Yonsei Medical Journal
Vol. 41, No. 4, pp. 517−521, 2000

Diffuse Cerebrospinal Gliomatosis with Extensive Leptomeningeal Spread

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Abstract

A case of diffuse cerebrospinal gliomatosis with extensive leptomeningeal spread is presented. The patient, an 18-year-old girl, was admitted due to progressive weakness and paresthesia of both legs, following rapid neuropsychiatric deterioration. An initial magnetic resonance imaging (MRI) study of the T-spine showed diffuse high signal intensities from T9 to T12 spinal cords on a T2 sagittal image and diffuse cord bulging at T1WI. This suggested an inflammatory lesion such as tuberculosis or fungal meningoencephalitis. A limited autopsy was performed. A microscopic examination revealed multifocal GFAP-positive astrocytic proliferations that were low grade astrocytoma in the cerebral leptomeninges, parietal, occipital and temporal lobes and anaplastic astrocytoma in the spinal cord and spinal leptomeninges. The high proliferative indices of the spinal lesion and aneuploidy correspond to a diagnosis of malignant astrocytoma and a rapid fatal clinical course.

Key Words: Cerebrospinal gliomatosis, subarachnoidal, leptomeningeal, meningoencephalitis, flow cytometry

INTRODUCTION

Diffuse cerebrospinal gliomatosis is a very rare entity, the third type of gliomatosis cerebri and it denotes an extensive glioma involving the supratentorial compartment, posterior fossa, and even the spinal cord. Moore described the first case of diffuse cerebrospinal gliomatosis masked by syphilis in 1954.[1] The clinical manifestations are variable consisting of motor weakness, sensory change, behavioral and mental changes that mimic motor neuron disease[2-4] and meningoencephalitis. There is no specific radiologic finding for the condition. Clinical diagnosis is therefore impossible and a tissue biopsy is necessary for the diagnosis.

We present an autopsy case of diffuse cerebrospinal gliomatosis predominantly involving the spinal cord, which demonstrated a high proliferating activity of the tumor cells, corresponding to a rapid fatal clinical course.

Received November 13, 1999
Accepted April 18, 2000
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CASE REPORT

An 18-year-old girl was admitted to the hospital because of progressive weakness and paresthesia of both legs. The patient had been well until two months before admission, when she began to experience a painful sensation similar to electrical stimulation on both feet. Fifteen days before admission, the patient developed a headache on the posterior head. One week before admission, paresthesia and weakness of the legs worsened and the patient had difficulty climbing stairs. There was no recent history of chills, cough or diarrhea, although she displayed temporary facial twitching and tinnitus 3 days before admission. Upon neurologic examination, the patient was alert and oriented. Cranial nerve functions were preserved. Motor power was 5/5 in the upper extremities without drift; the lower extremities were partially paralysed; leg strength was 2/5 in both hip flexors; 4/5 in the quadriceps, hamstrings, dorsiflexors, and plantarflexors of the feet. She reported normal sensation in the upper extremities. There was marked dysesthesia below the level

of T11 upon pin prick test. Temperature and vibration sensations were 50% of normal below the level of T11 and the joint position was slightly impaired. Deep tendon reflexes were +++ in the lower extremities and ++ in the upper extremities; bilateral Babinski signs were not noted.

The results of the lumbar puncture are presented in Table 1. Blood chemistry and urinalysis were normal. The levels of urea nitrogen, creatinine, bilirubin, calcium, alkaline phosphatase, and uric acid were normal, as were tests for α-fetoprotein and CEA. Serum total protein and albumin were 8.3 g/dl (nl: 6.0−8.0) and 5.6 g/dl (nl: 3.3−5.3), respectively. Serum electrophoresis revealed slightly increased albumin and total protein levels. Tests for antibodies to the human immunodeficiency virus, and VDRL were negative, as were tests for varicella antibody and Epstein-Barr virus antibody in serum and cerebrospinal fluid. Skin tests for *Paragonimus westermani* and *Clonorchis sinensis* were negative. The brain stem auditory evoked potential and median nerve soma-

Table 1. Results of Lumbar Puncture Findings

Variable	First admission	Second admission
Opening pressure (mmHg)	300	260
Glucose (mg/dl)	44	
Total protein (g/dl)	5.3	4.5
Red cells (mm^3)	1	10
White cells (mm^3)	45	
polymorphonuclear cells		
mononuclear cells	93%	
eosinophils		
others	7%	
Cryptococcal Ag	negative	negative
ADL (u/L, 37°C)	16.0	
Cytology	cell paucity	negative
Oligoclonal band negative		
ELISA tests*	negative	

* ELISA tests for cysticercus, paragonimus, sparganum, clonorchis.

