Progressive hearing loss and cerebellar ataxia in anti-Ma2-associated autoimmune encephalitis

Perda auditiva progressiva e ataxia cerebelar na encefalite autoimune associada ao anti-Ma2

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A 38-year-old Brazilian man presented with a two-year history of progressive bilateral hearing loss, cerebellar ataxia, emotional lability and hypersexuality. A limbic-brainstem syndrome was suspected and neuroimaging studies performed, disclosing marked brainstem signal changes (Figure). Cerebrospinal fluid analysis disclosed a mild protein increase. During paraneoplastic screening, testicular ultrasonography disclosed a heterogeneous complex mass in the right testicle (Figure) and serum anti-Ma2 antibodies were detected.

Anti-Ma2-associated encephalitis classically emerges in the context of seminomatous and nonseminomatous testicular tumors and presents with a complex spectrum of neurological manifestations[1], including limbic encephalitis, atypical parkinsonism, cerebellar ataxia, brainstem dysfunction, myelopathy, radiculoplexopathy, REM sleep behavior disorder and narcolepsy[1,2].

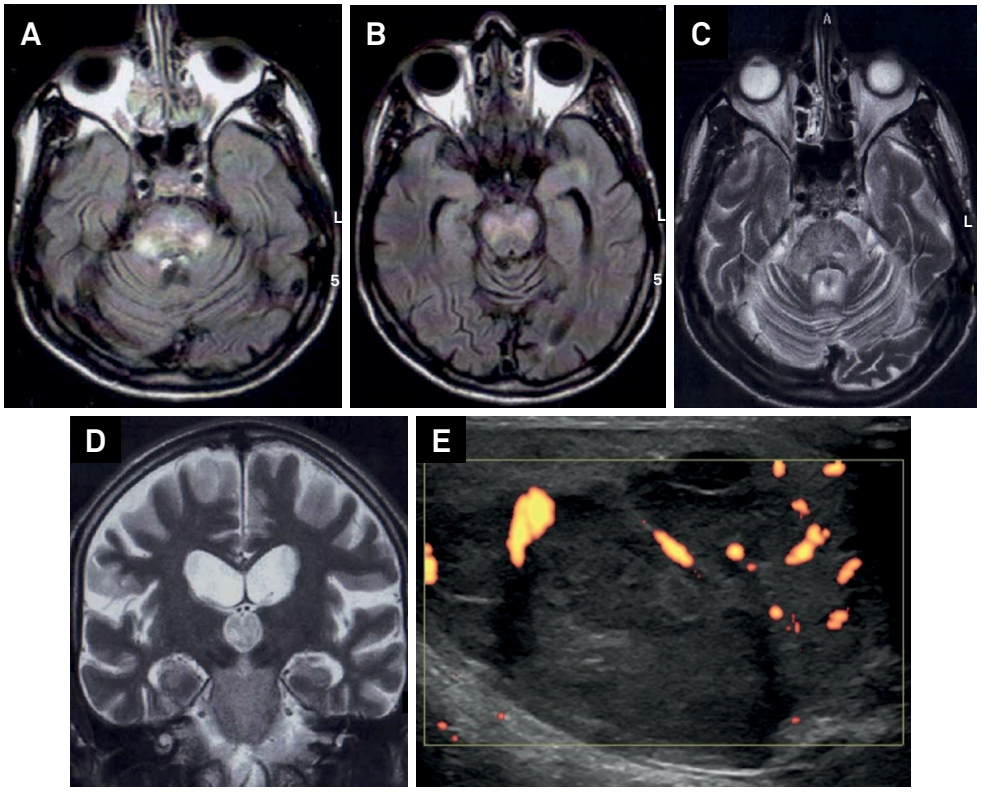

Figure. Brain MR imaging studies disclosing cortical atrophy, bilateral hyperintensity in the mesial temporal lobes and marked hyperintense signal change in the pons and superior cerebellar peduncles in axial FLAIR sequence (A,B) and axial (C) and coronal T2-weighted images (D). (E) Ultrasonography of the testis disclosing a heterogeneous complex mass in the right testicle with the presence of flow inside the lesion in the Doppler study.

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Ethical statement: Full consent was obtained from the patient's family for the case report. This study was approved by our Ethics Institution.

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