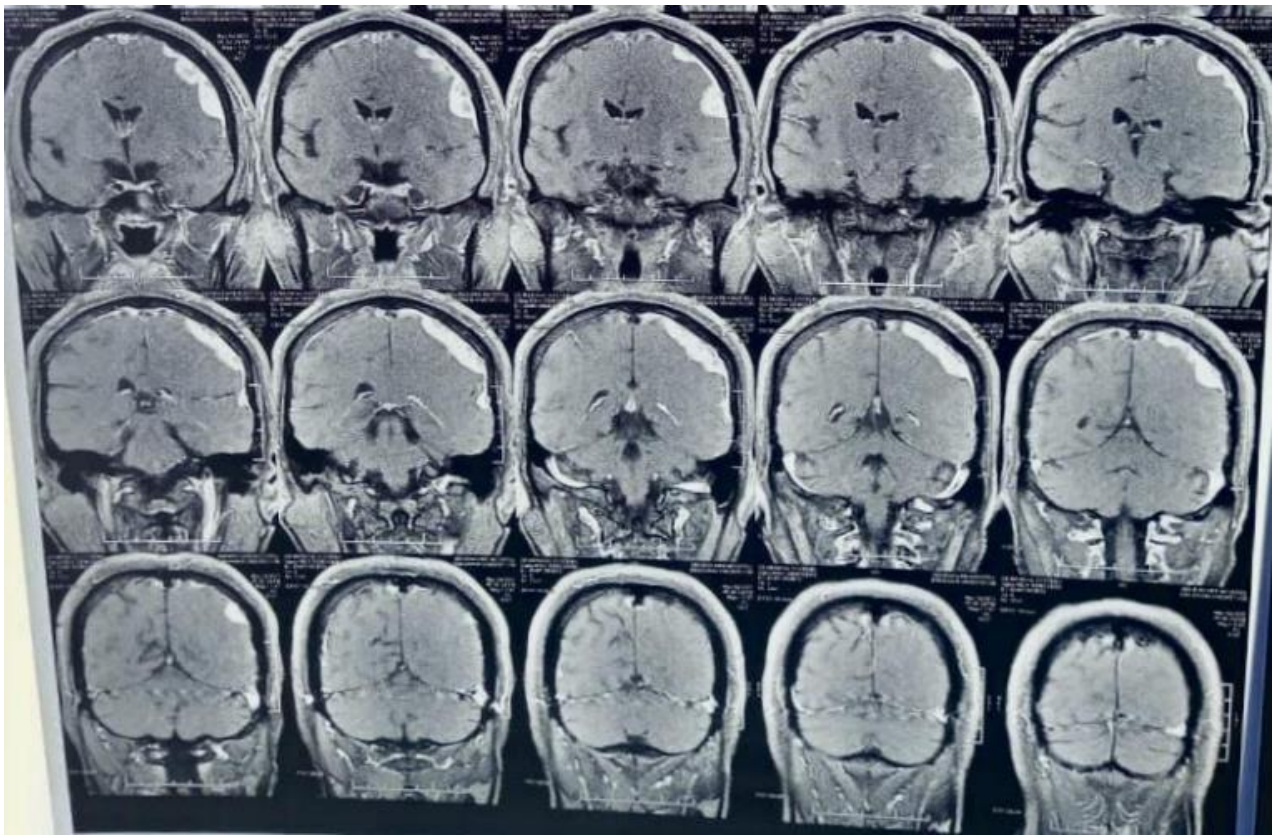

Figure 4. *MRI Brain Coronal Section.*

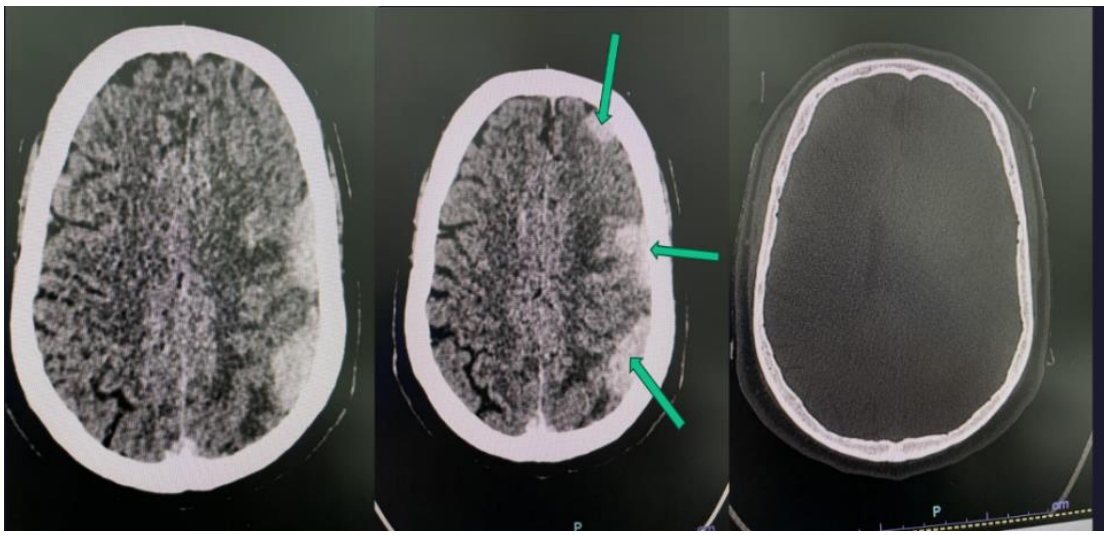

Figure 5. CT Brain showing multiple dural based extra-axial enhancing lobulated lesion along left frontoparietal convexity without bony erosions and hyperostosis.

2.1. Clinical and Radiological Diagnosis

MRI Brain done showed hypointense dural convexity lesion at left frontal and parietal region with perilesional edema. CT Brain showed multiple dural based extra axial enhancing lobulated lesion along left frontoparietal convexity without bony erosions and hyperostosis. Clinical and radiological diagnosis of en-plaque meningioma of left frontoparietal convexity compressing the underlying parenchyma was made.

2.2. Surgical Treatment

Patient was planned for craniotomy and surgical excision of lesion. Intraoperatively after removing the calvarium at left fronto -parietal aspect. Dura was visible with some nodular lesions underneath it. On opening the dura we found three lesions lying over cortical surface of parietal and frontal lobes, yellowish nodular oval shaped well defined lesions attached with dura with no clear demarcation/plane of dissection between lesion and brain tissue. Suspecting some high grade lesion en mass excision was done except the part of nodule present over middle frontal gyrus which was in proximity of pial vessel to avoid pial violation. Tissue sample was sent for histopathological examination.

