Lymphomas–Part 2

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KEYWORDS

• Lymphomatosis cerebri • Intravascular lymphoma • Lymphomatoid granulomatosis

KEY POINTS

- Lymphomatosis cerebri (LC) is a rare type of infiltrative lymphoma that usually does not enhance.
- In LC, restricted diffusion is minimal or absent.
- Magnetic resonance spectroscopy (MRS) may distinguish LC from gliomatosis cerebri (GC).
- Intravascular lymphoma (IVL) may mimic ischemic lesions.

SPECIAL LYMPHOMA TYPES

Lymphomatosis Cerebri

LC is a rare variant of primary central nervous system (CNS) lymphoma (PCNSL) pathologically characterized by diffuse cerebral infiltration of a noncohesive mass of malignant lymphoid cells.[1–6] The clinical picture is variable and includes abnormal behavior, personality changes, gait disturbance, seizures, memory deficits, and rapidly progressive dementia and weight loss.[4–8]

White matter abnormalities in LC affect all of the brain. MR imaging findings are extensive, diffuse T2 and FLAIR-weighted hyperintense lesions without formation of a cohesive mass and no contrast enhancement in both cerebral hemispheres and brainstem (**Fig. 1**).[6–11] Subtle or patchy enhancement may be seen.[8,10] There may be a transition from nonenhancing to enhancing lesions suggesting that progression and evolution is associated with disruption of the blood-brain barrier.[4] Restricted diffusion may be minimal or absent (see **Figs. 1**F, G).[10] Many cases respond to steroids alone, at least initially. To achieve complete remission, steroids are usually followed by radiotherapy, cisplatin, or methotrexate.[12]

Pearls

- LC is a diagnostic challenge.
- In patients presenting with diffuse, bilateral, asymmetric signal abnormalities in white matter, infiltrative lymphoma should be considered, especially if there is callosal involvement.
- Consider brain biopsy in rapidly progressive cognitive decline to allow earlier therapy for a potentially curable disease.[4]
- Awareness of this rare disease and early biopsy are required for preventing a poor clinical outcome.[6]

Intravascular Lymphoma

IVL was originally described in 1959[13] and designated as "angioendotheliomatosis proliferans systemisata." Since then, it has been referred to as neoplastic angioendotheliosis, malignant angioendotheliomatosis, and angiotropic large-cell lymphoma.[14–17]

IVL is a rare subtype of extranodal diffuse large B-cell lymphoma with a distinct presentation. Anatomically, it is characterized by proliferation of clonal lymphocytes within small vessels with

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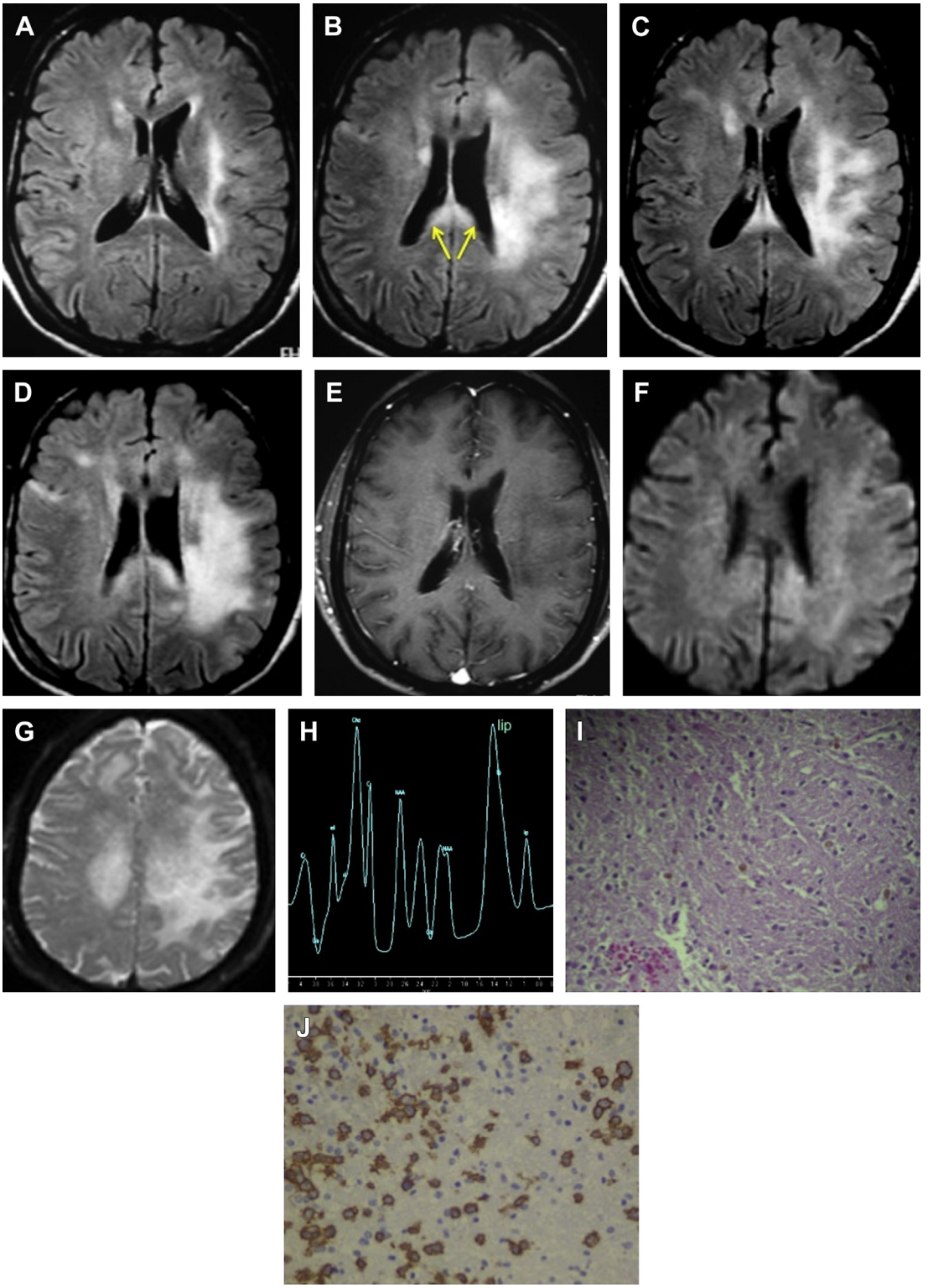

Fig. 1. LC. A 40-year-old man with status epilepticus. There is an ill-defined infiltrating lesion in the left frontal and parietal lobes ([*A, B*] axial FLAIR), extending to the splenium of the corpus callosum ([*B*] *arrows*). MR imaging 1 month later ([*C, D*] axial FLAIR) shows progression. No enhancement is demonstrated ([*E*] axial T1 with contrast) and there is no restricted diffusion ([*F*] DWI, [*G*] ADC map). (*H*) MRS shows low NAA and mI, high Cho, and lipid peaks, characteristic of LC ([*I, J*] confirmed at pathology). ADC, apparent diffusion coefficient. (*Courtesy of* Dr Leonardo Avanza, Vitória, Rio De Janeiro, Brazil; and Leila Chimelli, MD, Rio De Janeiro, Brazil.)