Primary Intraventricular Lymphoma with Diffuse Leptomeningeal Spread at Presentation

Dear Editor,

Primary central nervous system lymphoma (PCNSL) is a rare tumour accounting for 4% of all brain neoplasms.[1] In immunocompetent patients, it tends to present as a large solitary hemispheric mass with periventricular lesions giving rise to ependymal seeding in 38% of the cases.[2] Leptomeningeal metastases of the PCNSL at presentation are rare, accounting for only 2.3% of the cases.[3] We present a rare case of primary intraventricular PCNSL with diffuse leptomeningeal spread at presentation. In this case study, we discuss the differential diagnoses and diagnostic approach to disseminated intraventricular masses.

Introduction

Primary CNS lymphoma carries poor prognosis with low curable rate, hence early detection and diagnosis are important in determining the survival rate.[1] It represents approximately 1% of Non-Hodgkin's lymphoma.[4] Secondary CNS lymphoma tends to present with leptomeningeal metastases in two-thirds of patients and intraparenchymal lesion in one-thirds of patients, while primary CNS lymphoma almost always demonstrates cerebral parenchymal lesion.[4] Our case was unique as the lesions were primarily intraventricular without intraparenchymal lesion and demonstrated diffuse leptomeningeal spread at presentation.

Case Report

A 77-year-old Chinese lady was brought to our Emergency Department with a history of 8 weeks of worsening headache, giddiness and slurred speech. A CT scan of the brain was performed to exclude cerebrovascular bleed or space occupying lesion. Non-enhanced and contrast enhanced CT brain revealed markedly enhancing diffuse hyperdense intraventricular lobulated masses with suprasellar extension and mild hydrocephalus.

Magnetic resonance imaging (MRI) of the brain showed diffuse intraventricular masses which were isointense to grey matter in T1W and T2W images (Fig. 1A). These masses demonstrated restricted diffusion on diffusion weighted imaging (DWI) and marked enhancement post gadolinium. The bulk of the disease was in the third ventricle which was totally obliterated with extension into the suprasellar recess (Fig. 1B). There was also enhancing nodular leptomeningeal thickening noted (Fig. 1C). An avidly enhancing extra-axial mass in the left cerebello-pontine angle and enhancing left foramen of Luschka lesion were also demonstrated further showing leptomeningeal spread of the disease (Fig. 1D).

Biopsy of the intraventricular masses was done through a left frontal craniotomy and histopathology revealed diffuse large B cell Non-Hodgkin's lymphoma. The cells were

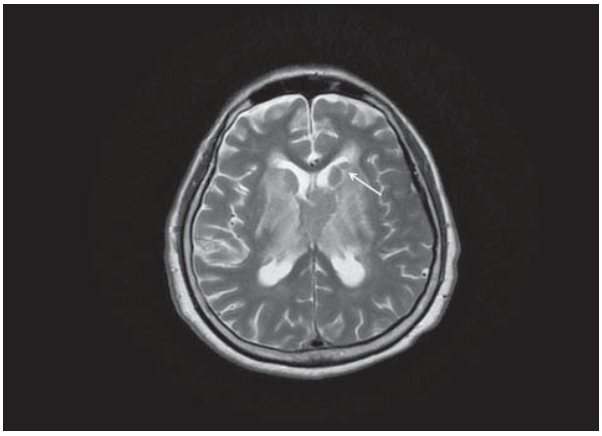

Fig. 1A. T2W axial MR image showing diffuse iso-intense intraventricular masses with CSF cleft (arrow).

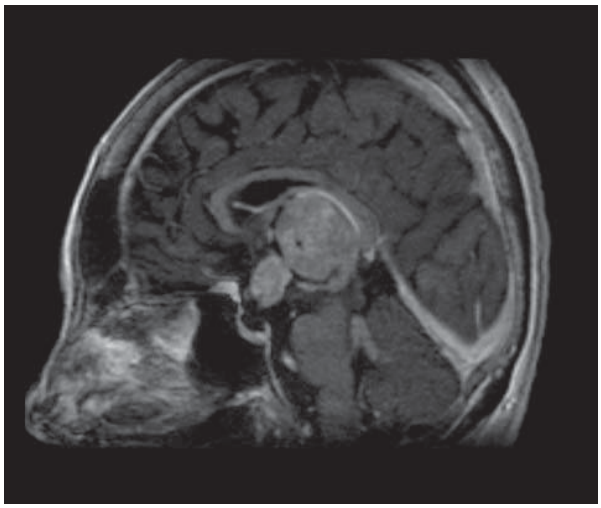

Fig. 1B. Post-gadolinium sagittal MR image showing heterogenously enhancing mass occupying the third ventricle extending to the suprasellar recess and outlining the 4th ventricle.