2.3. Post Operative Period

On post operative day 3, patient had an episode of focal seizure in right hand. CT brain was done which showed patchy edema in the left parietal lobe, involving the postcentral gyrus and to a lesser extent the left frontal lobes with few foci of hemorrhage. No increase in mass-effect since preoperative study. No significant midline shift or brainstem herniations.

2.4. Histopathological Examination

Histopathological examination shows histiocytic cells, admixed with plasma cells, lymphoid cells, emperipoletic activity, CD68 positivity suggestive of IgG4 related hypertrophic pachymeningitis. Tissue blocks were reviewed which showed excessive proliferation histiocytes admixed with plasma and lymphoid cells. Emperipoletic activity was seen along with positivity for S-100 and CD68 proteins. Diagnosis of Rosai Dorfman was made.

2.5. Follow Up Period

Follow up period after 1 month was uneventful with only 3 episodes of focal seizures and improvement in right hand grip. Patient antiepileptic medication and low dose steroid was continued. Whole body PET CT was negative except Solitary FDG uptake in left frontal lobe area. Antiepileptic medication was continued and steroids were gradually tapered and stop. Patient was advised to follow up after 3 months with repeat MRI Brain.

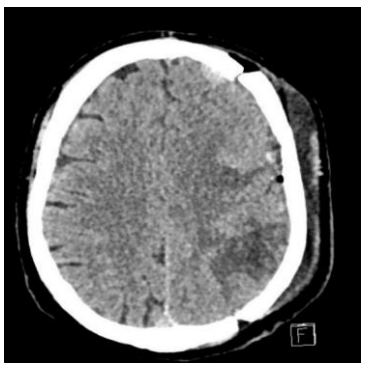

Figure 6. Post operative CT Brain.

