Intracranial Erdheim-Chester Disease Mimicking Parafalcine Meningioma: Report of Two Cases and Review of the Literature

Disclosures

None
Introduction

- Erdheim-Chester disease (ECD) is a rare, non-Langerhans cell histiocytosis that typically occurs in middle-aged patients, usually characterized by multifocal osteosclerotic lesions of the long-bones.

- While, many cases have extraskeletal involvement, central nervous system (CNS) involvement is unusual and isolated CNS involvement in ECD is rarely seen.

- We report 2 cases of dural-based ECD mimicking meningioma, where the lesion was the only site of disease.
Methods

• We review the clinical presentation, imaging, workup, management, and most recent clinical status of two patients with a CNS lesion as the initial presentation of ECD. We include pathologic analysis at our institution, as well as classic findings and genetic analysis.

• We review literature on CNS involvement in ECD, including a spectrum of imaging findings
Results

- The first patient was a 60M with a dural-based mass found on headache workup.

- The lesion progressed on serial imaging, and the patient underwent uncomplicated surgical resection (Figure 1).

- Pathology was consistent with ECD (Figure 2), and BRAF-V600E testing was negative. Extensive systemic workup showed no other sites of disease.

- The patient recovered well, and has no evidence of residual or recurrent disease.
Results

Figure 1. Coronal (A) and axial (B) T1 post-contrast MRI 1 year later showing left parafalcine homogeneously enhancing lesion

Figure 6. A) H&E stain showing a mixed population of mature plasma cells and lymphocytes, no eosinophils. B) CD68 showing abundant staining of histiocytes. C) FXIIIa with focal strong positivity
Results

• The second patient was a 42F with parafalcine mass discovered on imaging obtained for worsening headaches

• The lesion progressed on surveillance imaging (Figure 3), and she underwent surgical resection

• Pathology was consistent with ECD, with negative BRAF-V600E. Extensive systemic workup showed no other sites of disease

• The patient experienced mild right-sided weakness post operatively, which improved with physical therapy

• Post-operative surveillance imaging shows stable residual lesion
Results

Figure 3. Coronal (A) and axial (B) T1 post-contrast MRI showing left parafalcine dural-based mass with homogeneous contrast enhancement and moderate local mass effect.

Figure 4. Coronal (A) and axial (B) T1 post-contrast MRI showing residual lesion.
Erdheim Chester Disease is a non-Langerhans histiocytosis generally characterized by involvement of the long bones.

Involvement of the CNS is common, but isolated lesions are exceedingly rare and can mimic other benign entities; presentation varies by location of the lesions(s), and imaging can resemble a multitude of other pathologies.

We present two cases and review the literature on isolated CNS lesions of ECD mimicking meningioma on MRI, with negative BRAF-V600E mutation.

Treatment is for systemic disease is improving with the advent of new targeted therapies.
ECD is a rare condition, with isolated CNS involvement even more uncommon.

Lesions that exhibit growth or have an unusual presentation should undergo surgery for tissue diagnosis.