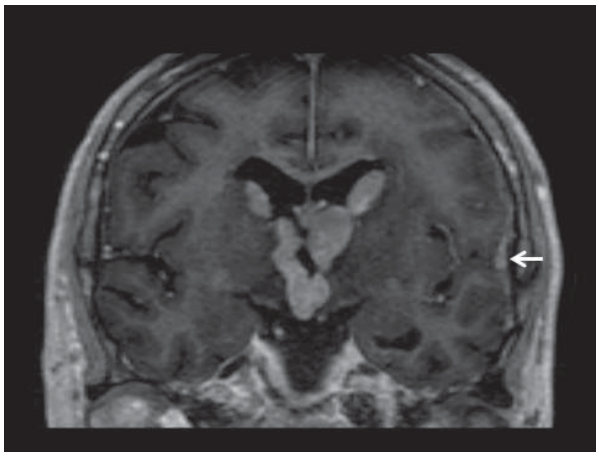

Fig. 1C. Post-gadolinium coronal MR image showing intraventricular masses extending through Foramen of Monro bilaterally into the third ventricle as well as leptomeningeal thickening in the left temporo-parietal region (arrow).

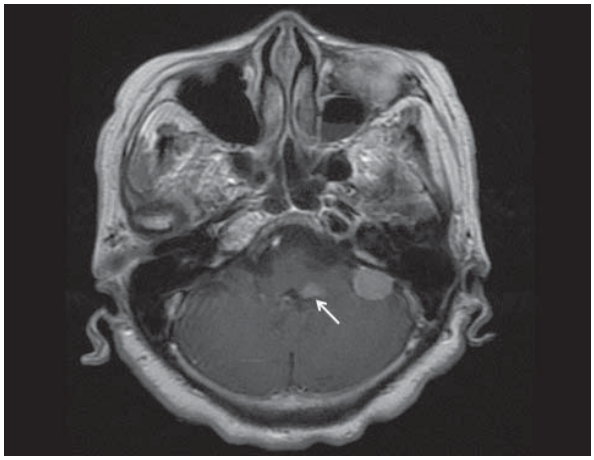

Fig. 1D. MR post-gadolinium axial image of the left CP angle mass and leptomeningeal seeding to the left foramen of Luschka (arrow).

positive for CD20, CD79a, Mum1 but negative for Cd3 and CD10. Lumbar puncture however, was not performed. As there was no involvement of the rest of the body, a diagnosis of primary CNS lymphoma was made.

The patient and her family then opted for traditional treatment. The patient died in her home a few weeks after the discharge.

Discussion

PCNSL may arise from different parts of the brain parenchyma with the periventricular deep white matter being the most common.[2] In immunocompetent patients, primary CNS lymphoma tends to present as a solitary intraparenchymal mass.[2] Typical lymphomatous lesions are hyperdense on unenhanced CT (70%) which was thought to be due to highly packed abnormal cells.[2] On MR imaging, majority of these lesions showed intermediate to low signal intensity on T1-weighted images and either isointense or hypointense signal relative to gray matter on T2-weighted images.[2] PCNSL lesion typically showed restricted diffusion, thus appearing hyperintense on DWI and hypointense on apparent diffusion coefficient (ADC) imaging due to its high cellularity.[4] On MR spectroscopy, PCNSL may show elevated lipid peaks with high choline/creatine ratio.[4] Despite having the characteristic MR imaging findings, none of this could unequivocally differentiate CNS lymphoma from other brain lesions.

Differential diagnoses for multiple intraventricular masses in our patient's age group include metastases and very rarely sarcoidosis. Multiple ependymomas may fit the description; however, they are less common in adults compared to the paediatric population.[5] Most of the supratentorial ependymomas (70%) are extra-ventricular in origin and those arising within the brain parenchyma have a propensity for cyst formation.[5] Secondary CNS involvement in systemic lymphoma is more commonly present with leptomeningeal spread.[4] Diffuse leptomeningeal carcinomatosis most commonly arise from adenocarcinoma, although any systemic cancer can also do so.[6] Symptomatic CNS involvement in sarcoidosis has been found in only 5% of cases and isolated CNS involvement which is rare, is estimated to occur in less than 1% of the patients.[7]

Metabolic imaging using radioisotopes such as positron emission tomography (PET), PET/CT fusion imaging and single photon emission computed tomography (SPECT) have been used in the evaluation of lymphoma. Hypermetabolic state of the lymphoma cells will show very high uptake of 18-fluorodeoxyglucose (FDG) on PET and is documented to show more pronounced metabolic activity than metastases and high grade gliomas, making FDG-PET more suitable for early therapeutic assessment.[4] PET performed using 11-C Methionine (Methionine PET) also shows very high uptakes in CNS lymphoma. However, the area of high uptake is usually larger than the area of enhancement on CT or MR, reflecting a larger tumour infiltration as compared to CT or MR imaging.[4] Methionine PET may provide better tumour volume delineation for therapeutic evaluation.[4]

Prognosis of PCNSL is poor for majority of patients, however, a substantial minority, representing 20% to 30% of cases can be cured with adequate treatment.[8] With radiotherapy alone, the median survival rate is usually short-lived ranging from 10 to 18 months while chemotherapy alone has been shown to prolong the survival rate to up to 36 months.[9] Consolidation after chemotherapy represents the best role for radiotherapy, however, it is associated with severe neurotoxicity and 30% mortality.[9] Age and performance status are the main independent prognostic

factors and treatment should be tailored accordingly for better treatment outcome.[8]

Conclusion

Central nervous system lymphoma is known to present in many different forms. As described in our case report, lymphoma should be considered as one of the differentials of diffuse intraventricular masses.

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