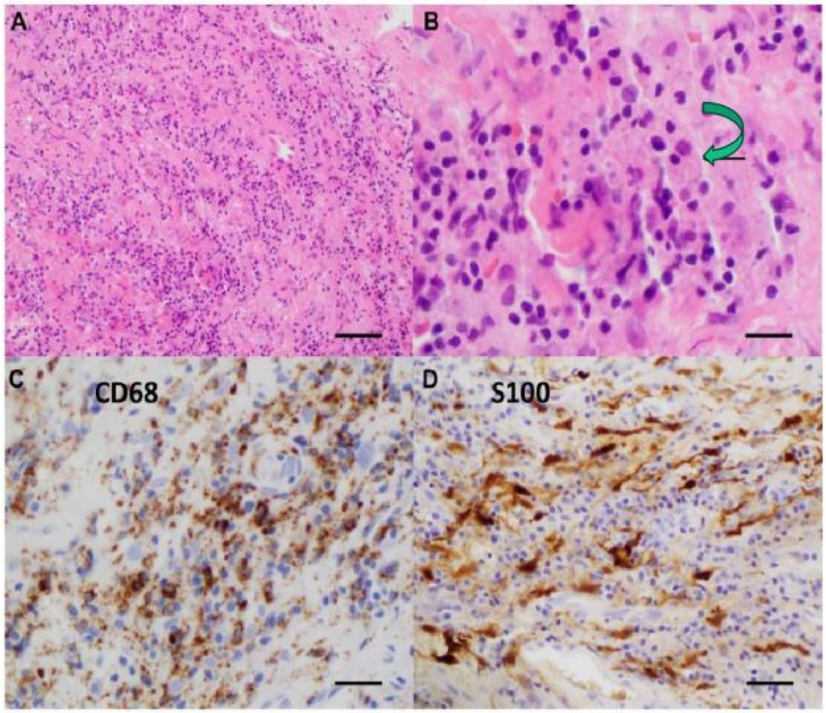

Figure 7. a) histiocytic cells, admixed with plasma cells and lymphoid cells. b) emperipoletic activity c) CD68 positivity d) S100 positivity.

3. Discussion

The definitive diagnosis of Rosai-Dorfman disease is based on histopathological examination and immunohistochemistry. [2] Histologically, Rosai-Dorfman disease is characterized by an infiltration of lymphoid plasma cells and histiocytes. Over 70% of the case, the histiocytes show emperipolesis that is characteristic of Rosai-Dorfman disease. Fibrosis in the background can be seen in extranodal involvement of disease. On immunohistochemical examination, histiocytes are immunoreactive for S-100 protein, HAM 56, α1 antitrypsin, α1 chymotrypsin, lysozyme, Mac 387 and Ki-1, but are negative for CD 1a and EMA. [5-8]. Due to masking of characteristic feature emperipolesis by storiform fibrosis, lymphocytic infilteration and rarity of CNS involvement in RDD isolated intracranial rosai dorfman disease is often misdiagnosed with meningioma and other en plaque dural lesions. Resection of lesion is the most effective treatment for isolated intracranial RDD followed by implementation of adjuvant therapies together with localized radiotherapy if neurologic symptoms persist. [4] Recurrence after surgical therapy is rare and limited to uncompleted debulking, multi-organ involvement and large mass. [11, 14]

4. Conclusion

Intracranial variety of RDD is very rare (5%) [6, 7], although rare it may lead to serious complications like blindness and deafness. So early and accurate diagnosis is very important. Isolated intracranial RDD as shown in the present case, may pose a diagnostic challenge both for the clinician, radiologist and the pathologist as radiological features mimics like en plaque meningioma and key characteristic feature emperipolesis was masked by fibrosis and lymphocytic infiltration leading to radiological diagnosis of en plaque meningioma and Histopathological diagnosis of IgG4 related HPM. Therefore uncommon presentation of isolated intracranial RDD should be considered as differential diagnosis of dural based lesions.

Abbreviations

A1 Antitrypsin	Alpha 1 Antitrypsin
A1 Chymotrypsin	Alpha 1 Chymotrypsin
CD1a	Cluster of Differentiation 1a
CD68	Cluster of Differentiation 68
CNS	Central Nervous System
EMA	Epithelial Membrane Antigen
HAM56	Human Alveolar Macrophage 56
HPM	Hypertrophic Pachymeningitis
IgG4	Immunoglobin G4
RDD	Rosai Dorfman Disease

Conflicts of Interest

The authors declare no conflicts of interest.

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