Case Report

A rare case of gliomatosis cerebri presenting as dementia

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Abstract

Dementia with the onset before the age of 65 years is classified as early-onset dementia. Although uncommon, it has considerable impact on the lives of patients and care givers, alike. A substantial subset of patients may have underlying reversible causes. Yet, many, especially those of the very young may be initially misdiagnosed. A case of young woman with rapid mental decay is described here. She was finally diagnosed with gliomatosis cerebri (GC) involving only right frontal lobe. This atypical radiological feature of GC with primary presentation as memory loss needs special attention and clinicians should be aware of such conditions.

Key words: Dementia, gliomatosis cerebri, MR spectroscopy, neoplastic, neurocysticercosis

INTRODUCTION

Early-onset dementia (at age < 65 years) is an uncommon but important group of disorders. It has devastating impact on the lives of patients and families and deserves attention of the clinicians. Dementia in younger people usually has some demonstrable secondary etiology (psychiatric illness, alcohol or drugs or metabolic disturbance, familial Alzheimer's disease, Wilson's disease, etc). Cognitive deficits secondary to brain tumors rarely occur, and there is little description in the available literature where it has been described in association with low grade gliomas, gliomatosis cerebri (GC), and primary brain lymphoma.[1] Gliomatosis cerebri is a highly aggressive, rare form of malignant astrocytic tumor. It is most commonly present as a diffusely infiltrating glial tumor of the cerebral cortex involving more than two lobes and occasionally infiltrating infratentorial structures and the spinal cord.[2] We describe a young woman with GC confined to a single lobe (right frontal) and presented primarily as a dementia. Before the advent of magnetic resonance imaging (MRI), diagnosis was generally not established until autopsy was done. Even with MRI, however, diagnosis is difficult. Management of GC is also difficult by virtue of their diffuse nature, surgery is not suitable and large field radiotherapy carries the risk of severe toxicity. As per our knowledge, only few cases have been reported that had involvement of single hemisphere and presented only with memory loss.[3]

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CASE REPORT

A 31-year-old woman was admitted to our hospital with a 3-year history of progressive memory loss and headache. The patient mainly complained of difficulties doing her usual daily routine activity job for about 1 year and misplacing personal objects, she was having difficulties doing simple calculations and drawing simple paper patterns, resulting in erroneous calculations and wrong measurements along with decreasing ability to perform complex tasks such as arranging a meeting with her old friend, pay bills, and marketing. Childhood and prior developments were apparently normal. No neuropsychiatric disorders could be assessed in the family. She did not take drugs, including alcohol. Her past medical history was uneventful and was negative for stroke, transient ischemic attack, chronic infection, or other chronic illness. Except for incontinence and a slight slurring of speech, the physical examination revealed no further neurological deficits, especially no speech impairment, seizures or myoclonia were observed. The Mini Mental

State Examination score was suggestive of dementia (21 of 30) and very low on the cognitive efficiency profile. There were also deficits on tests of visuospatial ability. Nevertheless, she experienced no changes in personality, appetite, or sleeping pattern noted, nor was she observed to be a snorer or have spells of apnoea, and there was no history of auditory or visual hallucinations. Blood count, biochemical profile, sedimentation rate, coagulation profile, and the results of radiography of the chest were normal. Cerebrospinal fluid analysis showed normal results. Results of Venereal Disease Research Laboratories testing were negative, and no malignant cells were identified by cytological examination. Serum vitamin B12, folic acid, and thyroid-stimulating hormone levels were within normal limits. Computerised tomography of the brain revealed mild left hemispheric mass effects. MR imaging and proton MR spectroscopy were performed on a clinical whole body 1.5-T imaging unit. T2-weighted and fluid attenuated inversion recovery FLAIR images revealed an ill defined area of signal intensity alteration in the left frontal region with involvement of rostrum of corpus callosum with minimal postcontrast enhancement. The lesion was causing effacement of cortical sulci [Figure 1]. The magnetic resonance spectroscopic imaging results showed an increased choline peak, a decreased *N*-acetyl aspartate (NAA) peak with an increased Ch/Cr ratio at the site of lesion. The patient was given supportive care for a period of 15 days following that she improved a little bit and the patient was given supportive care for a period of 15days following that she improved a little bit and discharged, and at discharge, her MMSE was 22/30 and she was also referred to higher centre.

