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Review article

Spinal primary central nervous system lymphoma: Case report and literature review

Feng Li [#], Chen Dingbang [#], Zhou Hongyan, Shen Cunzhou, Wang Haiyan, Sun Xunsha, Liang Xiulin, Chen Ling [*]

Department of Neurology, National Key Clinical Department and Key Discipline of Neurology, The First Affiliated Hospital, Sun Yat-sen University, Guangzhou 510080, China

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ABSTRACT

Primary central nervous system lymphoma (PCNSL) is a very rare tumor of increasing incidence. It is often misdiagnosed due to the unspecific presentation or unavailable biopsy, and results in poor prognosis. PCNSL involved the spinal cord is extremely sparse. Here we report a gentleman presented with one-year history of progressive tremor in the left limbs and slight dysarthria as well as three-month history of paraparesis, tinnitus and insomnia. MR images disclosed the swollen cerebellum and cauda equine, with contrast enhancement in both meninges and nerve roots. The cerebrospinal fluid (CSF) revealed extremely high protein level. Tubercular meningitis was considered and anti-tuberculosis therapy was given for weeks but without relief. With progressive deterioration, the PCNSL was eventually presumed according to positive CSF cytology and exclusion of systemic involvement. However, the patient passed away within days. We then reviewed the current diagnostic methods of PCNSL. The biopsy, as the gold standard for PCNSL diagnosis, is not eligible for all patients suspected PCNSL. The presurgical diagnostic algorithm of PCNSL has been fixed by clinicians and we suggest the early and repeated CSF cytology should be included for definitive diagnosis.

1. Introduction

Primary central nervous system lymphoma (PCNSL) is an aggressive malignancy tumor which represents about 1%-2% of all non-Hodgkin lymphomas and 2%-6% of all brain tumors [1–4]. It exclusively involves the central nervous system, including the brain, spinal cord, eyes, meninges and cranial nerves [5]. The spinal involvement is very rare and only a small number of cases were reported in literature [3,6–8]. Population aged 50–65 years old is especially vulnerable to PCNSL, which is diversified in clinical presentation and image characters [9,10]. Here we report a PCNSL case which involved the meninges and nerve roots mimicking tubercular meningitis and being prescribed with anti-tuberculosis drugs but without relief. Positive cerebrospinal fluid (CSF) cytology attributed to the definitive diagnosis of PCNSL. However, the patient was eventually passed away after weeks of deterioration.

2. Case report

A 45 year-old gentleman was admitted in our neurology department, who presented with one-year history of progressive tremor in the left limbs and slight dysarthria as well as three-month history of paraparesis, tinnitus and insomnia. He then developed severe dysarthria, sialorrhea, incompetent closure of the eyelids, constipation, atrophy in the left limbs as well as paralysis and numbness in the left lower limb in two months. CSF test revealed normal pressure, slightly decreased glucose (3.0 mmol/L in the CSF, 8.0 mmol/L in the plasma) and strikingly increased protein (18,962 mg/L). The magnetic resonance imaging (MRI) showed the swollen cerebellum and cauda equine, with enhancement in both meninges and nerve roots (Fig. 1). The electroencephalogram (EEG) was normal. Out of the fear of surgery, the patient refused to take biopsy of cauda equine. According to the epidemiology consult, the tubercular meningitis was suspected and the anti-tuberculosis recipe including Isoniazid (0.6 g QD), Rifampicin (450 mg QD), Pyrazinamide (1500 mg QD) and Ethambutol Hydrochloride (750 mg QD) in concomitant with the intrathecal injection of isoniazid and dexamethasone was prescribed. The next three lumbar punctures during treatment all disclosed normal pressure, mild increased cell count (300–700 * 10^6/L), extremely high

* Corresponding author at: Department of Neurology, National Key Clinical Department and Key Discipline of Neurology, The 1st Affiliated Hospital, Sun Yat-sen University, No. 58 on Zhongshan 2nd Road, Guangzhou, Guangdong 510080, China.
E-mail address: chen12@mail.sysu.edu.cn (L. Chen).
[#] These authors contribute equally to this work.

