

IMAGING HIGHLIGHTS

Central nervous system intravascular lymphoma in a Malaysian

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Intravascular lymphoma (IVL) is a rare subtype of extranodal diffuse large cell lymphoma, characterized by intravascular proliferation of B or T lymphocytes within small blood vessels; which may lead to occlusive symptoms, its neurological involvement has been said to be uncommon among Asians.¹ We describe a Malaysian with central nervous system IVL, to demonstrate that IVL is an important differential diagnosis in diffuse brain pathology also among Asians.

CASE REPORT

A 48 year-old Chinese Malaysian woman presented with seizure, following two weeks history of forgetfulness, speech disturbance and difficulty in walking. Clinical examination revealed expressive dysphasia, weakness both lower limbs and extensor plantar responses. Erythrocyte sedimentation rate and C-reactive protein values were raised at 32 mm/hr and 1.3 mg/dL respectively. Serum lactate dehydrogenase (LDH) was elevated (565 IU/L). The rest of blood investigations were normal.

Initial Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) showed diffuse multiple asymmetrical hyperintense white matter lesions on T2 weighted and FLAIR sequences on MR imaging (Figure 1a and 1b). Some of these lesions had mild contrast enhancement after gadolinium administration (Figure 1c) and showed hyperintensity on diffusion-weighted and ADC images (T2 shine through effects). Initial diagnosis of acute demyelinating encephalomyelitis (ADEM) was made and treatment with methylprednisolone resulted in clinical improvement.

A month later, she was readmitted with sudden onset of left lower limb weakness, urinary retention and worsening of neurological symptoms. MRI of the spine was normal. CT of the thorax and abdomen revealed enlargement of the adrenal gland (Figure 1f). A repeat brain

MRI showed progression of the white matter lesion with new areas of subcortical white matter hyperintensity (Figure 2a), areas of cortical laminar necrosis and gliosis at previous lesions sites (Figure 2a and 2b). The constellation of worsening clinical manifestations, widespread radiological abnormalities in brain and adrenal gland, and raised LDH, raised the suspicion of intravascular lymphoma.

Imaged guided biopsy of the brain was performed. The procedure was uneventful. The histology showed proliferation of large B lymphoid cells within vascular lumina with CD20 histochemical staining positivity (Figure 4). There was no B-cell lymphomatous deposition found in the extravascular regions of the brain. She was treated with chemotherapy comprising cyclophosphamide, doxorubicin, vincristine and prednisolone (CHOP) with rituximab and intrathecal methotrexate. Follow up MRI using 3.0 Tesla GE scanner showed reduction in the size of the hyperintense T2 lesions and the intensity of enhancement. The patient recovered neurologically. Autologous transplantation was subsequently performed. The patient remained well on last follow up two years after the initial diagnosis.

DISCUSSION

IVL is a disease that affects individuals of the 5th to 7th decade with no gender preponderance. Whereas

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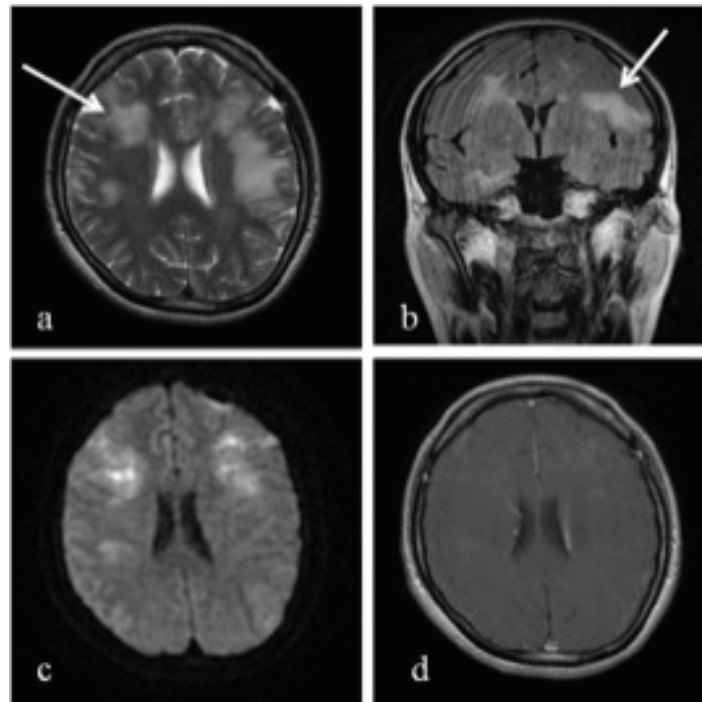


Figure 1: MRI of the brain at first presentation showing diffuse bilateral asymmetrical predominantly subcortical hyperintense white matter lesions in T2WI (a) and FLAIR (b) with hyperintensity on DWI (c). Involvement of the grey-white matter region is seen in some of the lesion (arrows). (d) Axial T1WI post contrast showed subtle enhancement.

