Case Report

DOI: 10.6966/EDMJ.202209_9(3).0005

A Diagnostic Dilemma: A Case Report of Tumefactive Multiple Sclerosis

Pao-Li Hsiao[1], Hung-Chang Kuo[2,]*

We present a case of central nervous system inflammatory lesion presented with tumor-like radiographic feature. Under the diagnosis of tumefactive multiple sclerosis, the patient received steroid therapy with evident clinical improvement alongside image confirmation for lesion regression. The case report seeks to highlight such diagnostic dilemma with a possibility of misdiagnosis for brain tumor. The physician therefore must integrate clinical picture with laboratory investigations and imaging techniques to avoid unnecessary surgical procedures.

Key words: tumefactive, multiple sclerosis, tumor-like, inflammatory

Case Report

A 39-year-old female with no prior medical illness visited our neurology clinic to seek medical advice for the chief complain of progressive left lower limb weakness and numbness over the course of 1 week. There was no recent history of trauma, fever, visual symptoms, neck rigidity, menstrual changes, sphincter problems, nor significant body weight changes.

Neurologic examination revealed left side upper and lower limb weakness, with muscle power of 4 and 4- respectively, as well as left side below-knee paresthesia. Other examinations were normal, including cranial nerves, muscle tone, preserved symmetric deep tendon reflexes and normal coordination.

Initial laboratory investigations, including complete blood count, metabolic profile, coagulation panel were within normal limits. Due to progressive symptom and clinical suspicion of brain organic lesion, Magnetic Resonance Imaging (MRI) with contrast enhancement revealed a 2 cm tumor-like lesion at right frontal-temporal region (Fig. 1A). An initial plan to undergo surgical biopsy was therefore suggested after discussing with neurosurgeon. However, upon closer examination, the lesion has a closed ring-enhancing pattern with perilesional edema and without diffusion restriction (Fig. 1B – 1D). There are also multiple small peri-ventricular subcortical and infratentorial white matter lesions, which appeared hypointense under T1 sequence, and hyperintense under both T2 and fluid-attenuated inversion recovery (FLAIR) sequence (Fig. 2). Some of the peri-ventricular lesions is perpendicularly oriented and consistent with the classic Dawson-finger sign (Fig. 2B). There were no cervical cord lesions identified.

From the [1]Department of Neurology, E-Da Hospital; [2]Department of Neurology, E-Da Cancer Hospital, Kaohsiung, Taiwan.

Received: March 15, 2021 Accepted: May 10, 2021

* Address reprint request and correspondence to: Hung-Chang Kuo, Department of Neurology, E-Da Cancer Hospital, No.21, Yida Road, Jiaosu Village, Yanchao District, Kaohsiung City 82445, Taiwan.
Tel: +886-7-615-0011 ext. 251027, E-mail: ed100256@edah.org.tw

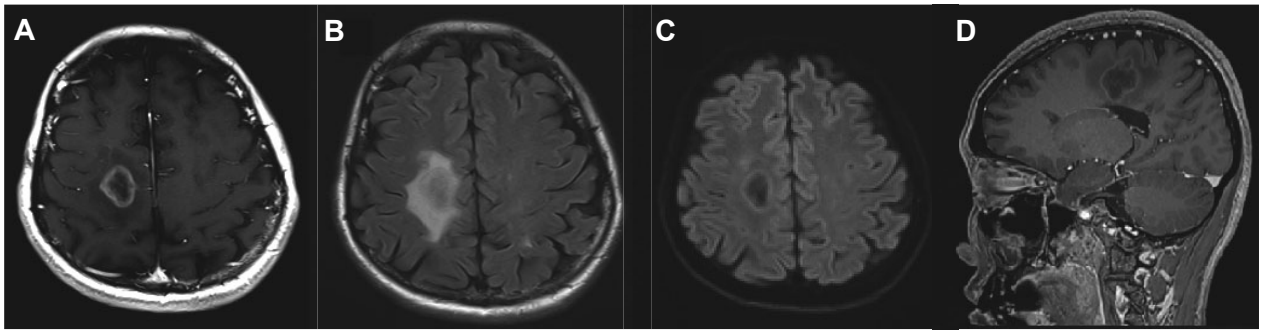

Fig. 1 Right temporal tumor-like lesion: (A) A 2 cm ring enhancing lesion under postcontrast T1-weighted axial MRI. (B) T2-FLAIR axial MRI showing perilesional edema. (C) Diffusion-weighted axial sequence showing no diffusion restriction. (D) Ring enhancing lesion under postcontrast T1-weighted sagittal MRI.

