Case Report

Tumor-like Manifestation, Uncommon Form of Multiple Sclerosis: Report of A Patient

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Multiple Sclerosis (MS) is a demyelinating disease of the central nervous system, characterised by focal neurological dysfunction with relapsing and remitting course. Acute widespread or tumor-like manifestation is one of the rare clinical variants and has poor prognosis. Here, the authors report a 36-year-old man who presented with left hemifacial and left hemibody anesthesia for one month. His symptoms gradually progressed. MRI brain showed multiple large hypersignal intensity lesions in both right and left frontoparietal lobes, surrounding with brain edema. Brain biopsy showed perivenous infiltration of mature lymphocyte with demyelination. He was dramatically improved with high dose steroid. However, he later developed transverse myelitis syndrome. The second MRI showed new foci in both sides of splenium of corpus callosum and T9-10 spinal cord. The findings were compatible with an unusual form of multiple sclerosis that is rarely seen.

Keywords: Multiple sclerosis, Tumor-like manifestation

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Case Report

A 36-year-old man from Pathumthani came to our hospital with left hemifacial and left hemibody anesthesia for one month. He also complained of headache, fatigue and weight loss. He did not have any eye symptoms. Neurological examination revealed facial palsy (upper motor neuron type), impairment of pain and temperature on left side of his face and body. The
motor power of all extremities and cerebellar function were normal. Laboratory data that included complete blood count, blood chemistry and anti-HIV test, was unremarkable. Computed tomography of the brain showed patchy low density lesions of about 1 x 1cm in white matter of both frontal lobes with partial rim enhancement in contrast study. There was mild degree of generalized brain edema (Fig. 1).

Two days later, his symptoms worsened. He developed left hemiparesis and had difficulty in walking. He was admitted to the hospital. Brain MRI was done. The lesions showed multiple irregular ring enhancements on post Gd-T1 weighted image, hypointense T1 weighted and hypersignal intensity T2-weighted lesions in both frontoparietal lobes surrounding with brain edema. Lesions at right frontoparietal lobe involved genu of right corpus callosum. There were some lesions in his left cerebellar peduncle and left side of pons (Fig. 2-4). He had undergone extensive investigations including chest x-ray, ultrasound whole abdomen, bone marrow biopsy and tumor markers to rule out metastatic tumor and hematologic malignancy. All of them were normal. He later underwent brain biopsy to rule out metastatic lesion or brain tumor. The result showed inflammatory lesion and periventricular infiltration of mature lymphocyte with demyelination. After pulse methylprednisolone therapy, he was dramatically improved and discharged with tapering dose of prednisolone.

Fig. 1 Contrast CT scan shows a cystic lesion with partial ring enhancement in right frontal region

Fig. 2 Sagittal T1-weighted MR image shows large hypointense lesion in frontal regions extended to the lateral ventricle but not extended to the cortex

Fig. 3 Axial FLAIR MR image shows multiple hypersignal intensity lesions in both frontoparietal lobes

After prednisolone was discontinued for 4 days, he experienced progressive paraparesis and numbness of both legs and perianal area. The neurological examination revealed asymmetrical weakness of both upper and lower extremities, grade 4/5 on the right and grade 2/5 on the left. Lumbar puncture was normal. The second MRI was done 3 months after the
first MRI. The study showed new foci at both sides of splenium of corpus callosum and T9-10 spinal cord (Fig. 5). The finding of two clinical attacks and evidence of multiple lesions in MRI were compatible multiple sclerosis due to McDonald diagnostic criteria(3). He was treated with pulse methylprednisolone 1g/day for 5 days. After treatment, his motor power was improved but still experiencing ataxia and paraparesis.

Discussion

Multiple sclerosis is one of the most important neurological conditions due to its prevalence, chronic nature and tendency to attack and eventually disable young adults. The diagnosis of multiple sclerosis largely depends on the clinical course, which features the exacerbation and remission of multifocal neurological deficits. Electrophysiological tests, CSF analysis and imaging studies can assist in the diagnosis. Clinicians have recently come to rely on MRI, because it is more sensitive for detecting white matter lesion than CT scan (5,7-11).

Imaging studies usually reveal multiple small plaques ranging from a few millimeters to 16 mm in size(5,11-13). However, sometime multiple sclerosis presents as a large solitary mass lesion that is indistinguishable from a brain tumor(4,5,12-14).

MRI in multiple sclerosis shows abnormalities typically in the form of high intensity T2-weighted lesions with asymmetrical, ovoid, well-demarcated border and oriented perpendicularly to the ventricular surface as well as in the brainstem and the spinal cord(14). Multiple sclerosis plaques typically found in the periventricular region, corpus callosum, centrum semiovale, and, to a lesser extent, periventricular white matter, basal ganglia. Lesions usually are arranged at right angles to the corpus callosum as if radiating from this area(15). Contrast enhancement can be observed in some of these lesions representing an acute inflammatory process(14).

Areas of increased signal intensity predominantly in the white matter are detected by T2-weighted pulse sequences in 70-100% of patients with clinically define MS(16). On the other hand, only 30% of patients with MS showed CT scan abnormalities, with 74% of these abnormalities being low-density lesions in the white matter(13,18). Low-density lesions with peripheral ring enhancement on CT are rare in MS(13,19,20).

