A case report of Primary Central Nervous System Lymphoma

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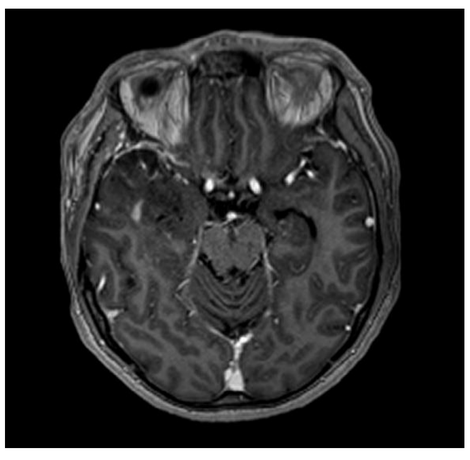
BACKGROUND: Acquired, progressive lesions involving both the corpus callosum and the periventricular white matter are found in association with a variety of conditions, such as demyelinating inflammatory diseases and neoplastic diseases. MRI imaging (T1-, gadolinium-enhanced T1- and T2-weighted imaging, fluid-attenuated inversion recovery, diffusion- and perfusion- weighted imaging, together with MRI spectroscopy) provides essential information for the differential diagnosis of this cases (1).

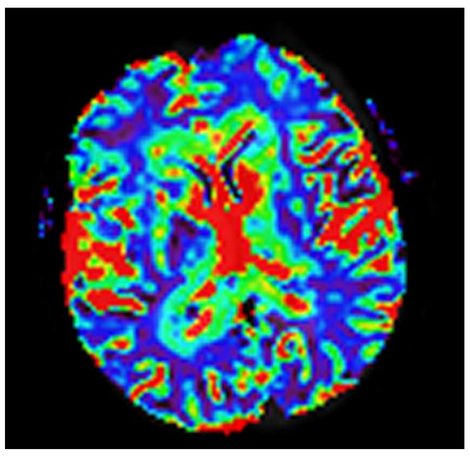
AIMS: We describe a case of Primary Central Nervous System Diffuse Large B Cell Lymphoma (PCNSDLBCL) presenting on brain MRI scans with extensive lesions involving the corpus callosum (CC) and the periventricular and periaqueductal areas.

MATERIALS AND METHODS: A 37-year-old man was referred for gradual onset of cognitive-motor slowing, headache and blurred vision. Brain MRI, whole-body PET, lumbar puncture, blood tests and brain biopsy were performed.

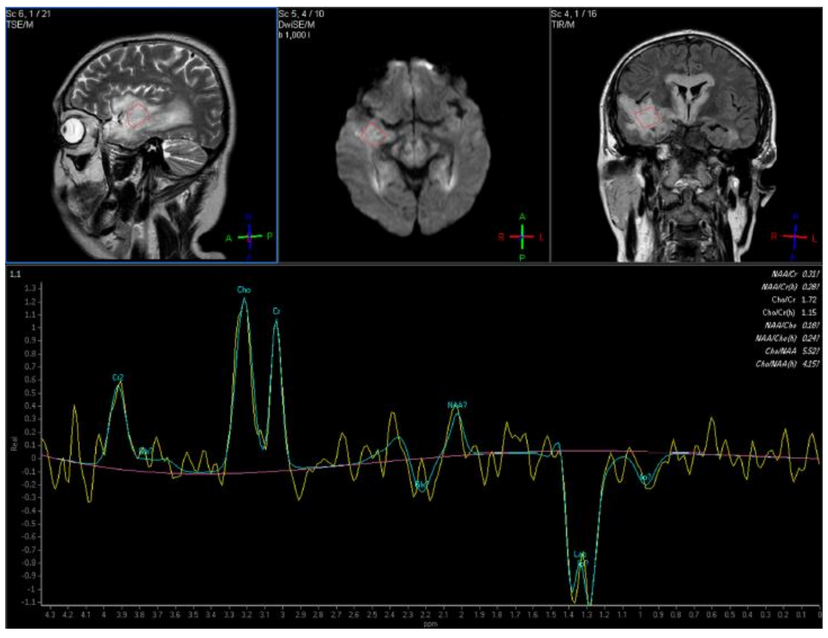
RESULTS: Brain MRI at onset revealed multiple enhancing and non-enhancing T2 hyperintense white matter lesions; CSF analysis showed the presence of oligoclonal bands (pattern 2), 10 mononuclear WBC/mmc and a protein concentration of 71 mg/dl; blood analysis detected high-titre MOG antibodies. He was treated with high- dose iv steroids with clinical improvement. Brain MRI at 6th month revealed radiological progression, although some previously detected lesions were disappeared; whole-body PET scan did not show areas of pathological hyperaccumulation; distribution of lymphocyte subsets in peripheral blood was normal. 4 months after discharge a severe worsening of cognitive and motor impairment occurred. MRI showed extensive T2 hyperintense white matter lesions outlining the fourth and third ventricle and the periacqueductal space, in contact with the ependymal surface, involving the corpus callosum in its entirety and extending to the temporal lobes and to the brainstem, with some enhancing nodular areas (fig. 1-7). The patient underwent stereotactic brain biopsy with histologic diagnosis of PCNSDLBCL (Ki-67 proliferative index: > 90%). He was treated with high-dose iv steroids. After a short period of stability, a new clinical worsening occurred. The patient was then admitted to the hematology unit and treated with high-dose iv methotrexate. He developed aplastic anemia and died of a septic shock after two weeks.