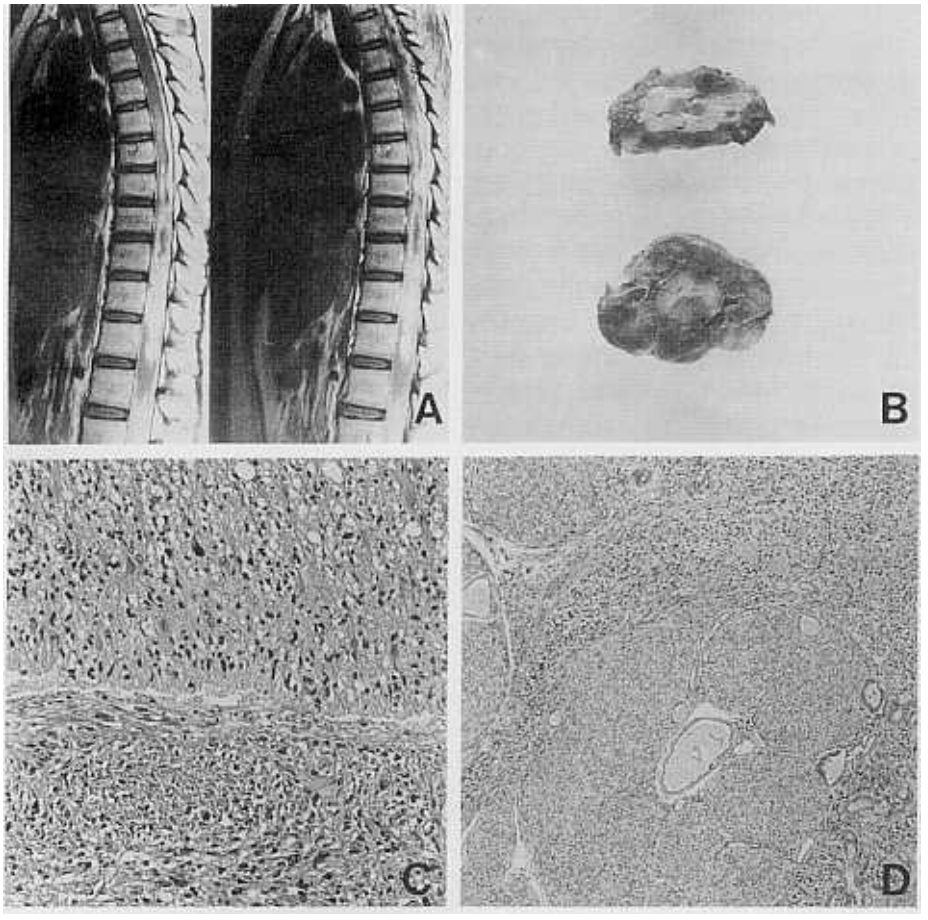

Fig. 1. Initial magnetic resonance imaging study demonstrated a diffuse irregular enhancing mass in leptomeningeal space (A). Transverse sections of the spinal cord showed an extensive leptomeningeal thickening and preserved cord contour (B) corresponding to the microscopic findings (C, D). H & E, ×200 and ×40.

tosensory evoked potential were within normal range, but the posterior tibial nerve somatosensory evoked potential showed conduction defects in the bilateral somatosensory evoked potential pathways. The magnetic resonance imaging (MRI) study of the T-spine showed diffuse high signal intensities from T9 to T12 of the spinal cords on a T2 sagittal image, and diffuse cord bulging at T1WI (Fig. 1A). The brain MRI was normal. In spite of large doses of steroid therapy (20 mg/kg/day) for 10 days, the symptoms slowly progressed. Symptoms of left 3rd, 4th, and 6th cranial nerve dysfunction (limitation of eyeball movements in all directions)were found on the 19th hospital day. Follow up brain MRI showed a thickening of the left cavernous sinus and carotid artery wall, and displacement of the left carotid artery to the anteromedial side (Fig. 2A). Although the patient had no medication for two weeks after self-discharge, eyeball movement progressively improved. However, weakness of the lower extremities and sensory deficits remained. One week before readmission, left vision became blurred and two days later right visual acuity also declined. She lost her bilateral eyesight for 3 to 4 days.