Necrosis, cyst formation and cavitation are rare in MS, although it is relatively common in other demyelinating diseases, such as neuromyelitis optica (Devic’s disease), Schilder’s disease, acute disseminated encephalomyelitis, and concentric sclerosis(19-21). The exact mechanism of necrosis in demyelinating disease is still obscure(20). Necrosis may result from edema and swelling occurring in and around the active plaque(22).

The pattern of contrast enhancement exhibited by demyelinating lesions may help to differentiate them from neoplasm or infection. This imaging pattern
consists of incomplete, C-shaped or “open-ring” enhancement, as compared with the completely circular ring of contrast enhancement that is characteristically seen with abscesses and tumors. This pattern has also been called the “white matter crescent sign” because in some cases the open portion of the partial ring abuts the cortical gray matter and the area of enhancement sweeps through the white matter in a crescent like pattern(23).

Recently, the proton MR spectroscopy has been applied to differentiate Tumefactive Demyelinating Lesions (TDLs) and high-grade gliomas. The finding shows significant differences in the N-AcetylAspartate/Creatine ratio (NAA/Cr) in their central regions, no other metabolite ratios could be used to distinguish high-grade gliomas and TDLs with confidence. This finding emphasizes the need for the cautious interpretation of spectroscopic findings(24).

In summary, the following CT scan and MRI finding are characteristic of multiple sclerosis: (a) multiple white matter lesions (predominantly in the periventricular white matter), (b) combined supratentorial and infratentorial lesions, and (c) minimal surrounding edema and mass effect relative to the size of the lesions.

In our case, the occurrence of patchy low density lesions with partial rim enhancement (open-ring) in CT scan raised questions of a cysticercosis, cystic glioma, brain abscess or metastases. MRI findings showed multiple large mass lesions surrounded with brain edema. In retrospect, the mass effect seemed to be minimal compared to the size of the lesions. The results of brain biopsy, subsequent clinical course, response to treatment, and repeated imaging findings lead to a diagnosis of multiple sclerosis.

Acute widespread, demyelinating pseudotumor or tumor-like manifestation is one of the clinical variants of multiple sclerosis. Classically, it presents as a solitary contrast enhancing mass lesion within the brain, which by radiographic examination, simulates a neoplasm. Mass effect may occur, with compression of the lateral ventricle and shift across the midline(10). The clinical abnormalities in such patients are variable. The abnormalities may be very minimal even in a patient with a massive lesion, while in another patient with similar lesions, confusion, hemiparesis, or neglect syndrome can be seen. Much of the T2 hypersignal lesion volume is often due to edema and may be responsive rapidly to steroid. However, this change with steroids may also occur with glioma or with CNS lymphoma and is therefore not a useful diagnostic criterion. Finally, brain biopsy is often required.

The nature of these large demyelinating lesions is uncertain. Poser et al. considered them to be a variant of multiple sclerosis, while Kepes suggested that they represent an intermediate entity between multiple sclerosis and acute disseminated encephalomyelitis(21,25,26).

Most reported cases respond to steroid therapy in the acute period and seems to follow clinical course of typical MS. However, there is no data from large series or consensus in treatment response or long-term prognosis in this specific group of patients.

References


อาการแสดงคล้ายก้อนเนื้องอกรูปแบบที่พบน้อยในโรคมัลติเปิล สเคลโรซิส: รายงานผู้ป่วย 1 ราย

ประวีณ โล่ห์เลขา, ก้องเกียรติ ภูมิภัททกุธาร

มัลติเปิล สเคลโรซิส เป็นโรคที่มีการปรากฏของปลอดประสาทในระบบประสาท ตัวเกิดจาก ทำให้มีอาการทางระบบประสาทตามตำแหน่งของโรค ส่วนใหญ่จะสัมผัสกับ หาย ๆ อาการแสดงคล้ายก้อนเนื้องอกในภาพถ่ายรังสี เป็นรูปแบบหนึ่งของโรคมัลติเปิล สเคลโรซิส ซึ่งพบได้น้อย รายงานได้เสนอผู้ป่วยชาย อายุ 36 ปี มีอาการชาใบหน้า และลำตัวด้านซ้ายมา 1 เดือน อาการของโรคเป็นมากขึ้นเรื่อย ๆ ภาพถ่ายทางรังสีพบว่า มีลักษณะคล้ายก้อนเนื้องอกในสมองทั้ง 2 ข้าง อาการชาใบหน้าเพิ่มขึ้น หลังจากได้รับการรักษาด้วยสีสัน ต่อมาผู้ป่วยกลับมีอาการของปลอดประสาท ด้านขวาอาการดังกล่าว หลังจากได้รับการรักษาด้วยสีสันใหม่ ภาพถ่ายทางรังสีที่ 2 พบว่ามีรอยโรคใหม่บริเวณประสาทไขสันหลังระดับช่องอกที่ 9-10 จากลักษณะทางคลินิกการด้านเนื้องอก และลักษณะทางภาพถ่ายรังสีฯ ได้กับโรคมัลติเปิล สเคลโรซิส ที่มีลักษณะคล้ายก้อนเบื้องต้นเช่นโรคตับ