

IMAGING HIGHLIGHTS

Nodular nerves and lumpy leptomeninges – neuroimaging in primary leptomeningeal lymphoma

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Primary leptomeningeal lymphoma (PLML) is a rare entity, representing 7% of all cases of primary central nervous system lymphoma and approximately 0.1% of primary central nervous system tumours.¹⁻⁴ Patients can present with symptoms of raised intracranial pressure, or cranial or spinal polyradiculopathies.¹⁻³ Even with extensive investigations, diagnosis in up to a third of patients can only be confirmed with meningeal biopsy. In this *Imaging Highlights*, we describe a patient who presented with multiple cranial nerve palsies with corresponding MRI nerve enhancement, and subsequent meningeal biopsy proven lymphoma.

CASE REPORT

A previously healthy 53-year-old man presented with acute onset of headache on a background of chronic giddiness of 9 months duration. Examination revealed a left III nerve palsy, a right relative afferent pupillary defect and an inferior temporal scotoma in the right visual field, with no other neurological deficits. Brain MRI revealed multiple enhancing nodules in the pontine cistern along several cranial nerves (Figures 1 and 2) and patchy nodular leptomeningeal enhancement (Figure 3). Neoplastic meningitis was considered likely. Differential diagnoses included sarcoidosis and Lyme disease⁵; the latter was deemed unlikely given epidemiological context and lack of travel history.

A review of systemic history and a complete physical examination did not provide clues to the aetiology or to a possible primary tumour; in particular, no travel history, rash, lymphadenopathy or hepatosplenomegaly was found. CSF showed lymphocytic pleocytosis; there were no radiological features of sarcoidosis in the HRCT thorax.

At this stage, neoplastic meningitis was considered more likely. However repeated CSF cytology and flow cytometry were negative. A whole-body CT and PET also did not demonstrate any primary tumour and spine MRI showed no further CNS lesions. Ultrasound testes revealed a diffusely hypoechoic left testis suspicious for

lymphoma and an orchidectomy was performed; histology, however, demonstrated no evidence of malignancy.

Given the MRI appearance of nodular leptomeningeal disease, negative systemic malignancy and sarcoidosis workup, we proceeded with a meningeal biopsy, which was taken from the right cerebellar region. This revealed diffuse large B-cell lymphoma. As there was no evidence of systemic or brain parenchymal involvement, a diagnosis of PLML was made.

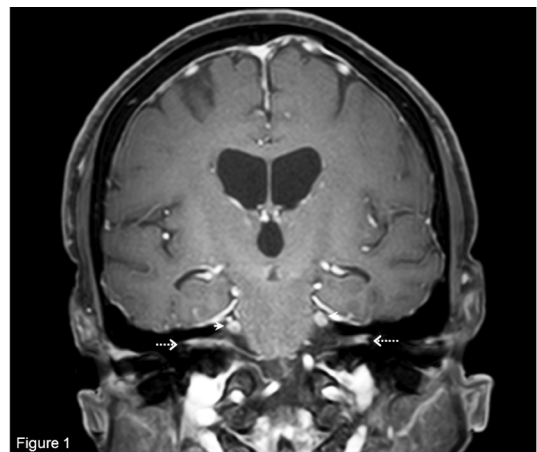


Figure 1. Coronal fat saturated post-gadolinium T1 weighted image: Nodular thickening and enhancement of bilateral V (arrowheads) and VII/VIII nerves (dotted arrows).

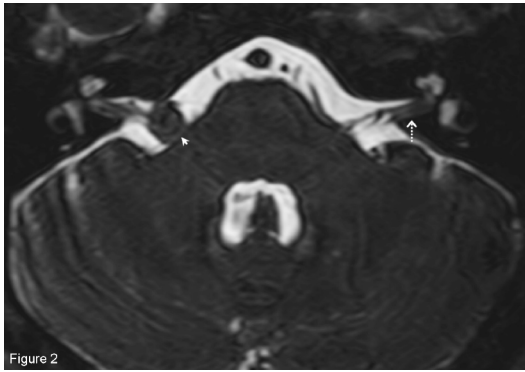


Figure 2. High resolution axial T2 weighted image: Nodule at right cerebellopontine angle (arrowhead) related to VII/VIII nerves; Thickening of left VII/VIII nerves (dotted arrows) within internal acoustic meatus.

