Primary CNS vasculitis presenting as thalamic mass: A case report

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ABSTRACT –
Central nervous system vasculitis is an uncommon condition that poses considerable diagnostic and therapeutic challenges. Clinical presentation of primary angitis of the central nervous system is highly variable and there is no set clinical presentation. Advances in neuroimaging & molecular testing has increased the chances of diagnosis. Biopsy of CNS tissues showing vasculitis is the only definitive test. Recent studies suggest that on early diagnosis and prompt initiation of treatment, favourable clinical outcomes may be achieved.

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INTRODUCTION –

Central nervous system vasculitis is a rare and complex disease of the vasculature of the central nervous system that poses considerable diagnostic and therapeutic challenges. It can be divided into primary angitis of the central nervous system (PACNS) or secondary angitis of the central nervous system when associated with systemic conditions. Clinical presentation can be extremely variable and there is no classical presentation.[1]

The median age of onset is around 50 years. Biopsy of the central nervous system tissues showing vasculitis is the only definitive test.[2]

Challenges in diagnosis of CNS vasculitis still exist due to the broad differential diagnosis and generally non-specific initial clinical manifestations. Differentiation between primary and secondary angitis is important in guiding treatment. Advances in neuroimaging and molecular testing have enhanced diagnostic decision making. Despite the limited use of targeted therapies, glucocorticoids and cyclophosphamide remain the main stays of the therapy in primary angitis of CNS.[3]

Mass lesion presentation of primary angitis of the central nervous system (ML-PACNS) is a special subtype of primary central nervous system vasculitis, which is difficult to be differentiated from other space occupying lesions. It poses a huge challenge in clinical practice.[4]

Contrary to historical reports, recent PACNS cohorts achieved favourable clinical outcomes when early diagnosis and prompt treatment was initiated.[1]

CASE REPORT –

A 50 years old female presented with complaints of weakness of the right side of the body with unresponsiveness to verbal commands for 3 days. The weakness was sudden in onset, affecting both upper and lower extremities. She was having history of diabetes mellitus type 2 and prior

appendectomy. There was no past or family history of hypertension.

On examination, she was afebrile, normotensive and without icterus. In respiratory system, bilateral AE were present. S1 & S2 heard normally. On abdominal examination it was soft. On CNS examination, GCS was E4VTMS, pupils were 1.5 mm and non-reactive, right hemiparesis was present. Power of upper limb was 0/5 while of lower limb was 0/5.

Patient came in the hospital with priorly done MRI. On evaluation of MRI it was found to be thalamic mass with hydrocephalus (Fig. 1 and 2).

