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NEURO-IMAGES

Primary central nervous system lymphoma revealed by multiple intraventricular mass lesions

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A 71-year-old man was admitted in our institution with a 1-month history of vertigo, vomiting, and weight loss. He had no relevant medical history. On admission, the patient was significantly disoriented and we observed a severe impairment of short-term memory. He was able to walk without aid. He needed help to wash and get dressed. Cranial nerves, motor, sensory, and cerebellar examination was normal.

Blood tests including basic metabolic panel, liver function panel, thyroid studies, markers of inflammation, vitamin B12, and folate were all normal, and infectious serum tests including HIV were negative. The patient had no immunodeficiency. He underwent contrast-enhanced brain magnetic resonance imaging (MRI) demonstrating multiple homogeneously enhancing intraventricular mass lesions, involving the whole ventricular system (Fig. 1). Cerebrospinal fluid (CSF) analysis showed lymphocytic pleocytosis (115 cells/mm³; $N \leq 5$) with increased protein (5.6 g/l; N: 15–45) and hypoglycorrhachia (0.41 g/l; CSF/serum glucose ratio = 0.33; normal ratio = 0.6). CSF flow cytometry was unremarkable and there was no evidence of malignant cells on CSF cytology. CSF analysis for *Mycobacterium tuberculosis* was negative. Biopsy of the intraventricular masses in the right temporal horn was performed to obtain a histological diagnosis. Histological examination (Fig. 2) showed diffusely distributed lymphoid cells with slightly-to-moderately abundant cytoplasm and visible nucleoli. Immunohistochemical analyses showed CD20 (+), Mum-1 (+), Bcl-2 (+), Bcl-6 (+), CD5 (marginally +), CD10 (−), and Ki-67 (index, 100%). The histologic features were consistent with a non-germinal center-like diffuse large B-cell lymphoma. Contrast-enhanced thoracic-abdominal-pelvic computed tomography scan revealing no co-existing systemic disease at the time of diagnosis cemented the diagnosis of PCNSL.

Three months later and after having received five cycles of chemotherapy with rituximab, methotrexate, procarbazine, and vincristine, the cognitive function remained dramatically impaired despite a significant radiological improvement on follow-up brain MRI (not illustrated). The patient could not perform any activities of daily living anymore.

Primary CNS lymphoma (PCNSL) is a rare form of extranodal non-Hodgkin lymphoma confined to the CNS. PCNSL represents approximately 2% of all primary brain tumors and is associated with substantial mortality [1]. Establishing its diagnosis is often challenging, since the clinical and radiological features may be variable and non-specific. The vast majority of PCNSL are supratentorial (thalamus, basal ganglia, corpus callosum, etc), but they

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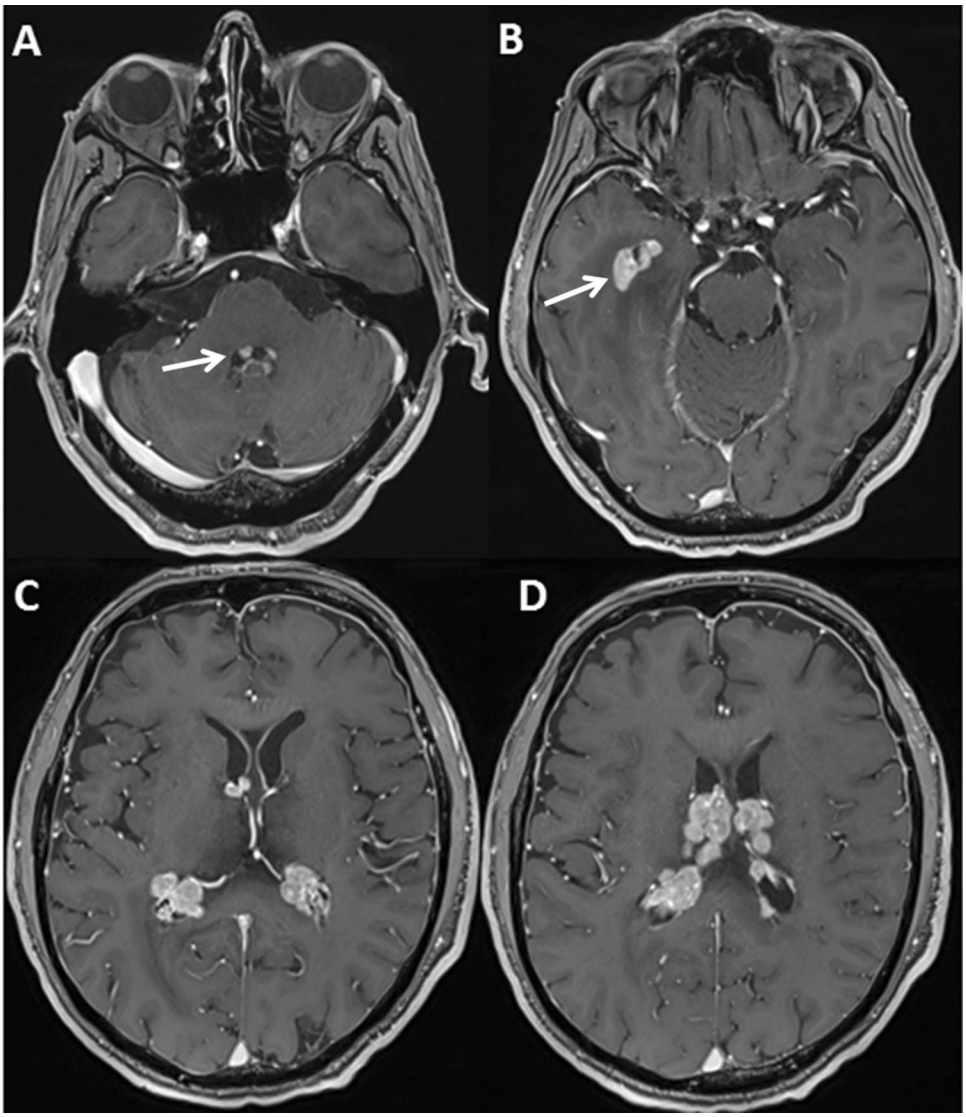

