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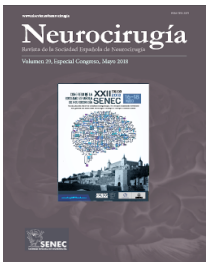

P0069 - ERDHEIM-CHESTER DISEASE MIMICKING AN INTRACRANIAL TEMPORAL MENINGIOMA: CASE REPORT

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Resumen

Objectives: Erdheim-Chester disease (ECD) is a rare, sporadic, non-Langerhans histiocitosis with unknown etiology. This disease is known to involve multiple organ systems, including skeletal, cutaneous, cardiovascular, respiratory, urinary, and central nervous systems. Neurological involvement has been reported in up 50% of patients. Here we present 1 case of ECD presenting as a dural-based lesion resembling a intracranial temporal meningioma.

Methods: A 35-year-old man was evaluated for right-sided peripheral facial nerve palsy. Brain CT scanning with bone windows revealed scalloping of the temporal bone and right petrous apex due to expansive mass of temporal lobe. Contrast-enhancing brain MRI showed a heterogeneously Gd-enhancing mass in the right middle cranial fossa, extending to the infratemporal fossa and petrous apex (mastoid air cells, and middle ear). A right temporal craniotomy was performed for resection of the mass. His post-operative course was uneventful and after an overnight stay in intensive care unit, he was discharged home and his neurological baseline on post-operative day 3. Pathology was consistent with ECD, and BRAF V600E mutation was negative.

Results: Isolated lesions of ECD in Central Nervous System are exceedingly rare and can mimic other benign entities. Presentations vary by location of the lesions(s), and imaging can resemble a multitude of other pathologies.

Conclusions: Lesions that exhibit growth or have an unusual presentation should undergo surgery for tissue diagnosis. If safe to do so, enlarging or symptomatic lesions should be resected completely.