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Erdheim-Chester Disease Mimicking a Precentral Meningioma: A Case of Rapid Parenchymal Recurrence and Multisystemic Progression

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Abstract: **Case Report**

Erdheim-Chester disease (ECD) is a rare non-Langerhans histiocytosis that remains a significant diagnostic challenge, particularly when it presents with dural involvement. We describe the clinical course of a 50-year-old male, originally treated for a presumed precentral meningioma based on a dural mass showing intense enhancement and an atypical T2-hypointense signal. Following surgical resection, the patient experienced a multisystemic flare (thoracic, hepatic, and adrenal) alongside a rapid multifocal parenchymal recurrence in the brain. Biopsy of the thoracic lesion confirmed a BRAF V600E-mutated ECD. This report emphasizes that a "black" T2 signal in a dural mass is a major clinical red flag. Radiologists should systematically correlate such findings with the patient's cardiac history to distinguish ECD from more common dural tumors and avoid unnecessary surgery.

Keywords: Erdheim-Chester Disease, Histiocytosis, Brain MRI, Meningioma mimic, BRAF V600E, T2 hypointensity.

1. INTRODUCTION

Erdheim-Chester disease (ECD) is a multisystemic myeloid neoplasm defined by the infiltrative spread of foamy, lipid-laden histiocytes [1]. Since the discovery of its frequent association with the BRAF V600E mutation, our understanding has shifted from a simple inflammatory process to a targeted oncological pathology. While the skeletal system is nearly always involved, neurological manifestations appear in about 40% to 50% of patients and are a leading cause of mortality [2]. The radiological spectrum of CNS-ECD is broad, ranging from neurodegenerative patterns to dural masses that mimic meningiomas due to their location and contrast uptake [3, 4]. We present a case that illustrates how the specific MRI "T2-hypointense signature" and a past history of a cardiac mass could have prevented a surgical misdiagnosis.

2. Case Presentation

A 50-year-old man sought neurosurgical consultation for a progressive weakness in his left lower limb. Interestingly, his records showed a prior surgery for a "left atrial mass" five months earlier, the nature of which had remained unclear.

Initial Pitfall and Surgery

The first brain MRI identified a solitary, right precentral dural-based mass. While the lesion enhanced vividly after gadolinium, its signal was strikingly dark on T2-weighted sequences. Misled by the location and the enhancement pattern, our team proceeded with a gross total resection under the presumptive diagnosis of an atypical meningioma. Post-operative histopathology revealed only non-specific fibrous changes and chronic inflammation, failing to provide a definitive diagnosis.

Systemic Unveiling

The diagnosis was only clarified months later when the patient presented with respiratory distress. CT imaging then revealed an extensive thoracic soft-tissue mass, bilateral adrenal thickening, and a nodular infiltration of the greater omentum. A CT-guided biopsy of the mediastinal mass finally demonstrated the classic CD68+, CD1a-, S100- histiocytic infiltration of Erdheim-Chester disease. Genetic testing confirmed the BRAF V600E mutation.

Neurological Relapse

A follow-up brain MRI was urgently performed. It showed the known surgical cavity but also revealed two new, well-defined nodular relapses: one in the right frontal region and another in the left parietal lobe. These new lesions were markedly hypointense on Coronal T2-weighted images **(Figure 1A)** and hyperintense on FLAIR **(Figure 1B)**. Peripheral micro-hemorrhages were noted on T2* sequences **(Figure 3B)**. Following contrast injection, both nodules showed the same homogeneous enhancement as the initial dural mass **(Figure 2)**. Diffusion imaging (DWI) and ADC maps showed no significant restriction, ruling out hypercellular malignancies like lymphoma **(Figure 3A)**. The patient was immediately started on BRAF-inhibitor therapy.

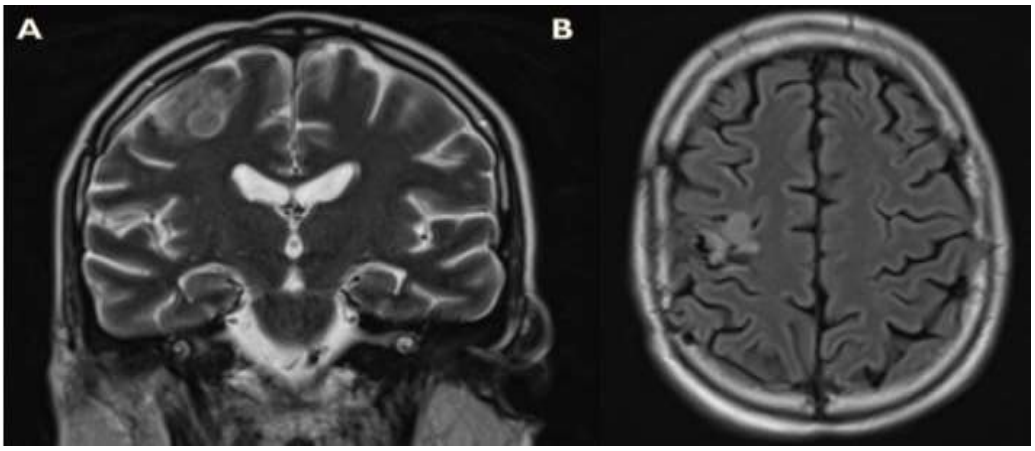

Figure 1: Morphological MRI signatures of CNS recurrence: (A) Coronal T2-weighted image demonstrating the marked hypointensity of the cortical-subcortical nodule (arrow). (B) Axial FLAIR image showing the hyperintense signal of the recurrence with associated gyriform infiltration