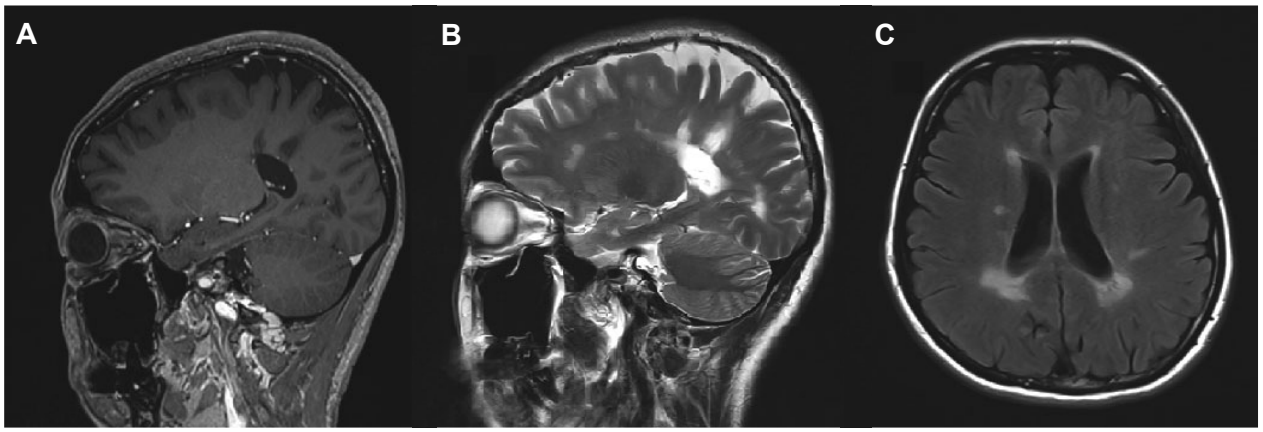

Fig. 2 Dawson-finger sign of Multiple Sclerosis: Multiple subcortical and periventricular lesion arranged in a perpendicular fashion. (A) Hypointense and non-enhancing lesion under T1-weighted sagittal MRI. (B) Residual focal edema under T2-FLAIR axial MRI. (C) Hyperintense under T2-FLAIR axial MRI.

Potential differential diagnosis included metastatic brain tumors or tumefactive multiple sclerosis. Follow up laboratory and imaging survey for evidence of underlying malignancy are all negative (including chest X-ray, tumor markers, computed tomography (CT) of abdomen and pelvis, sonography of breast with biopsy and Positron emission tomography (PET) scan). The cerebrospinal fluid analysis showed unspecific elevation of immunoglobulin G (IgG) index without positive oligoclonal band. The main tumor-like lesion appears to be hypometabolic on F18-FDG PET scan.

The patient received intravenous corticosteroid therapy with dexamethasone 5 mg every 8 hours for 12 days, with marked improvement of her left upper and lower limb weakness. She was discharged with oral steroid tapering over 2 weeks. At one month follow up, clinical examination showed only residual numbness over left lower limb, with improvement of Expanded Disability Status Scale (EDSS) score from 2 to 1 after treatment. Repeated brain MRI showed partially regressive change of the right parietal tumor-like lesion with residual perifocal edema (Fig. 3).

Discussion

Tumefactive multiple sclerosis is an atypical manifestation of the central nervous system (CNS) inflammatory demyelinating disease. The pseudo-tumoral reference in naming suggest it is often mistaken for neoplasm due to overlapping radiologic features. The uncertainty during diagnostic process therefore may result in unnecessary brain biopsy, or delay in appropriate treatment.

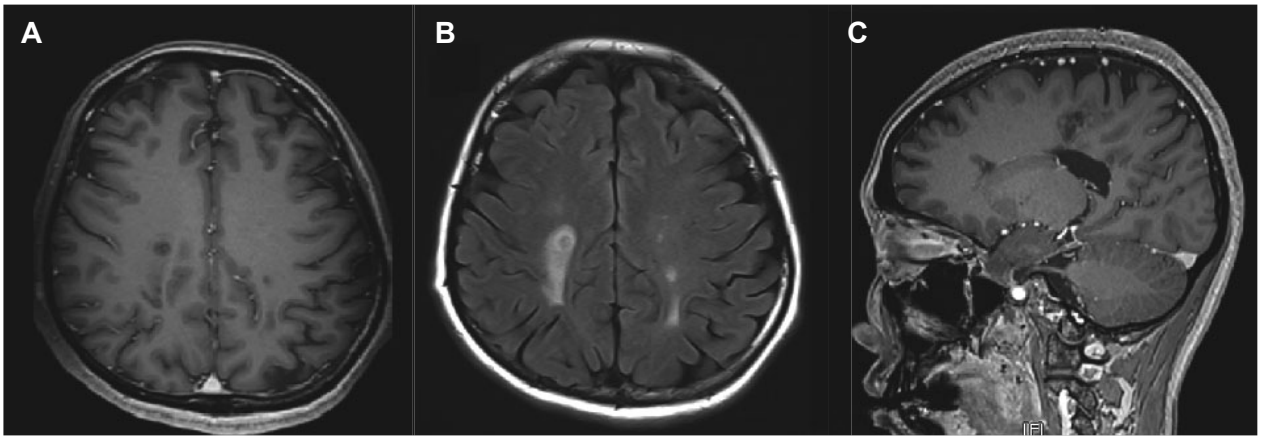

Fig. 3 Post-treatment MRI: (A) Regressive change under T1-weighted axial MRI. (B) Residual focal edema under T2-FLAIR axial MRI. (C) No contrast enhancement under T1-weighted sagittal MRI.

