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Case Report

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Isolated Intracranial Rosai-Dorfman Disease: A Diagnostic Challenge

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Abstract

Isolated intracranial Rosai-Dorfman disease (RDD) is an extremely rare, idiopathic histo-proliferative disorder. RDD is associated with the proliferation of histiocytes and emperipolesis. Central nervous system (CNS) involvement is extremely rare and due to rarity of RDD, it is not usually proposed in intracranial lesions diagnosis. RDD radiologically mimics meningioma and dural metastasis as dural-based lesions and histologically mimics plasma cell granuloma, Langerhans cell histiocytosis (LCH) and lymphoproliferative disease. We report a case with isolated intracranial RDD. A 47-year- old man presented with focal seizure. This case preoperatively was misdiagnosed with meningioma. Histopathological examination revealed pale histiocytes displaying emperipolesis which were positive for S-100 and CD68 proteins and negative for CD1a marker. BRAF V600E mutation was negative. In this case, total resection was performed and clinical symptoms were regressed completely.

1. Case Summary

A 47 year male presented in Department of Neurosurgery with chief complaints of focal seizure involving right side of face and right upper limb and tingling sensation in right upper limb from 1 year. There were 5 to 6 episodes of focal seizure per day. Each episode used to last for 40-50 secs. 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.

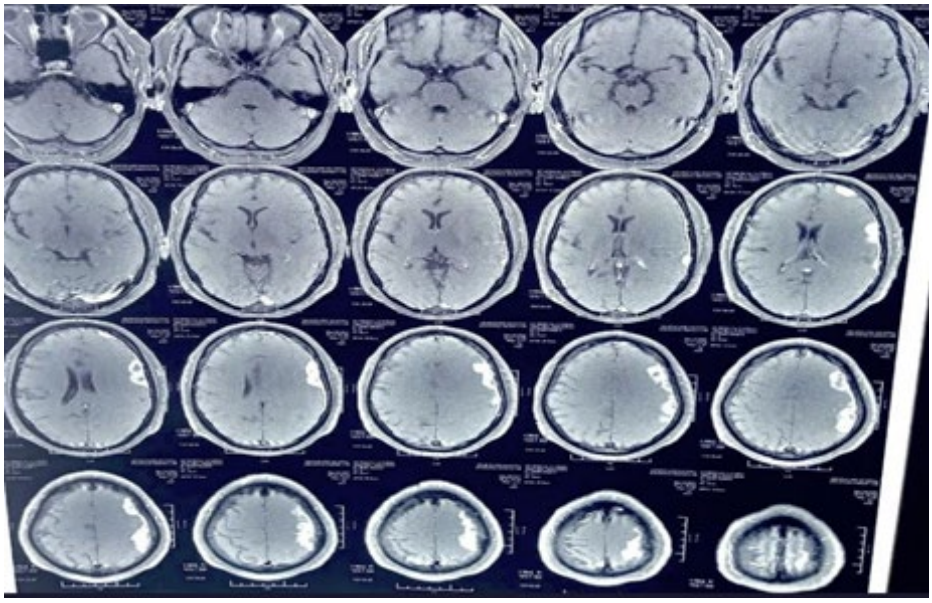

Figure 1: MRI Brain T1 Weighted Post Contrast Image

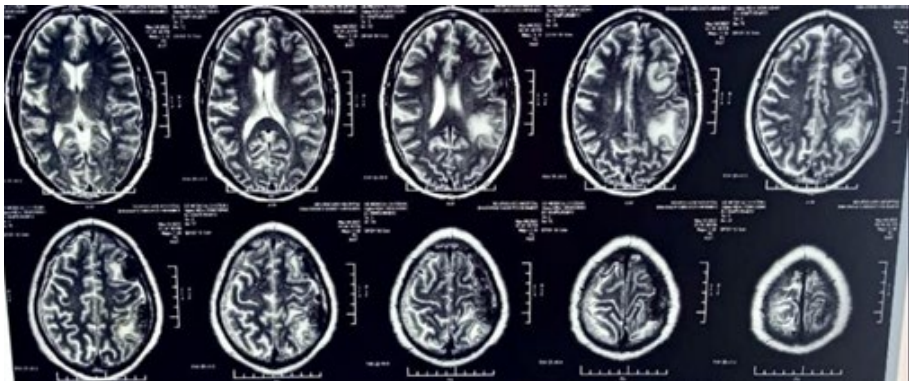

Figure 2: MRI Brain T2 Weighted Images Showing Hypointense Dural Covexity Lesion at Left Frontal and Parietal Region with Peri Lesional Edema

