A Case of Rapidly Progressive Dementia With Multifocal Brain Lesions

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Abstract

Objective: The purpose of this article is to describe and discuss the clinical manifestations of a patient with the rapidly progressive dementia which may have been caused by multifocal brain lesions.

Case Report: A 65-year-old man developed rapidly progressive dementia 4 months prior to presentation. He had a large decline in the immediate and recent memories, as well as abilities in basic and instrumental activities of daily living (ADLS and IADLS), within a short period of time. However, the other features of dementia, such as delusions, hallucinations, and depression, were not found. Computed tomography (CT) of the brain showed only mild brain atrophy, an enlarged pituitary gland, and on the contrary, multifocal brain lesions were detected by magnetic resonance imaging (MRI).

Conclusion: The case reminded us the importance of a thorough evaluation on an unusual patient experiencing rapidly progressive dementia. Even though brain CT indicates only non-specific findings, further delicate imaging studies (brain MRI) should be performed to evaluate those with unusual clinical course of cognitive decline.

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Key words: dementia, brain lesion, magnetic resonance imaging (MRI)

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Introduction

A variety of disorders and conditions can lead to dementia. For most etiologies, dementia is a chronic, progressive, and irreversible morbid condition without available treatments or interventions currently. However, its clinical course may be arrested or ameliorated to varying degrees with some specific managements. Potentially reversible conditions include drug- and alcohol-induced dementias, metabolic conditions (such as vitamin B\textsubscript{12} deficiency and hypothyroidism), and dementia syndrome of depression [1]. Dementias caused by structural brain lesions such as subdural hematoma, normal-pressure hydrocephalus, and brain tumors are also somewhat potentially reversible to a certain extent.

We present a case of dementia that progressed rapidly during a four-month course, caused probably by multifocal brain lesions detected by magnetic resonance imaging (MRI) rather than computed tomography (CT).

Case report

A 65-year-old married patient was admitted to our geropsychiatric ward because of his inability to find the way home twice in one week. Essential personal information of the patient were summarized as follows: he graduated from elementary school and had a medical history of diabetes mellitus (DM) under control by oral hypoglycemic agents in the past 10 years. For many years, he earned his living by selling ready-to-wears in a night market. In February 2006, his family apparently observed his repetitive counting of money and impairment of calculation. His memory function had continued to decline since then, and he got lost around 5 kilograms rapidly in March 2006. He was once found lying on the floor at home unconsciously and sent to the emergency department of a local hospital. His blood sugar measured up to 800 mg / dL, and hyperglycemic hyperosmolar nonketotic coma (HHNK) was diagnosed by the doctor. As his families discovered later, he had forgotten to take his hypoglycemic medications for one month. He was hospitalized for one week to recover with his blood sugar stabilized under careful control since then. After being discharged from the local hospital, the patient was monitored by his families to adhere to his medications. In mid April, he could still play cards at his friend’s house. Since late April, it had become difficult for him to find the way to his friend’s house, so he was forced to stay at home all day long. In the beginning, he had been observed to forget the place of wardrobe and bathroom everyday and used others’ towels and
toothbrushes. By early May, activities of daily living (ADL), such as dressing and bathing, were performed in a careless manner and required the assistance from his wife. Two weeks prior to the admission, he insisted that the house in which he had been living was not his home. In the last week before admission, he lost his way home twice while with an unsteady gait observed. He was admitted to the geropsychiatric ward at the end of May 2006. Upon admission, his mental status examination showed poor concentration, disorientation to place, blunted affect, confabulation, and repetitive behavior; there was, however, no delusion, hallucination, or depression. Mini Mental State Examination (MMSE) score emerged to be 15, and the Clinical Dementia Rating (CDR) and Hachinski Ischemia score were 2 and 5 respectively. Findings of the neurological examination were normal except for a wide-based gait. Results of routine laboratory tests, including complete blood count (CBC), urinalysis, serum biochemistries (electrolytes, liver and renal functions, vitamin B₁₂, folic acid), thyroid function, serological tests for syphilis, and tests for human immunodeficiency virus antibodies, were all negative. However, hyponatremia (129 mEq/L, reference range 135 – 147 mEq/L) was noted. Electroencephalography (EEG) showed moderate slow pattern bilaterally. Brain CT showed slightly generalized atrophic change of the brain parenchyma and an enlarged pituitary gland. The results of the other laboratory tests were as follows: serum prolactin 19.11 ng/mL (reference range 2.0 to 14.7 ng/mL), follicle-stimulating hormone 2.46 MIU/mL (reference range 1 to 8 MIU/mL), luteinizing hormone 2.83 MIU/mL (reference range 0.6 to 12 MIU/mL in follicular phase), 8:00 AM serum cortisol 136 ng/mL (reference range 50 to 250 ng/mL), and anti-diuretic hormone 0.28 pg/mL (reference range 0.4 to 2.4 pg/mL).