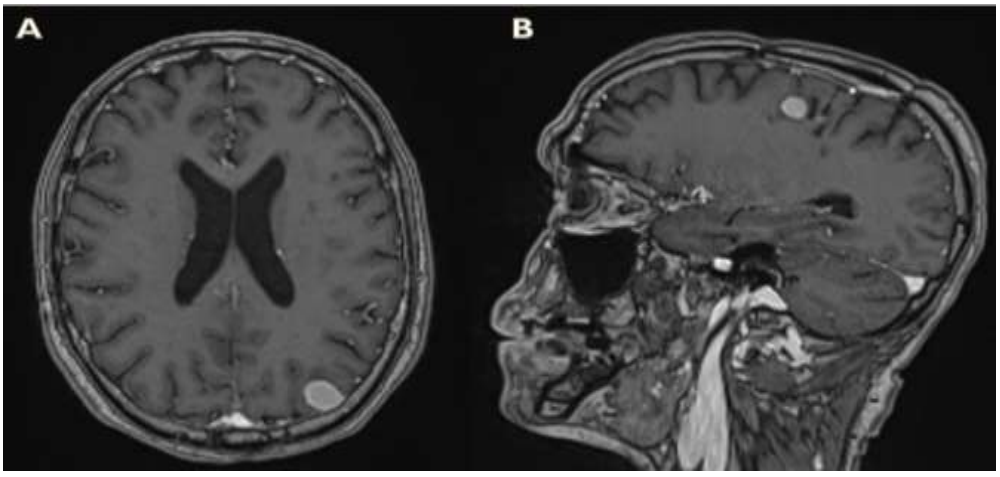

Figure 2: Enhancement pattern: Axial (A) and Sagittal (B) T1-weighted images after gadolinium injection showing intense, homogeneous enhancement of the lesions, mimicking the appearance of extra-axial dural tumors

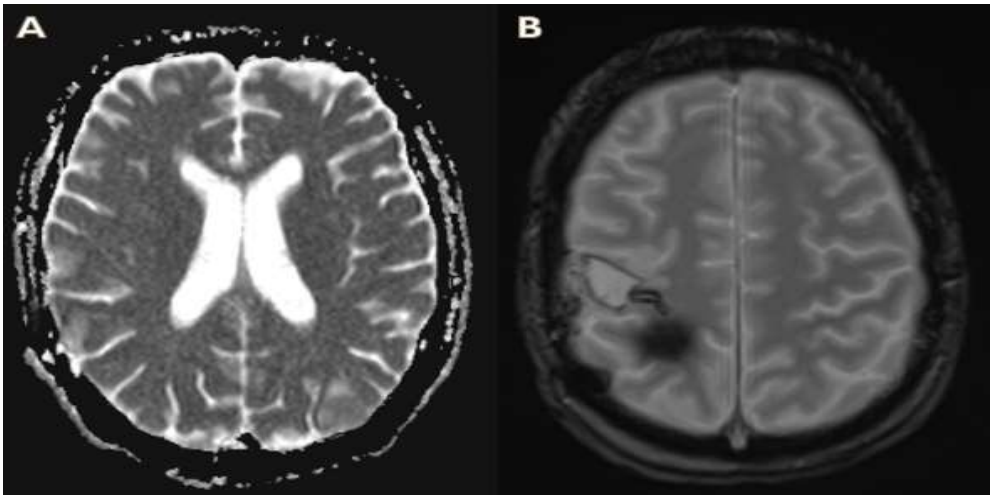

Figure 3: Advanced MRI sequences: (A) ADC map showing facilitated diffusion (high signal) within the lesion, helping differentiate it from hypercellular tumors. (B) Axial T2* sequence revealing peripheral hemorrhagic stigmata (hypointense signal)

3. DISCUSSION

Dural ECD is a well-known "mimic" of meningiomas. As noted by Johnson *et al.*, these lesions often present as enhancing masses on standard T1-weighted sequences, frequently leading to unnecessary neurosurgical interventions [5].

The T2 Signal: A Crucial Discriminator

Our case highlights that the key to the diagnosis lies in the T2 signal. Typical meningiomas are isointense or hyperintense on T2 sequences. In contrast, ECD lesions are almost always "T2-black" because of the intense fibrosis and the high density of xanthomatous histiocytes [2]. In our patient, this signal was pathognomonic and was present in both the initial dural lesion and the subsequent parenchymal recurrence.

The "Coated Heart" Connection

The patient's "left atrial myxoma" was a retrospective clinical smoking gun. Cardiac involvement occurs in over 70% of ECD cases, often presenting as a pseudotumoral infiltration of the right atrium or atrioventricular groove [6]. These masses are frequently misdiagnosed as myxomas when viewed in isolation. Had this prior cardiac event been correlated with the dark T2 brain lesion, a systemic CT or PET-scan would likely have established the diagnosis of ECD non-invasively.

Progression and Molecular Targeted Therapy

The transition from a solitary dural mass to multifocal parenchymal nodules illustrates the aggressive potential of BRAF-mutated ECD. However, the prognosis of CNS-ECD has been revolutionized by targeted therapies (such as vemurafenib or cobimetinib), which often lead to spectacular radiological regression where conventional chemotherapy failed [1, 2].

4. CONCLUSION

Erdheim-Chester disease should be high on the differential list for any dural mass that is notably hypointense on T2-weighted MRI. Clinicians must meticulously check for a history of cardiac "pseudotumors" or systemic infiltrative signs. Recognizing these radiological patterns early is essential to bypass surgery and direct the patient toward molecular testing and targeted therapy.

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