Tumor-like presentation of multiple sclerosis

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ABSTRACT

Multiple sclerosis patients may present with clinical data suggestive of cerebral tumor, however, most of the lesions do not show expansive signs in computerized tomography of brain or magnetic resonance imaging. We report in this paper, 2 patients who had shown expansive radiological signs suggestive of neoplasm. Cerebral biopsy was an important diagnostic procedure in these 2 cases which revealed the diagnosis of demyelinating disease.

Keywords: Tumor, demyelination, multiple sclerosis.

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The spectrum of primary demyelinating disease includes various pathological entities,[1] multiple sclerosis (MS) being the most common.[2] These diseases uncommonly have histological confirmation except where the diagnosis is uncertain, such as a single or large lesions with mass effect simulating glioma or other neoplasm.[3-5] Accurate diagnosis removes uncertainty when patients with MS present with clinical data suggestive of cerebral tumor.[2] Primary demyelinating disease of the central nervous system (CNS) generally does not produce a focal diffuse mass lesion, a feature that has been used to distinguish demyelinating lesions from tumor.[6] We present 2 cases, where the clinical presentation and neurologic findings were suggestive of CNS tumors and brain biopsy proved the histological nature of demyelinating disease.

Case Report.

Patient 1. A 25-year old Saudi male, school teacher, had 2 weeks history of vomiting, dizziness and difficulty in walking. Clinical examination revealed an ill-looking young man with horizontal and vertical rotatory nystagmus and right hemiparesis with mild limb and truncal ataxia. Hemogram and biochemisty were normal. Cerebro spinal fluid (CSF) examination showed 200 cells 100% lymphocytes, sugar normal and protein slightly elevated (0.77 g/L [N=0.15 -0.45 g/L]) IgG index normal, oligoclonal band negative, cytology normal. Screen for tuberculosis, brucella and fungals were negative. Electroencephalogram (EEG) showed focal slowing in the right temporo-parietal region. Evoked potentials and EEG were normal. Computerized tomography (CT) of brain showed a low attenuated area deeply-seated on the right temporo-parietal which is minimally enhancing with no mass effect seen. Brain magnetic resonance imaging (MRI) revealed multiple increased signal lesions without mass effect (Figure 1). Three foci of abnormal signal intensity; the largest located in the right thalmic and posterior basal ganglia involving the posterior limb of the internal capsule, a lesion showing focal enhancement with gadolinium, but no mass effect; scattered and small subcortical foci of increased signal intensity throughout both cerebral hemispheres while the other lesion in the medulla posteriorly on the right shows no mass effect for enhancement; minimal enhancement seen in the left

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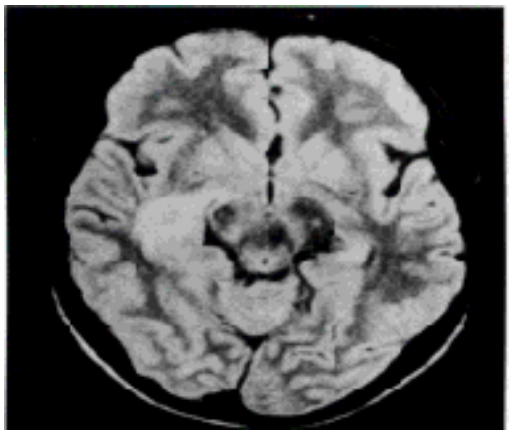

Figure 1 - MRI axial proton density cuts showing the largest right thalmic and posterior basal ganglia hyperintense lesion involving the posterior limb of the internal capsule.

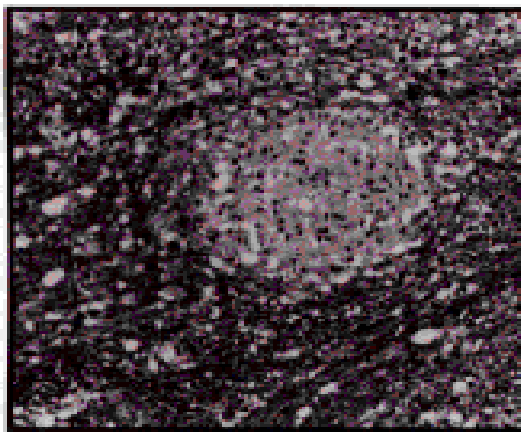

Figure 2 - Myelin basic protein staining of biopsy showing focus of demyelination with marcophage infiltrate.

cerebellar lesion. MRI angio showed normal intracranial vessels. Two days after admission, the patient deteriorated rapidly with difficulty in breathing, drowsiness, and dense left-sided hemiplegia which necessitated intensive care management and ventilation. CT brain showed the previously seen hypodense area had increased in size producing a mass lesion in the right temporo-parietal region with the effect of moderate compression of the body of the right lateral ventricle together with mid-line shift towards the left side. These changes were highly compatible with tumorative behavior. Steriotactic brain biopsy for the right temporo-parietal region performed at this stage showed demyelinating changes (Figure 2). The patient responded dramatically to pulse therapy with methylprednisolone and was extubated with good recovery of the left hemiplegia. He walked without assistance and was discharged home in good condition. The patient was readmitted 8 weeks later with recurrence of his symptoms and signs and improved partially after steroid pulse therapy.