The demographic prevalence of such cases is rare, but may be as high as 8% of all multiple sclerosis patients as estimated by large multicenter studies. The number is slightly lower in Taiwan, at around 6.3% as estimated by Taipei Veterans General Hospital with their study which enrolled 190 patients from 1985 to 2010.[1]

Patients with either Tumefactive, or multiple sclerosis in general, may present with a wide variety of clinical symptoms depending on the anatomical location of demyelinating lesion. Although physicians may make an educated guess through differences in the clinical course of disease progression, the presentation itself is often nonspecific for differentiating the etiology, whether being neoplastic or demyelinating in nature.[2]

Radiographically, studies have shown that certain features may help in distinguish between tumor and demyelinating tumefactive lesions. The currently recognized definition of a tumefactive demyelinating lesion under MRI is a T2-weighted sequence with a size of equal or larger then 2 cm.[3] Other characteristics supportive for demyelinating lesion include minimal surrounding edema, open ring enhancement and other associated lesions showing more typical Dawson finger sign.[4] However, there are also reviews suggesting peripheral diffusion restriction, if evolve dynamically during imaging follow up, is more common in demyelinating disease compared to tumors or abscesses.[5] FDG-PET scan can sometimes be helpful, as brain tumors, especially if malignant, is likely to show increased glucose metabolism, which is opposite to our case.[6]

The next step in diagnosis often requires laboratory investigations, including metabolic profile, coagulation panel, autoimmune profile and tumor marker survey. Cerebrospinal fluid examination is essential as the presence of oligoclonal bands with the absence of atypical cells favors inflammatory process over neoplastic etiology.[7]

Excluding tumor etiology, there are still differential diagnosis regarding demyelination, including acute disseminated encephalomyelitis (ADEM), neuromyelitis optica spectrum disorders (NMOSD) and myelin oligodendrocyte glycoprotein antibody associated disease (MOGAD). Some experts suggested that all patients diagnosed with tumefactive demyelinating lesion should be tested for both aquaporin 4-IgG and MOG-IgG, as treatment of each conditions may differ.[8]

In conclusion, albeit challenging, the definitive diagnosis between demyelinating or neoplastic lesion is crucial to establish an appropriate treatment or investigation plans. If the lesion is highly suspicious for neoplasm, brain

biopsy might be unavoidable for diagnostic confirmation. Therefore, the benefit of surgical biopsy with risk for comorbidity weighted against a trial of pulse steroid and follow-up imaging, with potential risk for delaying neoplasm treatment,[9] must be thoroughly discussed between first-line neurologists with neuroradiologists and neurosurgeons, in order to establish the best approach for the patient's prognosis and wellbeing.

References

1. Kuan YC, Wang KC, Yuan WH, et al: Tumefactive multiple sclerosis in Taiwan. PLoS One 2013;8:e69919. doi: 10.1371/journal.pone.0069919.
2. Hayashi T, Kumabe T, Jokura H, et al: Inflammatory demyelinating disease mimicking malignant glioma. J Nucl Med 2003;44:565-9.
3. Lucchinetti CF, Gavrilova RH, Metz I, et al: Clinical and radiographic spectrum of pathologically confirmed tumefactive multiple sclerosis. Brain 2008;131:1759-75. doi: 10.1093/brain/awn098.
4. Hu W, Lucchinetti CF: The pathological spectrum of CNS inflammatory demyelinating diseases. Semin Immunopathol 2009;31:439-53. doi: 10.1007/s00281-009-0178-z.
5. Abou Zeid N, Pirko I, Erickson B, et al: Diffusion-weighted imaging characteristics of biopsy-proven demyelinating brain lesions. Neurology 2012;78:1655-62. doi: 10.1212/WNL.0b013e3182574f66.
6. Wong TZ, van der Westhuizen GJ, Coleman RE: Positron emission tomography imaging of brain tumors. Neuroimaging Clin N Am 2002;12:615-26. doi: 10.1016/s1052-5149(02)00033-3.
7. Hardy TA, Chataway J: Tumefactive demyelination: an approach to diagnosis and management. J Neurol Neurosurg Psychiatry 2013;84:1047-53. doi: 10.1136/jnnp-2012-304498.
8. Ramanathan S, Prelog K, Barnes EH, et al: Radiological differentiation of optic neuritis with myelin oligodendrocyte glycoprotein antibodies, aquaporin-4 antibodies, and multiple sclerosis. Mult Scler 2016;22:470-82. doi:10.1177/1352458515593406.
9. Siri A, Carra-Dalliere C, Ayrignac X, et al: Isolated tumefactive demyelinating lesions: diagnosis and long-term evolution of 16 patients in a multicentric study. J Neurol 2015;262:1637-45. doi: 10.1007/s00415-015-7758-8.