DISCUSSION

The differential diagnosis of young-onset dementia is extensive, including sporadic hereditary etiologies, neurodegenerative diseases, adult presentations of inborn errors of metabolism, other metabolic or storage diseases, and other syndromic diagnoses.[4,5] Although investigations

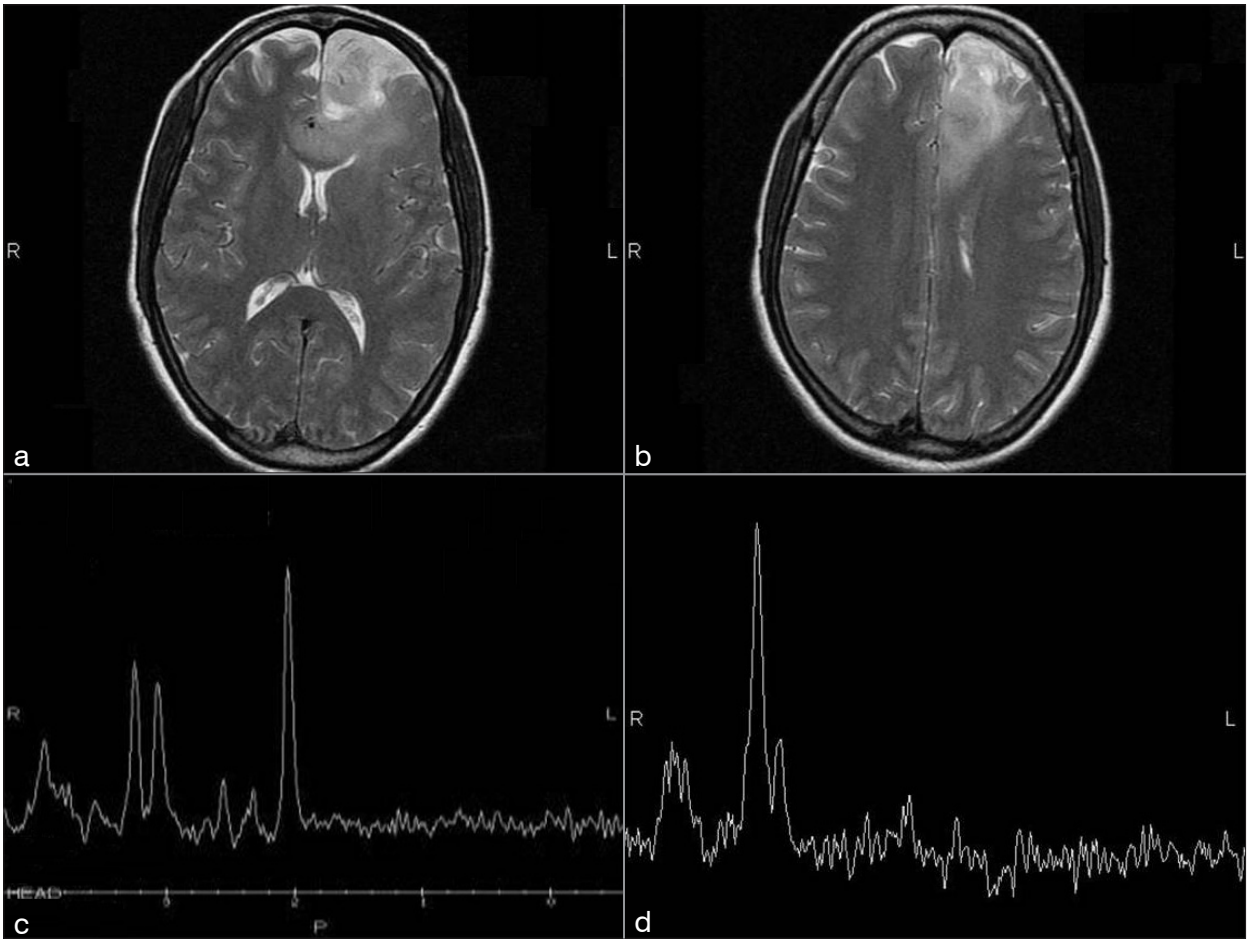

Figure 1: (a) Axial view T2 FLAIR image demonstrating tumor-related infiltration involving left frontal lobe involvement of rostrum of corpus callosum. (b) Axial view T2 FLAIR image shows extensive hyper intensity involving the left frontal lobe and rostrum of corpus callosum, with a mild mass effect. (c) The MR spectroscopic imaging results showed an increased choline peak, a decreased NAA peak with an increased Ch/Cr ratio at the site of lesion. (d) The MR spectroscopic imaging results showed an increased choline peak, a decreased NAA peak with an increased Ch/Cr ratio at the site of lesion

of the etiologies and prevalence of early-onset dementia have been performed,[4,5] in some individuals, the etiology remains indeterminate even after brain biopsy.[4] In young adults (up to 40 years of age), it is very rare to develop dementia without other features of neurological disease, or without features of disease elsewhere in the body. Most cases of progressive cognitive disturbance in this age group are caused by psychiatric illness, alcohol, or other drugs or metabolic disturbance. However, certain genetic disorders can cause true neurodegenerative dementia at this age. These include familial Alzheimer's disease, spinocerebellar ataxia 17 (dominant inheritance), adrenoleukodystrophy (X-linked), Gaucher's disease type 3, metachromatic leukodystrophy, Niemann–Pick disease type C, pantothenate kinase-associated neurodegeneration, Tay-Sachs disease, and Wilson's disease (all recessive). Wilson's disease is particularly important since cognition can improve with treatment.[4,5]