The patient was readmitted to the hospital because of progressive quadriplegia, paresthesia below the T5 level, visual loss of both eyes, posterior headache, neck stiffness and mild fever. She had severe neck stiffness and urinary incontinence. Often she had complained of visual and auditory hallucinations. Upon neurologic examination, the patient was alert and showed an appropriate verbal response to questions. Dilated pupils without direct and indirect light reflexes were found in both eyes. The patient neither noticed hand movement nor light perception. During left lateral gaze, a limitation of eyeball movement of the left eye was observed. Other eyeball movements were relatively intact. Motor power was 3/5 in the upper extremities; the lower extremities were paralyzed and flaccid. There was marked dysesthesia below the level of T5 upon testing for pin prick sensation, temperature sensation, vibration, and joint position. The deep tendon reflexes were + in the upper extremities and absent in the lower extremities; bilateral Babinski signs were not noted. The symptoms rapidly progressed and on the 3rd hospital day,

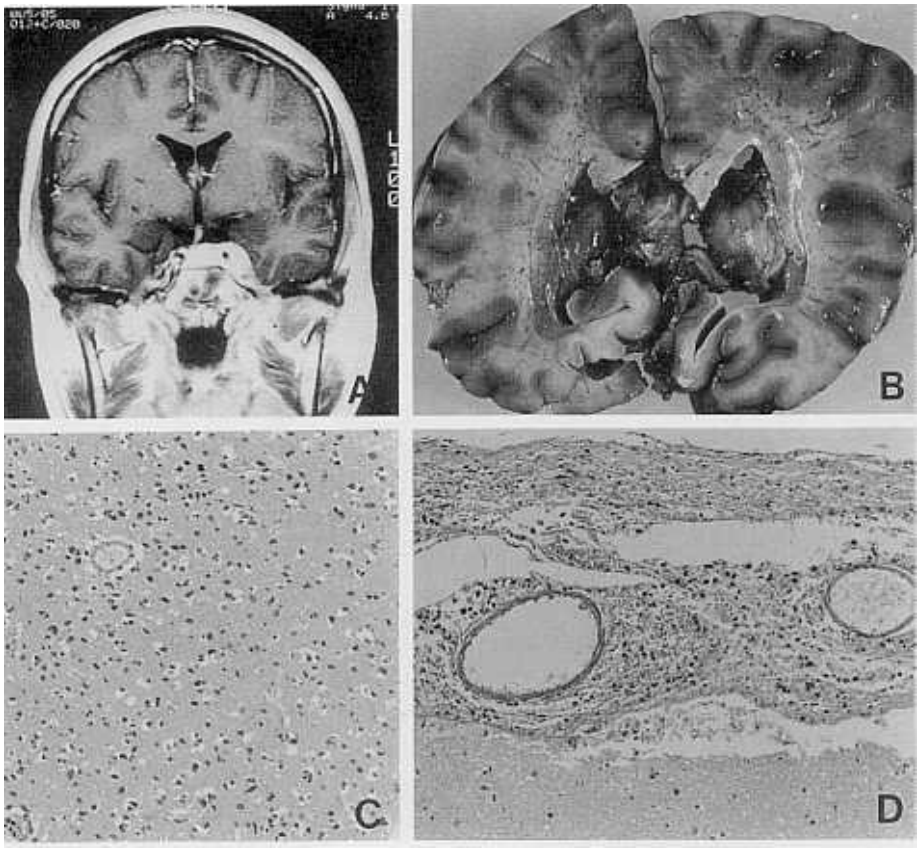

Fig. 2. Brain MRI showed an enhancing lesion in the left cavernous sinus without cerebral parenchymal mass lesion (A). Coronal sections of the brain revealed an irregular ventricular surface. Note the normal cerebral contour without gross abnormality in the hemispheres (B). Microscopic photographs show a gliomatous area involving temporal lobe (C), H & E, ×200 and subarachnoidal extension (D). H & E, ×100.