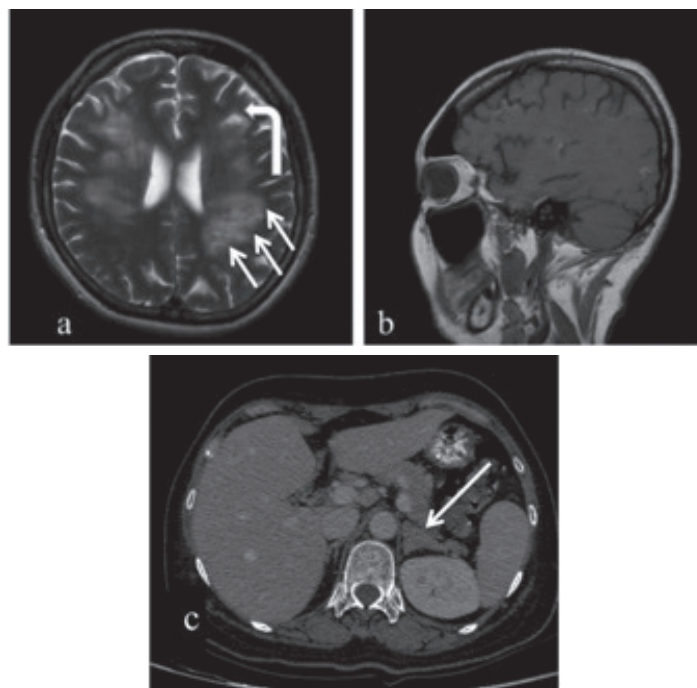


Figure 2a, T2WI MRI of the brain in axial, following readmission showing new subcortical white matter lesions (arrows). The previously seen subcortical white matter lesion (curved arrow) now shows infarct evolution with gliosis and cystic change. Figure 2b in sagittal T1WI showing evidence of gyriform hyperintense signal (arrows) consistent with cortical laminar necrosis. Figure 2c, CECT showing left adrenal enlargement.

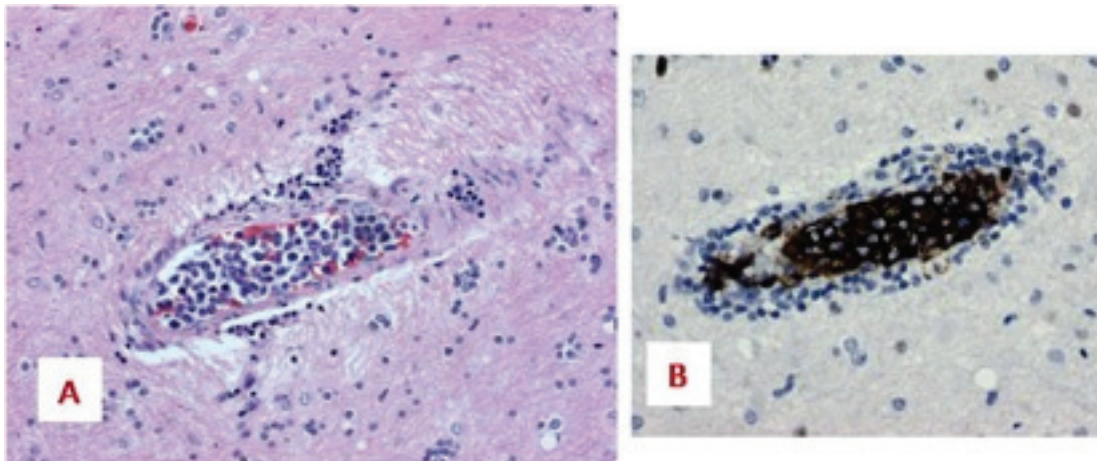


Figure 3A, Photomicrograph of brain biopsy showing blood vessels filled with immature lymphoid cells. H&E X 100. Figure 3B, Immunohistochemistry showed these cells to express intense positivity for CD20. IHC for CD20 X 100.

brain and skin involvement is more common in the Western populations, haemophagocytic syndrome is mainly found in Asia with similar immunohistochemical characteristic. The Asian patients usually present with overt splenomegaly and/or hepatomegaly with no associated lymphadenopathy, and only rarely with neurological and skin involvement.¹

A myriad of neurological presentations may be observed in IVL, manifesting as stroke and transient ischaemic attack; dementia, subacute encephalopathy or myelopathy.²⁻⁴ The main underlying pathological mechanism attributing to these symptoms is ischemia of the neural tissue secondary to occlusions of the small vessels by lymphomatous cells.¹

In IVL, the most commonly reported imaging abnormalities were hyperintensities on T2 weighted imaging, multifocal, subcortical in location and scattered throughout the brain and spinal cord.⁵⁻⁸ The main differential diagnosis are thromboembolic disease, vasculitis and ADEM. Contrast enhancement is variable in IVL, and may involve the parenchyma as seen in our patient. Meningeal, leptomeningeal, ring like enhancement may also be seen⁵⁻⁸. The elevated LDH level in our patient helped to alert the possibility of IVL. Raised LDH level has been observed in previous reports^{4,9,10}, though this is non specific. The radiological survey to detect other sites of involvement such as the adrenal gland in our patient, may also help in the differential diagnosis. Clinical awareness of this rare but potentially treatable disease is crucial as currently, the combination of chemotherapy has resulted in good treatment outcome.⁴

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