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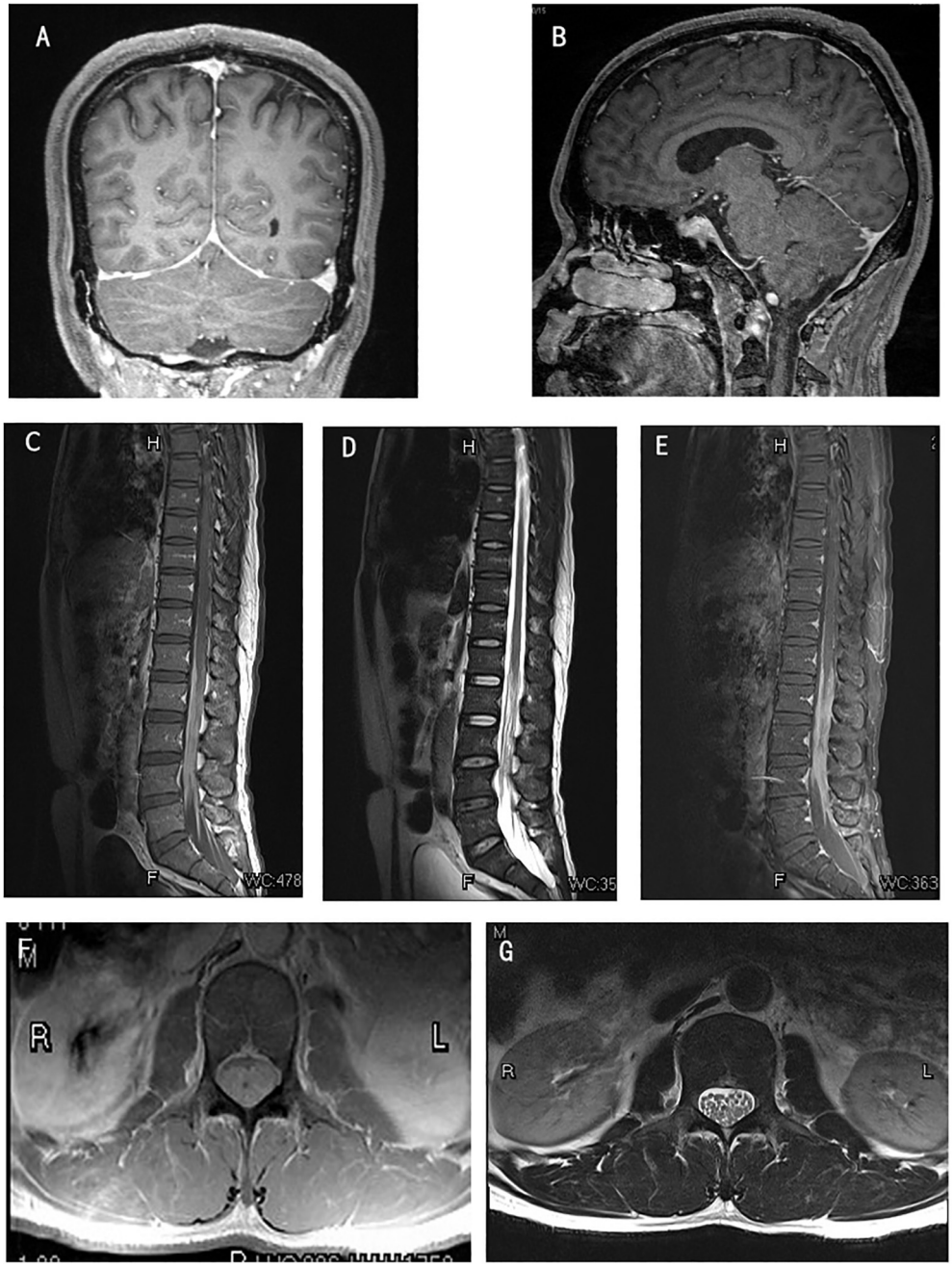

Fig. 1. Coronary (A) and sagittal (B) MRI showing enhancement of meninges. The swollen cauda equina was of contrast enhancement (C-E) and disclosed sagitally on T1-weighted (C), T2-weighted (D) and flair sequence (E), as well as transversely on T1 weighted (F) and T2-weighted (H).

quality of protein (an average of 25,000 mg/L) in the CSF. After two weeks of anti-tuberculosis therapy, no improvement had been approached and the patient was even admitted in our ICU due to respiratory failure.

On examination in ICU, he was alert and orientated. Neurological examination demonstrated complete left peripheral facial paralysis, dysphagia, bucking, paraparesis and amyotrophy in all four limbs with flaccidity. The rest of neurological examination was unremarkable. The laboratory tests for blood WBC, lymphocyte, erythrocyte sedimentation rate (ESR), antineutrophil cytoplasmic antibodies (ANCA), malignancy tumor biomarkers, tubercular antibody (PPD-IgG), T-spot and HIV were all negative. The serum lactate dehrdrogenase (LDH) was slightly increased. CT of the neck, chest, abdomen and pelvis were all negative.

The repeated epidemiology consult after admitted in ICU excluded the diagnosis of tuberculosis and the anti-tuberculosis drugs were then suspended. The fifth lumbar puncture suggested too little CSF to gain, which might attribute to the excessive swollen cauda equine. We hence creatively improved the conventional methods of lumbar puncture. Two needles were punctured separately but simultaneously in the L3/4 and L4/5 intervertebral spaces. Isotonic saline (0.9%) was injected from L3/4 to wash the cauda equine and diluted CSF was gained at L4/5(Fig. 2a). Cytological examination of CSF revealed abundant of lymphocytes with macronucleoli (Fig. 2b and c). Hematology consult was placed and suggested PCNSL. The flow cytometric analysis was required for definitive diagnosis. Unfortunately, the patient and his family refused further medical intervention for economical reasons. He

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