He was referred to medical oncology for further management, and received chemotherapy, including intravenous R-CHOP (rituximab, cyclophosphamide, vincristine, doxorubicin and prednisolone) and intrathecal methotrexate, followed by whole brain radiation therapy.

Interval brain imaging showed resolution of leptomeningeal enhancement. However, he relapsed with brain and spinal metastases, and failed to respond to subsequent treatment. He passed away within one year of disease presentation.

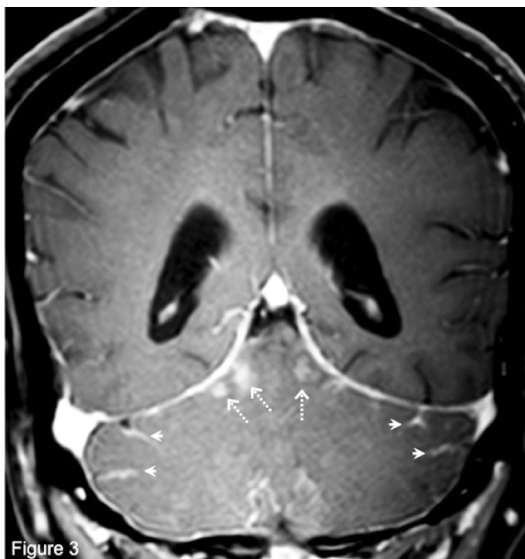


Figure 3. Coronal fat saturated post-gadolinium T1 weighted image: Nodular leptomeningeal enhancement (dotted arrows); Other areas of leptomeningeal enhancement (arrowheads).

DISCUSSION

Primary leptomeningeal lymphoma (PLML) is a rare entity.^{1,2} The majority of existing literature consists of case reports and small case series.² The largest international case series reports 48 patients over a period of ten years.¹ Our patient reflects the clinical and radiological findings, as well as the diagnostic challenges associated with this disease.

Patients with PLML present with multifocal symptoms;¹ headache and cranial neuropathies are common findings.^{1,2} Our patient’s initial findings are consistent with these descriptions.

Most patients (81%) demonstrate abnormalities in their brain and/or spine imaging.¹ Our patient had the striking MRI finding of nodular enhancement of both cranial nerves and leptomeninges; it is noteworthy that 3 of 9 patients in the second largest PLML series had similar nodularity of either cranial and/or spinal nerves.² However such MRI findings are non-specific; differentials include sarcoidosis and Lyme disease.⁵

Abnormal CSF findings in PLML include leukocytosis, raised protein concentration and hypoglycorrhachia.¹ Negative CSF cytology, flow cytometry and gene rearrangement studies were seen in up to 33% of patients; these patients eventually required a meningeal biopsy for diagnosis.¹ Our patient’s CSF cytology was negative, hence meningeal biopsy was deemed necessary, allowing us to clinch the diagnosis of PLML.

Our case report highlights three points. Firstly, PLML is a diagnostic challenge. Secondly, nodular enhancement of cranial/spinal nerves and leptomeninges in the MRI, as demonstrated in our patient, can be a useful radiological clue to the diagnosis of PLML. Lastly, these MRI findings, along with non-diagnostic systemic and CSF workup, should prompt the neurologist to consider the uncommon diagnosis of PLML and to proceed with a meningeal biopsy.

ACKNOWLEDGEMENTS

None

DISCLOSURE

Dr Nigel Choon Kiat Tan has received research funding from the National Medical Research Council, Singapore (NMRC/0752/2003 & NMRC/0998/2005), and holds an unrestricted educational grant from UCB Pharma. He holds stock in Abbott Laboratories, Novartis, Pfizer,

Johnson & Johnson, Stryker, and Proctor & Gamble.

Conflict of interests: None

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