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Letter to Editor

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Diffuse leptomeningeal glioneuronal tumor (DLGNT) with hydrocephalus as an initial symptom mimicking tuberculous meningitis: A case report

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Leptomeningeal enhancement involves the subarachnoid spaces. The etiologies include infection, inflammation or neoplasms. Neoplasms can be primary or secondary leptomeningeal metastasis from tumors such as the breast, lung, ovarian malignancies, lymphoproliferative disorders, or melanoma.[1]

Diffuse leptomeningeal glioneuronal tumor (DLGNT), a recently characterized neoplastic entity by World Health Organization (WHO) (2016), shows the characteristic imaging appearance of diffuse leptomeningeal enhancement and multiple sub-pial cysts.[2]

Hydrocephalus can be the initial presentation. The imaging appearance and indolent clinical features may mimic chronic meningitis like tuberculosis (TB).[3-5]

Our case was a 38-year-old man presented with headache on April 2021. In another center, initial brain computerized tomography (CT) scan and brain magnetic resonance imaging (MRI) showed acute hydrocephalus and diffuse leptomeningeal involvement more in basal cisterns.

The patient underwent ventriculoperitoneal

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shunt and stereotactic biopsy of cerebellar mass, and symptoms improved. 10 days later, he had severe neck pain and weakness in his right limbs, and after one week, he returned to our center. At the initial examination, he was alert and oriented. Saccadic and pursuit movements of eyes were impaired. Pupils were reactive to light. Muscle forces of upper and lower extremities were 5/5. He had right side clumsiness, scanning speech, bilateral abnormal finger-to-nose testing (FTN) and Heel to Shin (HTS) tests, and both plantar reflexes were up. His gait was wide base, and could not walk without help.

The report of cerebellar biopsy was acute inflammatory response without granuloma.

We performed brain and whole spine MRI which showed innumerable enhancing nodules in basal cisterns with extension to posterior fossa, to sulci of cerebellar hemispheres, and also in all of the cerebrospinal fluid (CSF) spaces (Figure 1).

A markedly enhancing intramedullary mass was noted in cord at C7-T1 level associated with extensive leptomeningeal enhancement in cervical spine (Figure 1).

There was also a nodular extramedullary mass in right side of cervical canal at C2-C3 level. Several enhancing mass lesions were depicted in bony structures in spine involving vertebral bodies and posterior elements. Diffuse syrinx formation along thoracic cord down to T10-T11 level was noted (Figure 1). Multilevel patchy dural and extradural enhancing components at thoracolumbar spine were evident with nodular components extend to sacral canal.

The bone marrow biopsy was normal. Malignancy work-up including chest and abdominopelvic CT scan, testis ultrasonography, tumor markers, and whole-body pet-CT scan were normal. In CSF analysis: white blood cell (WBC): 0, protein (Pr): 159 g/l, and glucose (GLU): 103 mmol/l. CSF cytology, TB polymerase chain reaction (PCR) and culture, Cryptococcus PCR, Brucella PCR were twice negative.