Patient 2. A 16-year old Saudi male student presented with a 3 weeks history of headache, progressive left-sided body weakness, speech and behavioral changes, agitation, non comprehensive speech, bouts of crying and laugher. Neurological assessment revealed positive primitive reflexes, emotional liability and left-sided hemiparesis. Investigations showed normal hemogram, erythrocyte sedimentation rate (ESR) and biochemistry. EEG and evoked potentials were normal. Brain CT showed a large area of low-attenuation in the right fronto-parietal lobe. MRI revealed multiple increased lesions in the white matter (Figure 3a and 3b). CSF examination showed no cells with normal sugar and protein and negative oligoclonal band. Steriotactic brain biopsy was

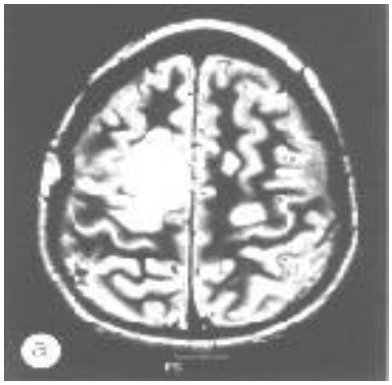

Figure 3a - Axial MRI T2 weighted images showing large right fronto parietal hyperintense lesion and 2 small white matter lesions.

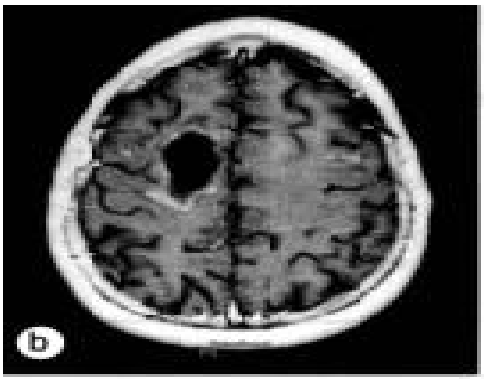

Figure 3b - Axial T1 cuts after contrast enhancement with gadalenium, showing right fronto parietal hypointense lesion with irregular rim enhancement.

consistent with a demyelinating benign lesion. The patient showed a good response to steroid pulse therapy and was discharged home in good condition. He was readmitted 4 months later with impaired cognitive function, dysphasia and left hemiparesis and responded remarkably to steroid pulse therapy.

Discussion. In the absence of pathognomonic clinical features or a definitive laboratory test, MS remains ultimately a diagnosis of exclusion.[2] Because of its heterogeneous manifestations, incorrect diagnosis is not rare. The rate of inaccurate diagnosis remains around 5 to 10%.[7] Several monophasic syndromes may represent the onset of MS, or may be self limited and never lead to further events. They include isolated optic neuritis, acute transverse myelopathy, and acute disseminated encephalomyelitis (ADEM). In these monophasic syndromes, CFS and MRI abnormalities consistent with MS increase the risk for development of MS over the next several years.[8] Multiple sclerosis can present as a cerebral mass lesion with clinical features and findings on imaging studies suggesting a neoplasm. Biopsy then leads to the surprising findings of inflammatory demyelination.[9] The converse situation, namely the mistaken diagnosis of a neoplastic process as demyelination, also rarely occurs. The most common neoplasm mistaken for MS is CNS lymphoma.[1-5,10] Central nervous system lymphoma is usually unifocal, but often may be multicentric. Cerebro spinal fluid examination may demonstrate cytologically abnormal lymphocytes but usually is normal or demonstrates only nonspecific abnormalities of protein or pleocytosis. Both the clinical and radiographic manifestations of CNS lymphoma may respond to corticosteriod therapy leading to further confusion.[11] Magnetic resonance imaging findings of CNS glioma may also look similar to those of MS with T2-hyperintense lesions in the cerebral white matter extending into corpus callosum and variable Gd-enhancement.[12] The radiographic distinction may be made by the demonstration of involvement of gray matter structures out of proportion to white matter, lesion behavior over time is more helpful. Enhancement of MS lesions rarely persist for longer than 4-6 weeks. Thus, persistent Gd-enhancement, particularly if there is a prominent component of enhancement along blood vessels, and continued enlargement of lesions strongly suggests a neoplasm. Ultimately, biopsy is necessary to make the correct diagnosis.[13] However, occasionally lesions of MS can appear to be very large on MRI and yet pathologically represent very small areas of demyelinating as in our case.[2] This may be due to an increase in tissue water from a disrupted brain blood barrier associated with inflammation and edema giving rise to increased signals that appear similar to signals from demyelination and gliosis.[13-14] Computerized tomography scan findings of mass effect and peripheral contrast enhancement in cases of MS have to lead to the mistaken diagnosis of glioma. In the series of Reth et al, 6 patients had an infiltrating lesion of the corpus callosum mimicking a glioma and underwent surgical biopsy.[15] In our cases the CT scan and MRI findings and the clinical presentation of focal neurological deficit were highly suggestive of CNS tumor. Our report emphasizes the importance of considering primary demyelinating disease in the differential diagnosis of a cerebral mass lesion especially in younger patients.

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