During the whole course of hospitalization, the patient’s hyponatremia improved with sodium supplement, and the blood sugar was controlled at around 110 to 130 mg/dL. However, cognitive impairment persisted. He had difficulty in finding his own room in the ward and was unable to perform ADLs without the assistance from his family. Because neither psychotic nor depressive symptoms were detected, no antipsychotic or antidepressant agents was prescribed.

Brain MRI was performed for the unusually rapid clinical course of dementia in this patient. T2-weight, MRI revealed multifocal high signal lesions on T1 involving the medial aspect of bilateral thalami, bilateral dorsal aspects of the brain stem, and the white matter of bilateral cerebral hemispheres.
T1-weighted images further indicated a pituitary tumor (about 11 mm) (Figures 1) and multifocal gadolinium-enhanced lesions in bilateral mesial temporal regions along the periventricular area, in the body of the corpus callosum (Figures 2 and 3), and in the periventricular regions along the occipital horn and body of bilateral lateral ventricles (Figures 4 and 5). The patient refused tumor biopsy and insisted to be discharged.

Discussion

According to the Diagnostic and Statistical Manual of Mental Disorders (DSM-IV) [2], the rapidly declining memory, attenuating execution of tasks, and growing difficulty in ADLs performance could be noted in the patient just within a 4-month interval are typical symptoms and signs of dementia. However, the clinical course of the patient progressed relatively faster than usual. Disturbance of sleep-wake cycle and disorientation were noted concurrently in this patient; it was necessary to examine whether the delirium presented or not. The condition, however, soon stabilized after hyperglycemia hyponatremia were corrected. Based on the morbidity history and long-term observations in the psychiatric ward, we concluded that the main diagnosis for the patient was dementia rather than delirium. Even if the patient’s cognitive function declined dramatically in just a few months, it didn’t favor primary degenerative disease, such as dementia of the Alzheimer’s type.

Compared with CT, MRI used more advanced techniques like spin echo, gradient echo, combination spin and gradient echo pulse sequences before and after intravenous administration of paramagnetic contrast agents; therefore, it was able to provide greater contrast resolution between structural abnormalities and adjacent brain parenchyma, and increases the sensitivity in detecting focal lesions of the brain. Early experience suggested that

Figure 1. The brain MRI showed an enlargement of the pituitary fossa with a increased mass image of the pituitary gland (arrow). (T1WI, with contrast)
Figures 2 and 3. The brain MRI showed multiple nodular enhanced lesions at the corpus callosum (arrow). (T1WI, with contrast)

