Primary Cerebellar Diffuse Large B-cell Lymphoma
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Section: Neuroradiology
Area of Interest: Neuroradiology brain
Imaging Technique: MR
Case Type: Clinical Cases
Authors: Abasin Tajmalzai MD1,2, Masoumeh Gity MD2, Sadegh Muradian MD2
Patient: 39 years, male

Clinical History:

A 39-year-old man presented to our hospital with progressive headache, vertigo, and visual impairment from one month without fever or other signs/symptoms of infections. The patient was not immune-compromised. A small left testicle mass was incidentally found on physical examination. Left radical orchiectomy was performed and histopathological examination revealed a testicular seminoma.

Imaging Findings:

Brain MRI with and without paramagnetic contrast agent was performed which revealed two infratentorial masses (26×27mm located in the middle and superior aspect of the cerebellum and another 35×45mm mass in the right cerebellar hemisphere) which were isointense to mildly hyperintense on T2WI and isointense to hypointense on T1WI images (relative to grey-matter) (Figs.1a,1b,1c). The mentioned masses were associated with mild surrounding oedema on FLAIR image causing mass effect on the fourth ventricle & resultant in mild hydrocephalus (Figs.2). After contrast injection, both masses intensely and homogeneously enhanced (Figs.3a,3b,3c,3d) and also showing restricted diffusion and low ADC values (Figs.4).

The above-mentioned imaging finding was typical and raises the suspension of cerebellar lymphoma, however, metastatic diseases (considering the patient known tumour) and other space-occupying lesions were in the differential diagnosis. Surgical biopsy for tissue diagnosis of the cerebellar mass was performed which confirmed the diagnosis of diffuse large B-cell lymphoma.

Discussion:

Background

Primary central nervous system lymphoma (PCNSL) which was first described by Bailey in 1929 as perivascular sarcoma is a rare, malignant non-Hodgkin lymphoma (NHL) that is confined to the CNS [1]. PCNSL is accounting for about 2-6% of all primary brain neoplasms and 1-2% of all NHLs and most cases are diffuse large B-cell lymphoma. PCNSL is an aggressive tumour that widely invades the brain parenchyma [2,3].

Immunocompromised individuals are considered most at risk of the disease, however, the incidence of PCNSL is increasing in immunocompetent populations [11]. PCNSL usually occurs as a solitary lesion, most commonly seen in the supratentorial brain and rarely in the cerebellum [4].
Clinical Perspective

The signs and symptoms of PCNSL are nonspecific and rapidly progressive. Patients usually present with personality changes, cognitive impairments, and focal neurological deficits. Headache, nausea, and vomiting, decreased alertness are less commonly observed at the time of diagnosis. A visible tumour on imaging is essential to raise the suspension of CNS lymphoma, which then can lead to an early histologic diagnose based on cytology of CSF or brain biopsy [5,6].

Imaging Perspective

Contrast-enhanced MRI is the most useful imaging modality [1,6]. It may reveal unifocal/multifocal, mainly supratentorial periventricular space-occupying lesions (SOLs) which usually show intense and homogeneous post-contrast enhancement [1,7]. Typically, the PCNSLs show significant diffusion restriction due to hypercellularity and also appear hyperintense on T2WI and isointense/hypointense on T1WI [1,7].

MR spectroscopy will show a large choline peak, reversed choline/creatinine ratio, and marked decreased N-acetyl aspartate. MR perfusion finding includes only a modest increase in relative rCBV and angiogenesis. The fluorodeoxyglucose PET/CT may show single or multiple high metabolic signals that exhibit SUVs elevation [8,12].

Imaging alone does not allow reliable discrimination from other malignant brain tumours or SOLs [5]. Histopathological confirmation of the diagnosis is essential which is typically achieved by stereotactic serial biopsies [9].

Outcome

CNS lymphoma is sensitive to radiation and chemotherapy, so the combination of chemotherapy and radiotherapy is the standard treatment. Surgical resection may also be performed for low-grade tumours. The median survival is weeks to months if treatment is only symptomatic [5,10].

Take-Home Message

PCNSL usually occurs as a solitary lesion, and most commonly seen in the supratentorial brain, but it can be multiple and infratentorial as in this case of an unusual form of PCNSL.

Written informed patient consent for publication has been obtained.

Differential Diagnosis List:  Primary cerebellar diffuse large B-cell lymphoma, CNS metastasis, Malignant glioma, Secondary CNS lymphoma

Final Diagnosis:  Primary cerebellar diffuse large B-cell lymphoma

References:


Haldorsen IS, et al. (2009). CT and MR imaging features of primary central nervous system lymphoma in Norway,


Description: Axial T2WI showing an isointense to mildly hyperintense mass (relative to grey-matter) in the middle and superior aspect of the cerebellum (arrows) Origin: Neuroradiology Section, Department of Radiology, Tehran University of Medical Sciences, Imam Khomeini Hospital Complex, Tehran, Iran, 2021
Description: Sagittal T2WI showing an isointense to mildly hyperintense mass (relative to grey-matter) in the middle and superior aspect of the cerebellum (arrows) Origin: Neuroradiology Section, Department of Radiology, Tehran University of Medical Sciences, Imam Khomeini Hospital Complex, Tehran, Iran, 2021
Description: Axial T1WI show a mildly hypointense mass (relative to grey-matter) in the middle and superior aspect of the cerebellum (arrows) Origin: Neuroradiology Section, Department of Radiology, Tehran University of Medical Sciences, Imam Khomeini Hospital Complex, Tehran, Iran, 2021
**Figure 2 a**

**Description:** Axial FLAIR image showing mild edema as a high signal in the cerebellum (white arrow) causing mass effect on the fourth ventricle (black arrow) **Origin:** Neuroradiology Section, Department of Radiology, Tehran University of Medical Sciences, Imam Khomeini Hospital Complex, Tehran, Iran, 2021
Description: Coronal T1 C+ MR shows two intensely and homogeneously enhancing masses in the middle and superior aspect of the cerebellum (white arrow) as well as in the right cerebellar hemisphere (black arrow) Origin: Neuroradiology Section, Department of Radiology, Tehran University of Medical Sciences, Imam Khomeini Hospital Complex, Tehran, Iran, 2021
Description: axial T1 C+ MR shows an intensely and homogeneously enhancing mass in the middle and superior aspect of the cerebellum **Origin:** Neuroradiology Section, Department of Radiology, Tehran University of Medical Sciences, Imam Khomeini Hospital Complex, Tehran, Iran, 2021
**Description:** axial T1 C+ MR shows an intensely and homogeneously enhancing mass in the right cerebellar hemisphere. **Origin:** Neuroradiology Section, Department of Radiology, Tehran University of Medical Sciences, Imam Khomeini Hospital Complex, Tehran, Iran, 2021
Description: Sagittal T1 C+ MR shows an intensely and homogeneously enhancing mass in the middle and superior aspect of the cerebellum. Origin: Neuroradiology Section, Department of Radiology, Tehran University of Medical Sciences, Imam Khomeini Hospital Complex, Tehran, Iran, 2021.
Figure 4

Description: Axial DWI and ADC map of the same patient shows the characteristic diffusion restriction within the tumor as a low ADC value (arrows) Origin: Neuroradiology Section, Department of Radiology, Tehran University of Medical Sciences, Imam Khomeini Hospital Complex, Tehran, Iran, 2021