Fig. 1 Brain MRI. Axial contrast-enhanced T1-weighted MRI of the brain showed multiple homogeneously intraventricular enhancing mass lesions involving the fourth (**a**, white arrow), the right temporal horn (**b**, white arrow), and the lateral ventricles (**c, d**)

may arise from other areas of the CNS, including brainstem, cerebellum, leptomeninges, and rarely spinal cord or structures of the eye. Our case is unusual in that the lesions are confined to the ventricular system, an extremely rare location. To date, less than 20 cases have been reported in the literature. Primary intraventricular CNS lymphoma may manifest as an isolated lesion (for example to the fourth ventricle) or as multiple mass lesions disseminated throughout the ventricular system [2–4]. The main differential diagnosis of intraventricular lesions includes tumor, neurosarcoidosis, and infectious diseases such as tuberculosis. Despite high morbidity and mortality, PCNSL is a potentially curable disease, so that the early diagnosis is essential to ensure the early treatment given that a delayed diagnosis may significantly affect survival and functional outcome. Biopsy with histological examination is the diagnostic procedure of choice.

In conclusion, this report calls for awareness of this uncommon radiological presentation of PCNSL which should always be considered in the differential diagnosis of isolated or multiple intraventricular mass lesions.

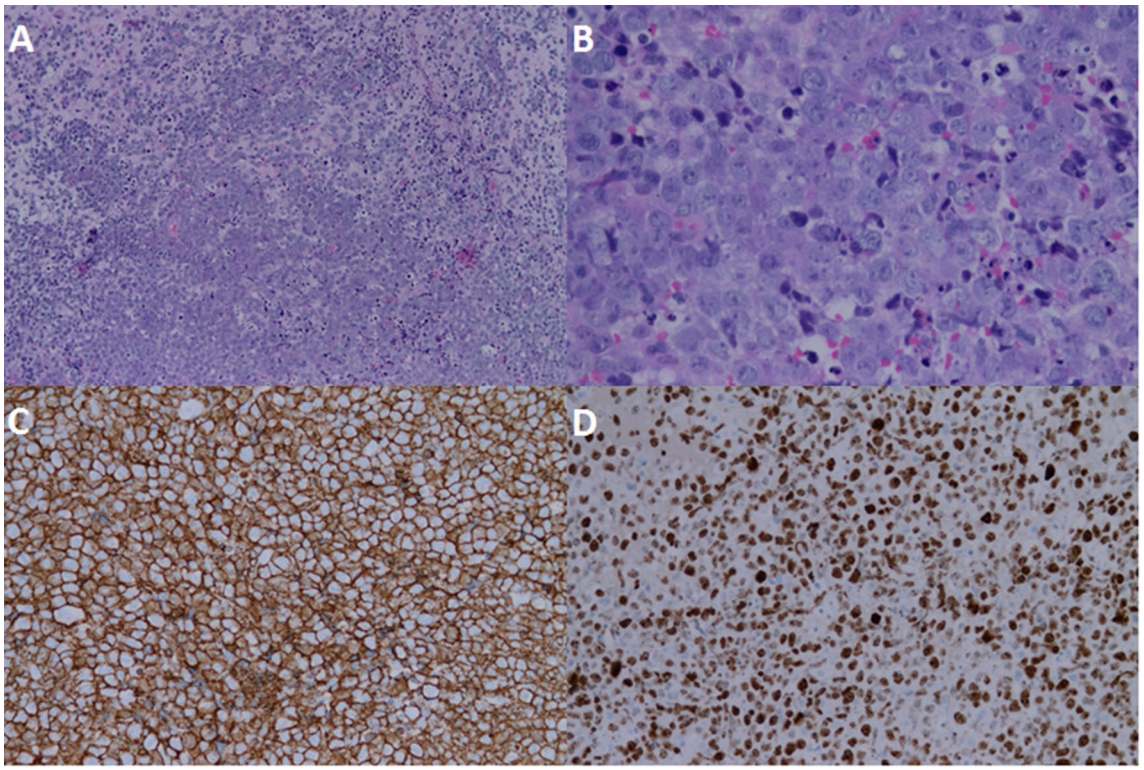

Fig. 2 Histopathological findings. The tissue biopsy from the temporal lesion showed diffuse infiltration of lymphocytes with irregular nuclei (**a**, hematoxylin–eosin, ×10; **b**, hematoxylin–eosin, ×40). Immunohistochemical studies show that neoplastic lymphocytes are positive for CD20 (**c**, ×20). The proliferation index (Ki-67) is greatly increased (**d**, ×20)

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Informed consent For this type of study, no informed consent is required.

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