Fig 1: Magnetic Resonance Imaging

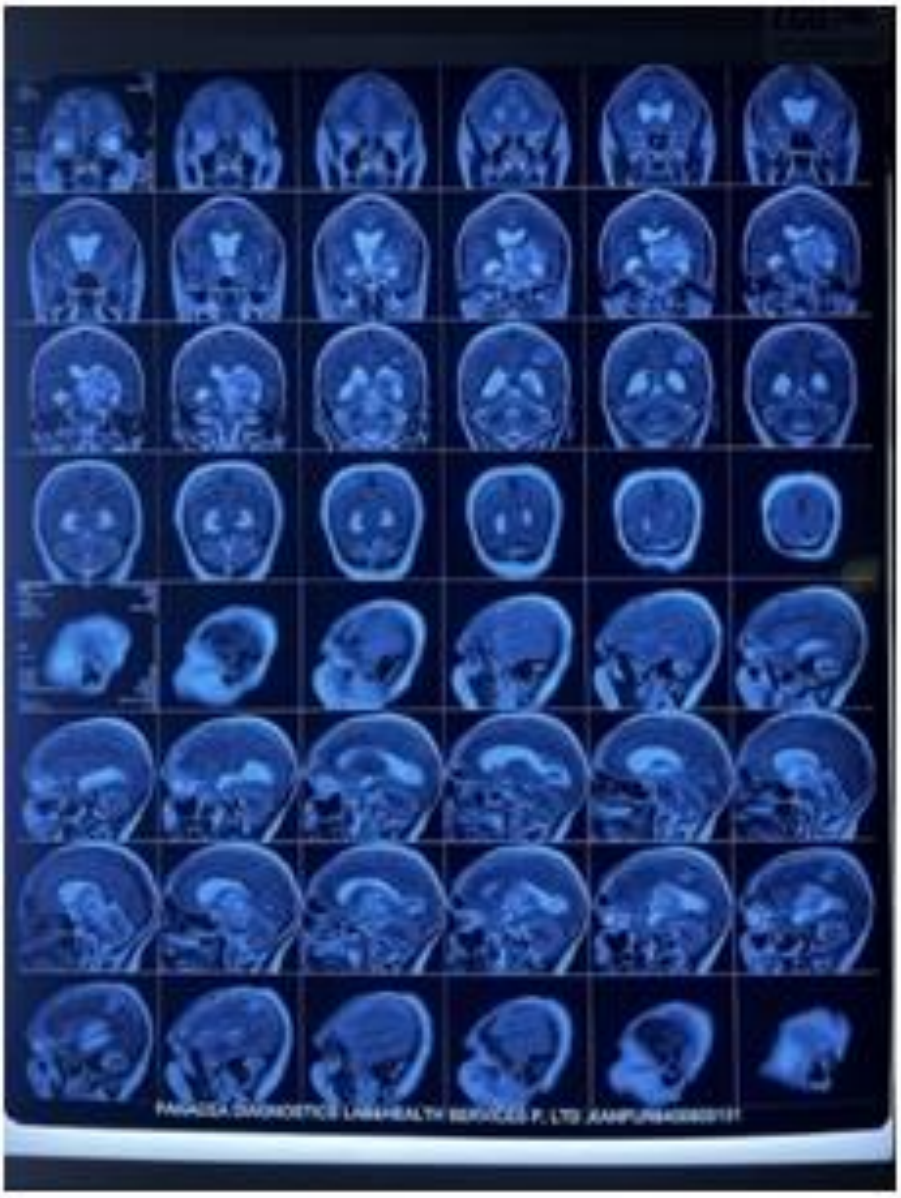
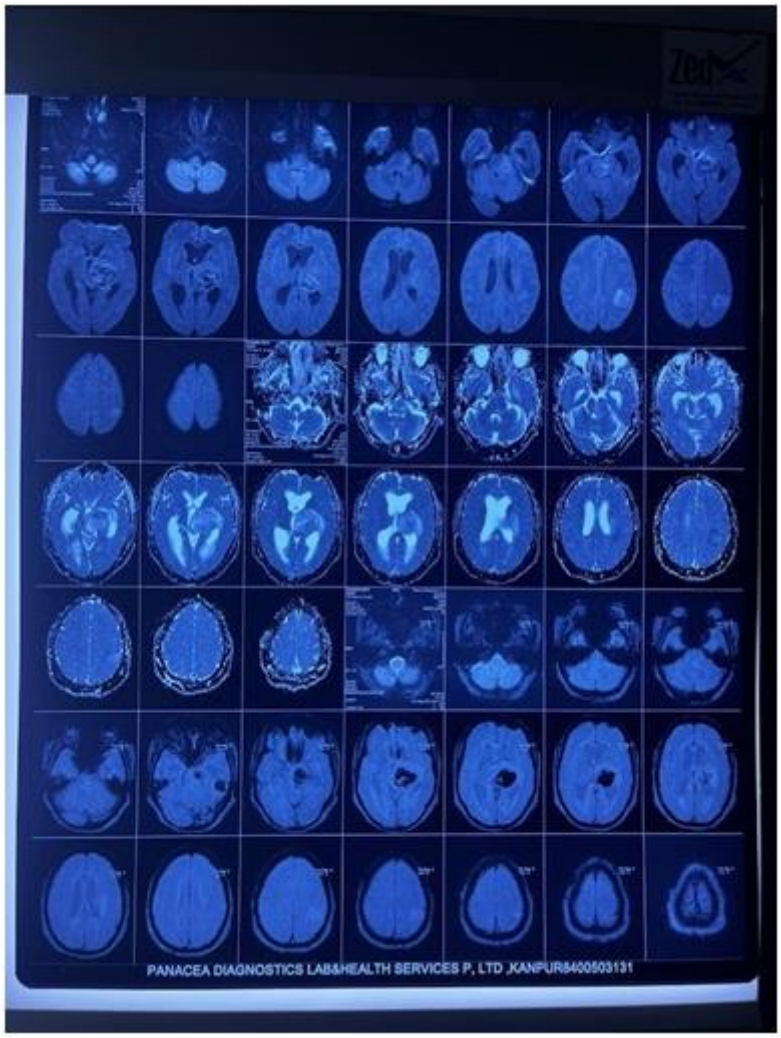


Fig 2: Magnetic Resonance Imaging shows thalamic mass with hydrocephalus



On blood investigation, Anti PR3 antibody were positive while Anti-nuclear antibody (ANA) were also positive with intensity of 3+ and pattern fine speckled. These blood test also indicate CNS vasculitis.

Endoscopic third ventriculostomy with biopsy of tumour tissue with omaya reservoir placement under GA. On evaluation of biopsy, primary angitis of central nervous system was the diagnostic decision.

DISCUSSION / CONCLUSION

ML-PACNS is one of the clinical subtypes of primary central nervous system vasculitis accounting for 5-15% of PACNS cases.[5] The median age of the onset was previously reported to be 37-45.5 years [4] but our case presented at 50 years of age. This variation in age may be because the lesion may be occurring late in India. Way of presentation in

our case was similar to other space occupying lesions (hemiparesis etc.). Biopsy remains the ultimate and confirmatory mode of diagnosis.

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