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CASE REPORT

Inflammatory demyelinating pseudotumor with hemorrhage masquerading high grade cerebral neoplasm

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KEYWORDS

Brain tumor;
Demyelination;
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Abstract Demyelinating pseudotumors are rare, benign, solitary intracranial space occupying lesions which masquerade cerebral neoplasms. Contrast MRI shows open ring enhancement which is fairly specific for this entity. Advanced MRI techniques like MR spectroscopy and magnetizing transfer techniques can help differentiating these lesions. NAA/Cr ratio is significantly elevated in central regions of demyelinating pseudotumors than in gliomas and other lesions. Presence of abundant foamy macrophages, lymphoid inflammatory infiltrates around blood vessels, sheets of gemistocytic astrocytes with well-developed processes, well defined border of the lesion absence of neovascularity and necrosis should help us diagnose demyelinating pseudotumor fairly confidently on histopathology.

1. Introduction

Demyelinating pseudotumors are rare, benign, solitary intracranial space occupying lesions which masquerade brain neoplasms clinically, radiologically and histopathologically (1–7). It is very difficult to differentiate these lesions from brain tumors as there is a considerable clinico-radiological overlap and it becomes nearly impossible to differentiate on routine cross sectional imaging when they present with hemorrhage or show complete rim enhancement (1,3,4,8). Subsequent relapse of symptoms and the presence of multiple abnormal signal intensity plaques, should raise suspicion of demyelinating pseudotumor (5). We describe a case of 43 year old female who presented to us with left parietal lobe hemorrhagic lesion which we initially presumed as glioma but biopsy revealed inflammatory demyelinating pseudotumor.

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2. Case report

A 43 year old female came to the emergency department with the history of sudden onset of loss of consciousness followed by generalized tonic-clonic seizures. On admission, her vitals were stable (Blood pressure – 146/90). Patient was not a known hypertensive or diabetic. She had recent complaints of febrile illness, headache, weight loss and weakness for which was admitted in local rural hospital and was treated. Apparently she worsened during treatment and she was referred to our hospital. On examination – pupils were normal and reacting to light, motor examination showed decreased power in right upper and lower limbs, plantar reflex was extensor on right side, sensory examination was unremarkable and lab investigations were within normal limits. MRI brain showed a 5×6 cm T1 hyperintense and T2 heterogenous signal intensity lesion in the left parietal lobe with extensive perilesional vasogenic edema (Figs. 1 and 2) and ovoid hyperintensities in periventricular white matter (Fig. 2). On contrast administration, there was complete rim enhancement of the lesion (Fig. 3). Initial differentials thought were glioma and venous infarct however later was ruled out as venous sinus flow voids were maintained. Presuming the lesion as glioma, right parietal craniotomy and excision was performed and the specimen was sent for histopathological examination. Sections showed numerous vascular channels dissecting into the parenchyma; vascular channels were lined by prominent endothelial cells and surrounded by scattered spindle cells. Scanty areas of necrosis were seen surrounded by golden yellow pigment representing hemosiderin. Numerous foamy macrophages were also noted. These features were consistent with demyelinating pseudotumor. Patient was given high dose corticosteroids and she recovered drastically from weakness (see Fig. 4).

3. Discussion

Inflammatory demyelinating pseudotumor (IDMP) is a puzzle to clinicians, radiologists and pathologists due to many overlapping features (1,3–5,7). Pathogenesis of IDMPs is not clear and it is different from multiple sclerosis and acute disseminated encephalomyelitis (9). Patients with IDMP or brain tumors can present with frequent headaches, giddiness, altered

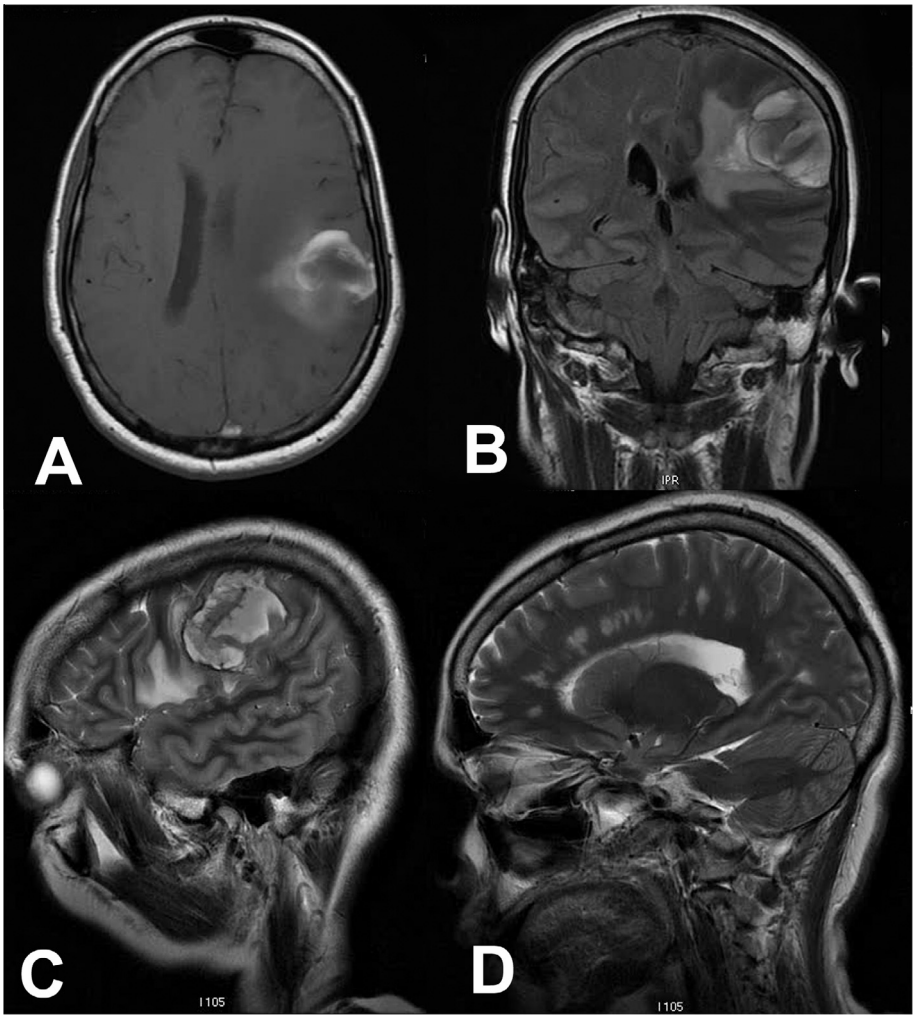

Fig. 1 Axial T2 weighted MR image showing large heterogenous predominantly hyperintense left parietal lobe lesion showing hypointense hemosiderin rim, perilesional extensive vasogenic edema and mild midline shift to right. Also note the periventricular perpendicular hyperintense lesions in right frontal lobe suggestive of demyelination foci.