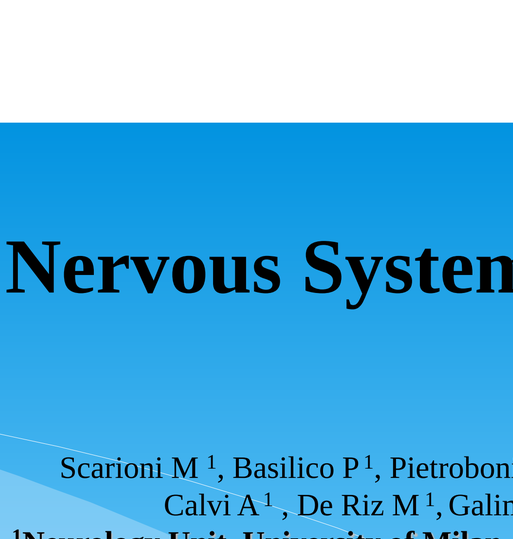
(Fig 1) Gd-enhanced T1-weighted MRI, axial section



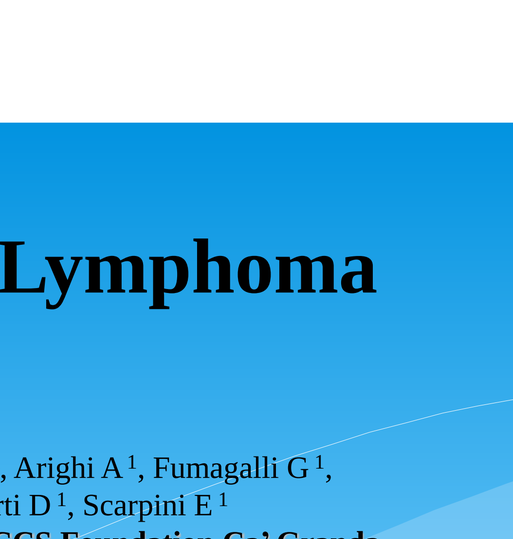
(Fig 2) Perfusion-weighted MRI, axial section



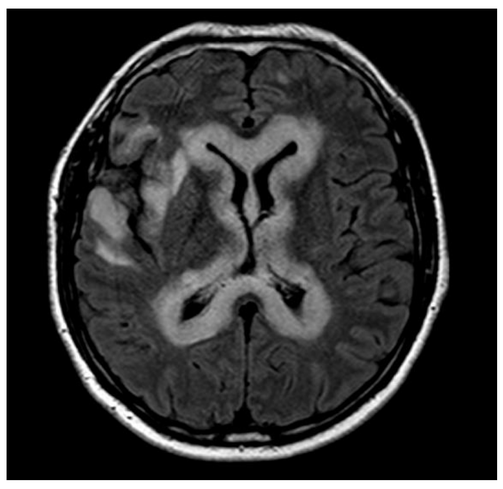
(Fig 3) MRI spectroscopy



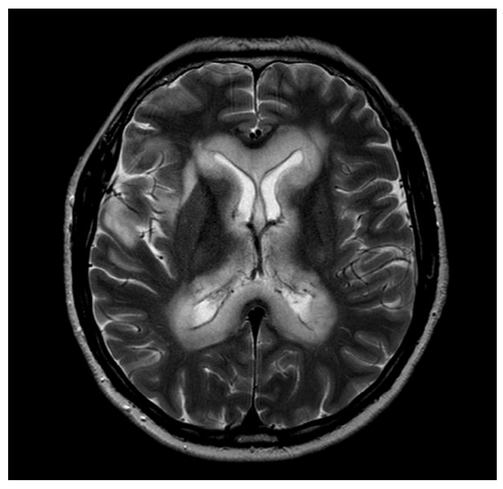
(Fig 4) T2-weighted MRI, sagittal section



(Fig 5) FLAIR, coronal section



(Fig 6) FLAIR, axial section



(Fig 7) T2-weighted MRI, axial section

DISCUSSION: Periventricular and corpus callosum white matter lesions improving with steroid therapy, in association with positivity for pattern 2 CSF oligoclonal bands, suggest multiple sclerosis. Besides, recent studies showed that brain MRI changes are extremely frequent in Neuromyelitis Optica and, when present, the periventricular white matter and the CC are the most frequently areas involved (1). CNS lymphomas may have characteristic imaging findings on MRI (such as their predilection for the periventricular regions, often in contact with ventricular surfaces); nevertheless, such findings cannot unequivocally distinguish CNS lymphoma from other brain lesions (2).

CONCLUSIONS: The diagnosis of CNS lymphoma is very difficult, especially in the absence of systemic involvement, because of clinical and radiological response to corticosteroids and lack of specific MRI findings that can rule out other pathologies. The majority of PCNSDLBCL are histologically diagnosed (3).

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