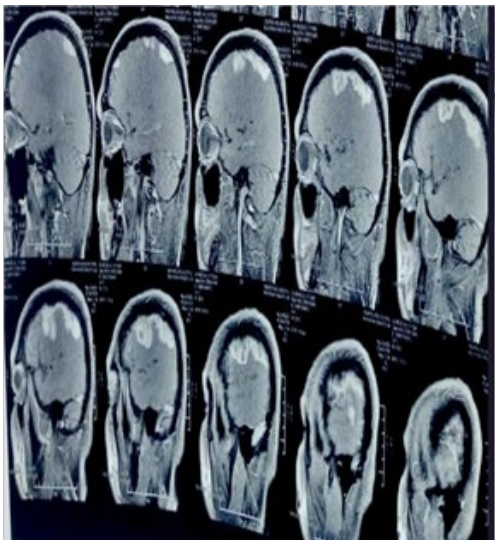

Figure 3: MRI Brain Sagital Section

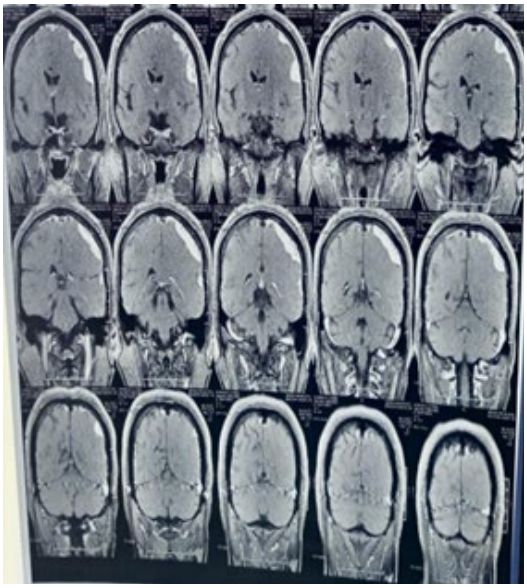

Figure 4: MRI Brain Coronal Section

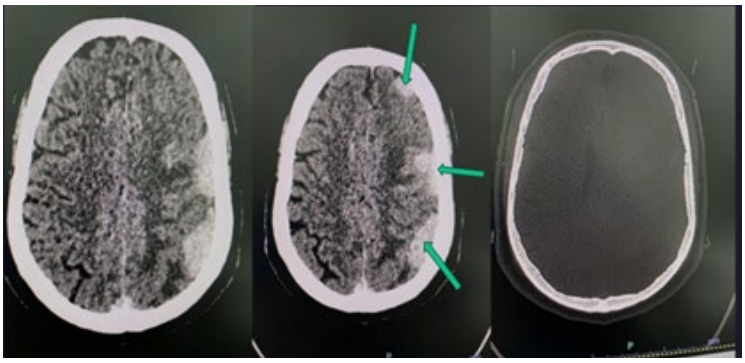

Figure 5: CT Brain Showing Multiple Dural Based Extra-Axial Enhancing Lobulated Lesion Along Left Frontoparietal Convexity Without Bony Erosions and Hyperostosis

Clinical and radiological diagnosis was of en-plaque meningioma of left frontoparietal convexity compressing the underlying parenchyma. Patient was planned for craniotomy and surgical excision of lesion. 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. On post op day 3 patient had an episode of focal seizure in right hand. CT brain was done which showed patchy edema in the subcortical white matter 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.

