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Teaching NeuroImage: Branching Dura Mater in Primary Central Nervous System, ALK-Positive, Anaplastic Large Cell Lymphoma

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A 22-year-old male presented with the new onset of gradually progressive diffuse headache for 2 months. He exhibited no apparent physical and neurological abnormalities except limited bilateral lateral rectus movement and bilateral papilledema on fundoscopic examination. The MRI showed multiple nodular enhancing lesions along both sides of the falx cerebri with perilesional edema. Additionally, these nodular structures formed a branching network of thickening dura mater to adjacent nodular structures (Figure 1). Due to the atypical features of dura mater, the differential diagnosis included tuberculosis, sarcoidosis, IgG4-related disorders, lymphoproliferative disorders, meningioma, hemangiopericytoma, and Erdheim-Chester disease. (1) A stereotactic biopsy was performed. The pathological and immunohistochemistry results, as well as systemic evaluation, were compatible with primary central nervous system anaplastic large cell lymphoma with ALK+. (2) After the total of six courses of high-dose methotrexate and ifosfamide followed by whole-brain radiation, the patient achieved complete remission and resolution of headache (Figure 2).

Abbreviation: ALK = Anaplastic lymphoma kinase, IgG = Immunoglobulin G

Ethical consideration: Informed consent was obtained from patient.

Author contributions: All authors contribute to the design & conceptualized the study, the acquisition of data, the interpretation of the data, and the writing / reviewing of the manuscript for intellectual content.

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Figure 1: The MRI brain with gadolinium prior to treatment. T1-weighted imaging with gadolinium revealed multiple bilateral nodular enhancing lesions along the falx cerebri, primarily located in the frontal lobe, and thickening of dura mater (Figure 1A – sagittal view, Figure 1B – coronal view, and Figure 1C – axial view). The red arrows demonstrated a branching network of thickening dura mater to adjacent enhancing nodular structures (Figure 1A). Axial T2-FLAIR image showed hypersignal intensity in the bilateral frontal lobes, suggesting the presence of perilesional edema (Figure 1D).

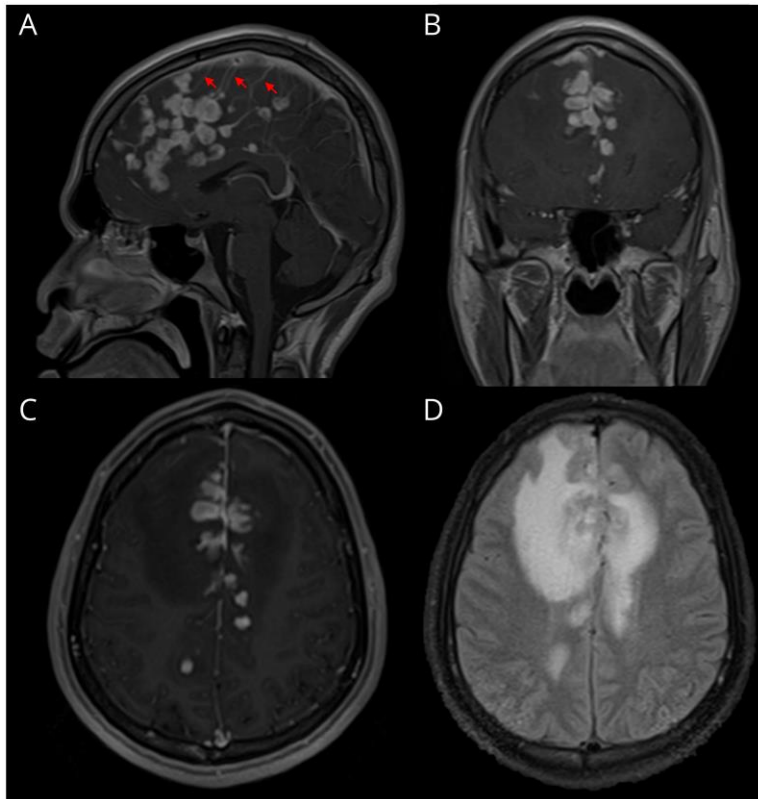
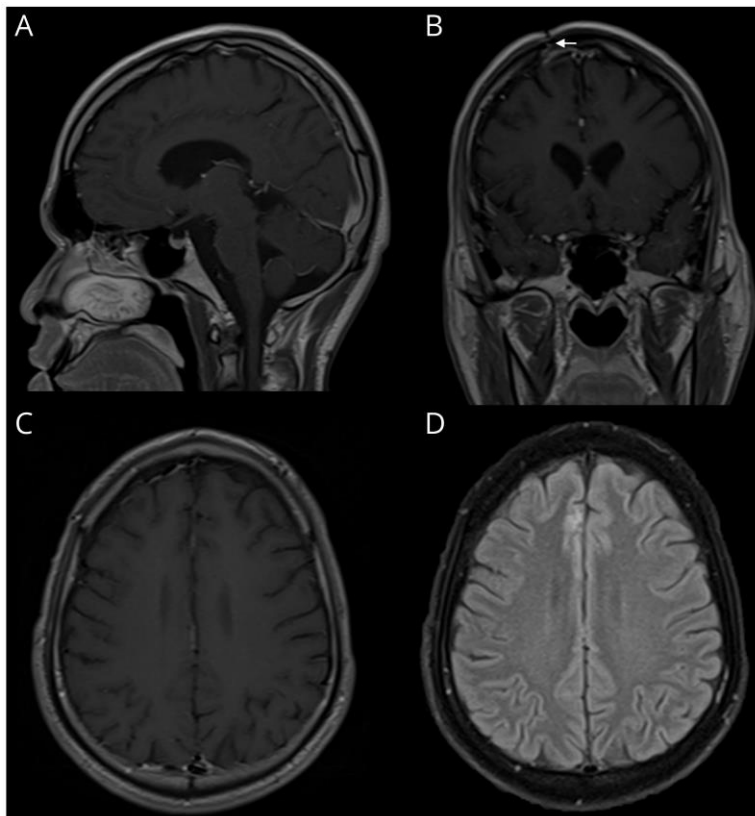


Figure 2: The MRI brain with gadolinium after 3 months of the total six courses of high-dose methotrexate and ifosfamide followed by 30.6 Gy/17 fraction of whole brain radiation. T1-weighted imaging with gadolinium revealed the nearly complete resolution of thickening dura mater, nodular enhancing lesions, and a branching network (Figure 2A – sagittal view, Figure 2B – coronal view, and Figure 2C – axial view). The white arrow demonstrated the presence of post-stereotactic brain biopsy and mild thickening of adjacent dura mater, primarily resulted from the post-operative change (Figure 2B). Axial T2-FLAIR image showed the resolution of hypersignal intensity in the bilateral frontal lobes (Figure 2D).



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