Dementia secondary to intracranial infections deserves a special attention especially in tropical and developing countries. Syphilis, HIV infection, Lyme neuroborreliosis, herpes virus, toxoplasmosis, cryptococcus, cytomegalovirus, or more rarely, Whipple diseases are few of the infectious causes of behavioral changes including dementia.[6] Besides few isolated reports of neurocysticercosis presenting with dementia, a recent controlled study reported dementia in 12.5% of the patients with active neurocysticercosis.[7,8] Brain tumors can cause dementia by mass effect on structures such as the hypothalamus or pituitary gland, which control hormone secretion and can also press directly on brain cells, damaging them. Treating the tumor, either medically or surgically, can reverse the symptoms in some cases.[9,10]

As in our case, the clinical course of GC is usually slow and long, especially when the neoplastic infiltrate is of low histological grade. The most common age group affected by GC is 50 and 60 years. In contrast, our patients is of young age.[3,11] The clinical course of GC is variable and includes pyramidal deficit, dementia, headache, cranial nerve changes, intracranial hypertension, seizures, and others. However, the present patient had a rare and atypical presentation in the form of dementia only. Before the inception of MRI, GC was diagnosed strictly in autopsy studies. However, MR imaging findings may be nonspecific, leading to a differential diagnosis that includes neoplastic, inflammatory, and vascular lesions. MR spectroscopy may further help in narrowing the differential diagnosis in favor of a neoplastic lesion by revealing increased Cho/Cr and Cho/NA and variably decreased NA/Cr.[9,10,12-14] Some non-neoplastic lesions such as encephalitis, demyelinating disease, and organizing hemorrhage, may mimic these spectral changes in rare instances.[15] If Cho/Cr or Cho/NA is not elevated, MR 5 spectroscopy may still help narrow the differential diagnosis in favor of GC. Specifically, *m*-Ins/Cr and *m*-Ins/NA should be measured because they can be elevated even when Cho/Cr is normal.[10,12] As a diffuse tumor, surgical treatment is usually not recommended, and radiology over extensive areas may cause serious toxicity.[13] This case highlights two atypical findings: (1) dementia in the young patient caused due to GC and (2) involvement of single lobe of brain which may be initial stage of growing GC, later on bilateral lobes may be involved.

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