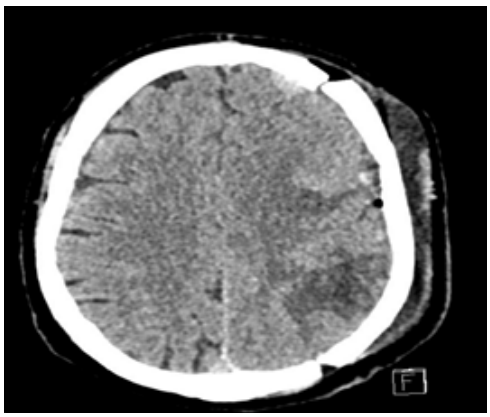

Figure 6: Post-Operative CT Brain

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 pale histiocytes displaying emperipolesis which were positive for S-100 and CD68 proteins and negative for CD1a marker. BRAF V600E mutation was negative rosai dorfman disease.

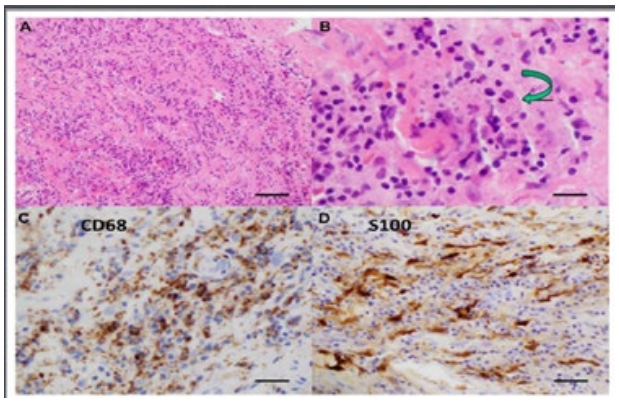

Figure 7: A: Histiocytic Cells, Admixed with Plasma Cells, Lymphoid Cells B: Emperipoletic Activity C:CD68 Positivity D: S100 Positivity

Post operative period was uneventful with no 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.

2. Discussion

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' and previously classified by the Working Group of the Histiocyte Society of 1987 as a non langerhans cell (LC) histiocytosis. It is a benign self-limiting condition characterized by the accumulation of activated histiocytes in the sinusoids of lymph nodes and/or extranodal tissues [1]. Classic RDD presents with massive bilateral painless cervical lymphadenopathy with associated fever, loss of weight and night sweats. Extranodal disease is seen in over 40% of cases and may rarely occur in the absence of nodal disease, usually in older patients with different demographics. It predominantly affects children and young adults, but the disease can be seen in the range of 1 to 74 years old. 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 [2]. They can occur in the suprasellar region, convexity, parasagittal region, cavernous sinus, petroclival region and cerebellum. CNS Rosai-Dorfman disease shows a predilection for males, and typically presents during the 4th to 5th decade with the mean age of 39.4 years old [3]. The definitive diagnosis is based on histopathological examination and immunohistochemistry. RDD can be misdiagnosed with different isotypes of meningioma and other en plaque dural lesions.

3. Conclusion

RDD is an idiopathic disorder classified as a histiocytosis, which typically presents with massive cervical lymphadenopathy. Intracranial variety is very rare (5%) it may lead to, although rare, serious complications like blindness and deafness. this can be treated satisfactorily with surgery and or corticosteroid. So early and accurate diagnosis is very important. Extranodal RDD, especially isolated intracranial RDD as shown in the present case, may pose a diagnostic challenge both for the clinician, radiologist and the pathologist as radiological and clinical features mimics like en plaque meningioma. Definitive diagnosis primarily relies on histopathological examination of biopsy tissue(emperipolesis,CD68,S100 staining), and resection of the intracranial mass is the most effective treatment for intracranial RDD. When neurologic symptoms persist or there are lesions around vital structures, the implementation of adjuvant therapies is recommended, together with localized radiotherapy. To explain regarding close follow up to the patient is very important as chances of recurrence is 14-29 %. RDD should be differentiated from IgG4 related disease as well .IgG4 related disease responds very well with corticosteroids alone.

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