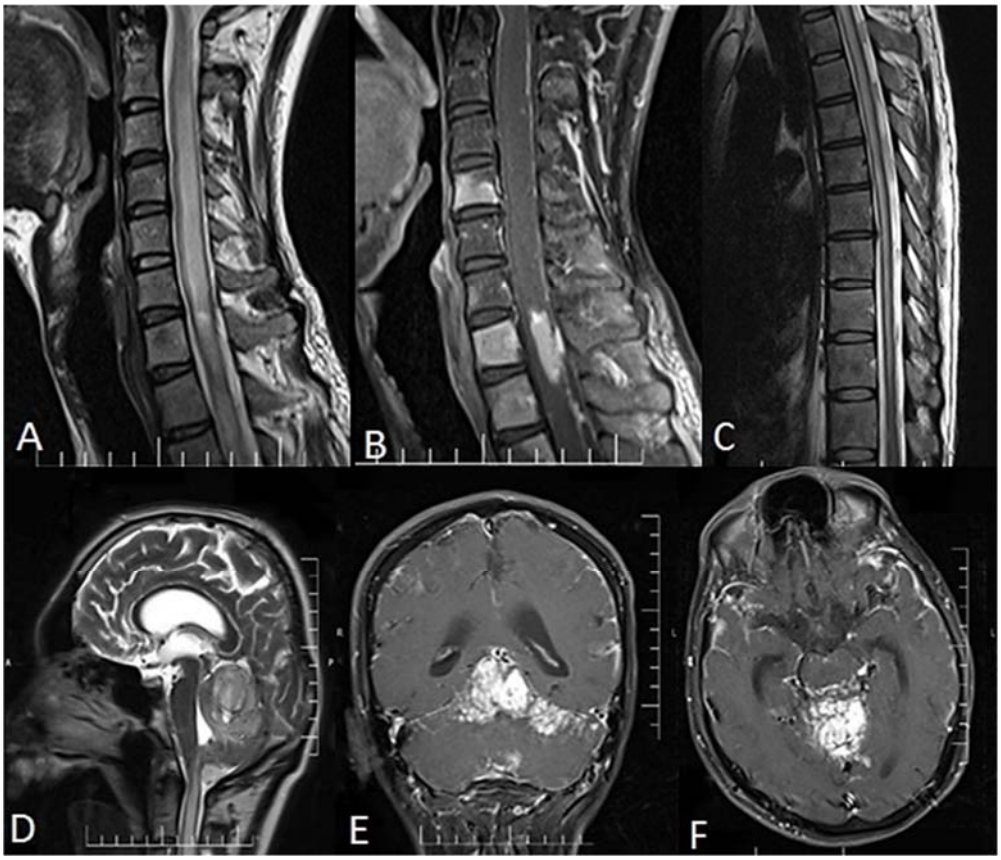

Figure 1. Cervical magnetic resonance imaging (MRI) showing markedly enhancing intramedullary mass with extensive leptomeningeal enhancement (A, B), diffuse syrinx formation along thoracic cord down to T10-T11 level (C), and innumerable enhancing nodules in basal cisterns with extension to posterior fossa and to sulci of cerebellar hemispheres (D, E, F)

Due to nodular patchy leptomeningeal enhancement with probable diagnosis of TB, he was discharged with four-drugs anti-TB treatment. 2 days later, he presented with aphasia and right hemiparesis. In brain CT, left temporoparietal intracranial hemorrhage (ICH) was detected. In control brain CT, he became hydrocephalus again due to shunt failure with persistent hiccups and shortness of breath in sleep. The patient then transferred to intensive care unit (ICU) and intubated. Meningeal biopsy was performed. In physical exam, he became vegetative and unfortunately died 4 months after onset of symptoms. In pathologic report, received specimen in formalin consists of multiple fragments of creamy-tan soft tissue. In microscopic description, sections show cerebellar tissue involved by a neoplasm showing sharp demarcation which included rather uniform tumor cells with round nuclei, fine chromatin, and inconspicuous nucleoli with eosinophilic clear cytoplasm arranged in sheets or vague pseudorosette pattern. Background was myxoid. Focal areas of increase in cellularity with vascular proliferation were evident showing some mitotic feature. In immunohistochemistry (IHC), olig2 was strongly positive in atypical cells, synaptophisin and sox10 were also positive. The overall histomorphology, IHC, and correlated imaging findings were in keeping with DLGNT.

The common manifestations of DLGNT include headache, altered mental status, behavioral abnormalities, paraphasia, and hydrocephalus.[3] Leptomeningeal thickening and enhancement, along with multiple sub-pial cysts involving the cerebrum, brainstem, and spinal cord, have been reported. As in our case, intramedullary lesions are also another feature of DLGNT.[2]

Diffuse leptomeningeal enhancement has a large differential diagnosis. Among all the causes, bacterial and viral meningitis are the most common, showing thin linear meningeal enhancement in contrast to fungal and carcinomatous meningitis which often demonstrate thick and nodular enhancement.[1]

Thus in an endemic region, thick nodular meningeal enhancement with hydrocephalus evokes a diagnosis of tuberculous meningitis (TBM), which is remarkably similar to DLGNT, as our case.

In conclusion, DLGNT is a recently characterized neoplastic entity which should be included as a differential diagnosis of chronic meningitis such as TB, sarcoidosis, and other meningeal infiltrating tumors.

Conflict of Interests

The authors declare no conflict of interest in this study.

Acknowledgments

None.

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