Figures 4 and 5. Multiple enhanced lesions were detected in the periventricular region along the left wall of the third ventricle and bilateral lateral ventricles. (T1WI)
3~30% more focal intracranial lesions could be identified by MRI than by CT [3]. Periventricular hyperintensities [4], white matter lesions (WML) [5, 6] and the corpus callosum (the main interhemispheric fiber connection) [7] were shown to be associated with dementia syndrome. Moreover, limbic structures of medial temporal lobe (MTL) were regarded as a potential key to the integrity of declarative memory function [8], and atrophy of the MTL was associated with the severity of dementia [9]. On T2-weighted MRI on our patient, obvious thalamus lesions involving the brain stem and white matter of the bilateral cerebral hemispheres were detected. On the T1-weighted images with contrast, lesions in the bilateral medial temporal regions, periventricular area and the corpus callosum were also shown. The lesion sites and clinical manifestations of our patient seemed to be consistent with those of several previous cases. According to the radiologist, the differential diagnosis of these lesions might include lymphoma, leukemia, tumor infiltration (primary to be determined) and unidentified infectious origins [10]. There was no significant finding on vital signs, CBC and biochemistry data (except hyponatremia). However, after performing the contrast MRI, remarkable multifocal lesions with high signal intensity were revealed on both the T1 and T2-weighted images, and the blood-brain barrier (BBB) at those areas reflected some degree of impairment. In general, tight intercellular junctions between capillary endothelial cells created the BBB that could block the penetration of the radionuclide or contrast medium into intravascular space [3]. Because there was no available pathology report for the brain lesions in this patient, we could not differentiate these lesions just by the findings of the brain images. As suggested in related literatures, in might primary CNS lymphoma, tumors appear to be one or more (usually multiple) firm or friable, centrally located, deeply seated masses with variable demarcation from adjacent cerebral parenchyma. Tumors were often located in close proximity to the ventricles and might infiltrate through and along the ependymal ventricular walls. Corpus callosum and basal ganglia were the common sites of involvement [11]. In secondary CNS lymphoma, as in primary Counterpart, multicentricity was common; however, in the case of our patient, the masses were predominantly peripherally, not deeply located. Both focal and diffuse thickening of the arachnoid occurred, and the infiltration also extended into the overlying dura [3]. By MRI, leukemia could appear as a solid mass extending inward from the meninges or within the brain parenchyma contiguous with the ventricular wall ependyma [12].
conclusion, according to the locations of our patient’s lesions, primary CNS lymphoma and metastatic tumors were the other probable diagnostic listing while secondary CNS lymphoma and leukemia were not likely.

The case report had several limitations. First, the pathology of the brain lesions was not available. Second, the hemoglobin A1C (HbA1c) was not measured at admission, so the glycemic control before hospitalization could not be assessed. The risk of DM with poor glycemic control, related atherosclerosis, vascular dementia and AD could not be comprehensively reviewed. Third, the association of hyponatremia with cognitive decline could not be ruled out [13]. Both the actual duration and severity level of hyponatremia remained uncertain. The patient’s rapid cognitive decline might be due to a mixture of all possibilities mentioned above.

In summary, this case illustrated the importance of a thorough evaluation on patients with rapidly progressive dementia with obvious neurological signs. Unlike other geropsychiatric patients with primary degenerative dementias, our patient exhibited a shorter course and gait disturbance. There was also no obvious delusion, hallucination or depression. The patient presented dementia features as well as several unusual attributes that did not lead themselves to a precise diagnosis of primary degenerative disease. Therefore, for an elderly patient showing early rapid deterioration of cognitive function associated with significant neurological signs, the geriatric psychiatrist should not assume the usual diagnosis of a primary degenerative dementia, such as Alzheimer’s disease. Brain CT and other imaging examinations, such as brain MRI, should also be arranged for more comprehensive evaluation.

References


與多處腦部病灶可能有關快速進展的失智症 —
個案報告

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摘 要

目的: 描述與討論1位與多處腦部病灶可能有關的快速進展的失智症
病患所呈現的临床表徵。

病例報告: 病人為65歲已婚男性, 因為近來迷路兩次而住院評估治療
。病人自住院前四個月開始工具性日常生活功能 (IADL)、日常生活功能
(ADL)、和瞬間或即時記憶與近期記憶皆快速退化。腦部電腦斷層檢查指
發現輕微腦萎縮與腦下垂體腫大, 但是在核磁共振檢查僅發現有腦部多發
性病灶。病程中沒有觀察到有妄想、幻覺、或憂鬱。

結論: 此個案彰顯臨床處理進展快速失智症病患時, 詳細評估的重要
性。當病患的認知呈現不尋常的退化病程時, 雖然腦部電腦斷層攝影沒
有明顯的發現, 仍需考慮更進一步精細的影像學檢查 (如核磁共振檢查)。
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關鍵詞：失智症、腦部病灶、核磁共振

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