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Primary sarcoma of the leptomeninges: unusual presentation and previously undescribed neuroimaging features

Sarcoma primário de leptomeninges: apresentação não usual e aspectos de neuroimagem previamente não descritos

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A 30-year-old patient presented with generalized tonic-clonic seizures. Magnetic resonance imaging and microscopic examination are demonstrated in Figures 1 and 2.

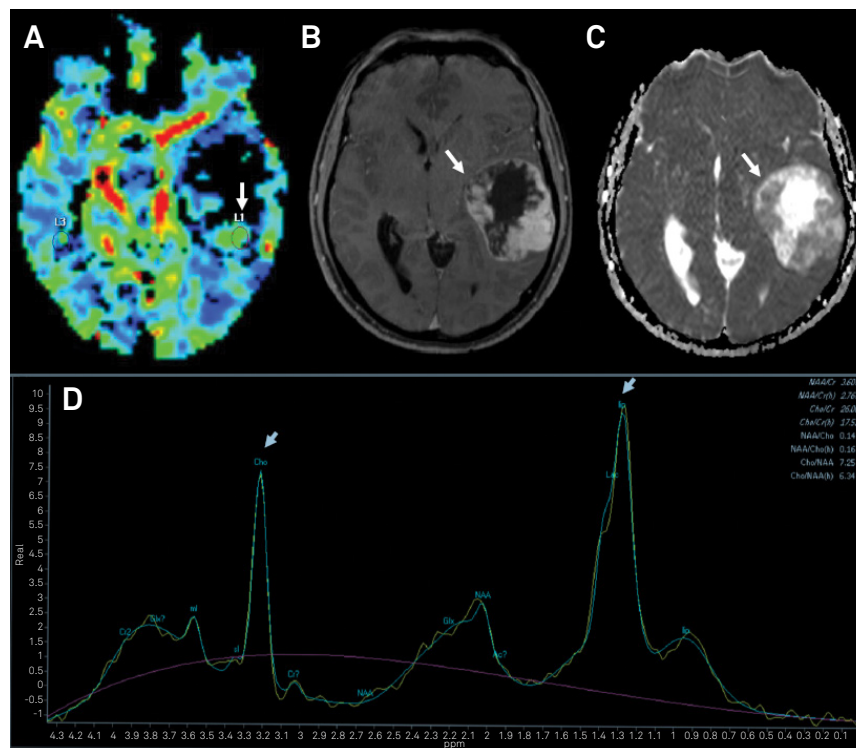


Figure 1. A. A high relative cerebral blood volume (rCBV) map can be seen in the left temporal lesion (arrow) and is represented in green. B. T1-weighted post-gadolinium axial depicts lesion in left temporal lobe with necrotic center and a peripheral solid component with heterogeneous enhancement (arrow). C. ADC map exhibits true restricted diffusion in peripheral solid component of the lesion (arrow). D. Spectroscopy shows a significant elevation of the choline peak in 3.2 ppm (indicating increased cell turnover) and of the lipids/lactate peaks (necrosis/anaerobiosis markers) in 1.3 ppm (arrows).

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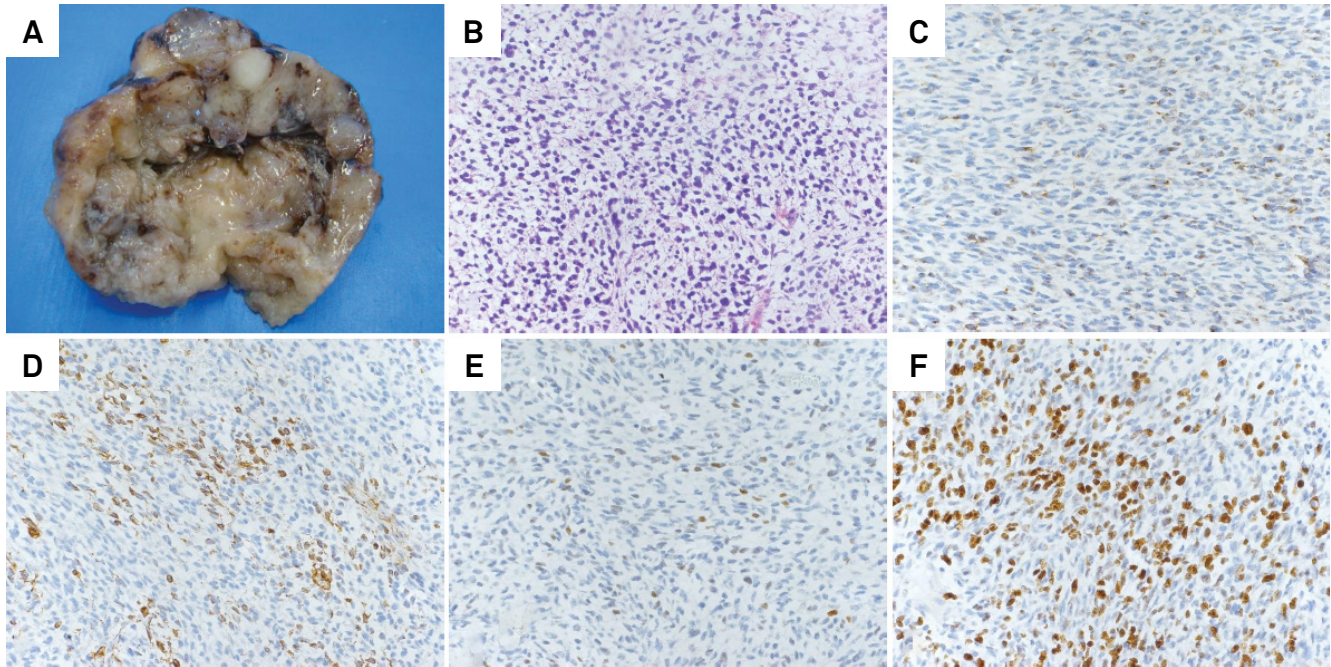


Figure 2. Neuropathology. Neoplasm with histological and immunostaining features compatible with an undifferentiated cerebral sarcoma of leptomeningeal origin. [A] Surgical specimen measured 7×3×3 cm and presented as a well-delimited, lobulated, friable yellowish and black mass. [B] On HE stain tissue was composed of small undifferentiated cells with basophilic nuclei and scant cytoplasm in a solid pattern. Fusiform and star-shaped cells were observed, as well as necrosis and apoptosis. On immunohistochemistry the lesion was focally positive for [C] epithelial membrane antigen (EMA), [D] vimentin and [E] p53. [F] Ki-67 was positive in up to 20% of cells in high-power fields. No staining was observed for AE1/AE3, S100 protein, CD34, desmin, GFAP, HHF-35 and 1A4. B, C, D, E and F: 20x magnification.

Histological analysis led to the diagnosis of undifferentiated cerebral sarcoma of leptomeningeal origin.

We illustrate a primary leptomeningeal sarcoma with hyperperfusion and high choline and lactate levels. Perfusion and spectroscopy are not yet discussed in the literature for these tumors.

Primary leptomeningeal sarcomas are aggressive tumors that account for 0.7% to 4.3% of intracranial neoplasm and mostly affect children^{1,2}.

Our case merits attention due to an unusual presentation concerning age and lack of apparent relation with the meninges.

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