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Case Report
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A Rare Cause of Large Cervical Spinal Cord Edema in the Absence of an Intramedullary Mass: Primary Diffuse Leptomeningeal Glioneuronal Tumor

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Abstract

Introduction: Primary Diffuse Leptomeningeal Glioneuronal Tumor (DLGNT) is a rare tumor of the central nervous system. MRI of the tumor usually reveals diffuse leptomeningeal thickening, leptomeningeal contrast enhancement and hydrocephalus without any parenchymal lesion. In these patients, the predominant clinical findings are related to hydrocephalus. Spinal cord findings are more diverse than intracranial findings.

Case Report: A 52-year-old female patient presented with a complaint of hydrocephalus. Her brain MRI showed hydrocephalus and diffuse hyperintense area between C2 and C7 in the spinal cord, consistent with edema.

Conclusion: The diagnosis PDLG is difficult due to the variety of clinical findings and the rate of antemortem diagnosis is very low. We present a case with long segment cervical spinal cord edema without an intrinsic tumoral lesion, which is, as far as we know, not reported previously.

Keywords: Glioneuronal tumor; Spinal cord edema; Hydrocephalus; Leptomeningeal contrast enhancement

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Case Presentation

A 52-year-old female patient complained of poor appetite, nausea, and vomiting. Gait disturbance was added thereafter, and she became unable to walk without support. Her neurological examination showed limited abduction in her left eye and ataxic gait. Her routine blood tests did not reveal any pathology. Her brain MRI showed hydrocephalus as well as leptomenigeal thickening and contrast enhancement around the brainstem Figure 1. T2-weighted spinal MR images revealed a diffuse hyperintense area between C2 and C7 in the spinal cord, consistent with edema. Post-contrast images revealed diffuse leptomeningeal contrast enhancement across spinal cord and prominent contrast enhancement in central anterior median fissure Figure 1, 2. Diffusion-weighted images revealed slightly increased diffusion in the cervical spinal cord lesion Figure 1. T2-weighted images revealed millimetric hyperintense lesions inside the thecal sac at L1 and L3 levels, which showed nodular contrast enhancement in post-contrast images Figure 2. Sedimentation rate, CRP, vasculitis and tumor markers, *Brucella*, hepatitis markers, *Treponema pallidum* test and ACE level were normal. Cerebrospinal Fluid (CSF) analysis showed a pressure of 40 cmH_2O, increased protein (350 mg/dl), low glucose level (60 mg/dl, concurrent blood glucose 110), and no cells. CSF mycobacterium PCR, *Brucella* and Lyme antibody, paraneoplastic panel were normal. Thoracoabdominal CT and PET scans did not reveal any abnormality. A 1 gr/day pulse steroid therapy was administered for five days as well as topiramate and acetazolamide to reduce hydrocephalus. Her ataxia resolved, left eye which was in midline, began to have abduction. She was discharged with oral steroid but hospitalized again due to nausea, vomiting and decreased oral intake. Her consciousness worsened to confusion. She had repetitive generalized tonic-clonic seizures and bilateral 6^{th} cranial nerve paralysis. She did not respond to pulse steroid and diuretic therapy. L1 lesion was removed by the neurosurgeon using laminectomy. Gram and ARB staining did not reveal any microorganism; there was no growth in cell culture. While her pathological assessment was pending, her consciousness deteriorated to lethargy and she died afterwards. Histopathological evaluation revealed a tumoral structure consisting of uniform oligodendroglioma-like cells with round nuclei, transparent or

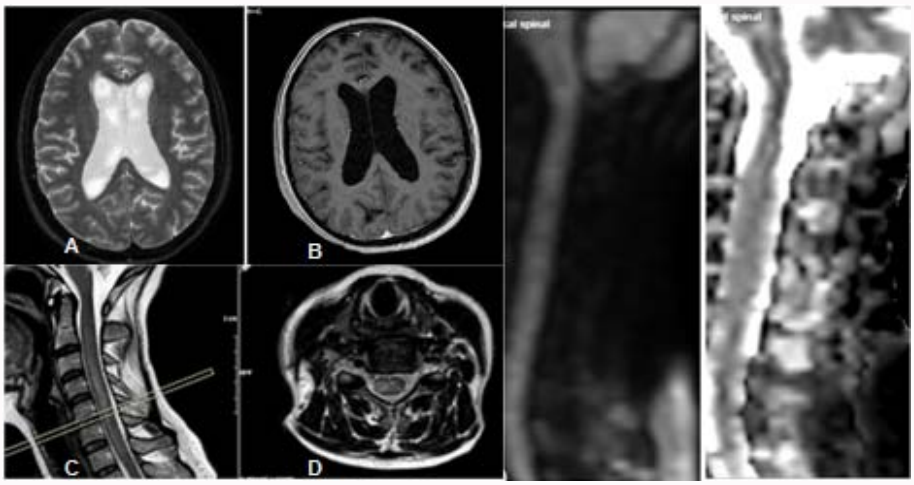

Figure 1: A/B: Hydrocephalus is seen on T2-weighted axial section and contrast-enhanced T1-weighted axial image. C/D: The brain parenchyma is normal. Diffuse hyperintensity of the spinal cord between C2-C7 on T2-weighted sagittal and axial images consistent with edema. E/F: On the sagittal DWI image and the corresponding ADC map a slight increase in diffusion is observed in the cervical spinal lesion.

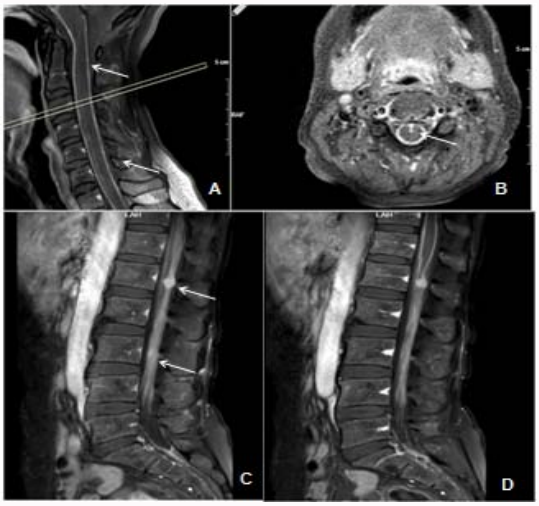

Figure 2: A/B: Contrast-enhanced T1-weighted sagittal and axial sections show diffuse leptomeningeal thickening and enhancement along the medulla oblongata and spinal cord. In addition, significant enhancement is observed in the anterior median fissure (arrows). C/D: In post-contrast T1-weighted sagittal sections and millimetric nodular enhancing lesions (arrows) are observed in the thecal sac at L1 and L3 levels. In addition, diffuse leptomeningeal enhancement is observed in the lumbar spine.

pink cytoplasm in the microvascular stroma. Immunohistochemical staining showed diffuse, strong immunoreactivity with GFAP and S100. Ki-67 proliferation index was about 3%. Thus, the diagnosis of PDLG tumor was established. The biopsy specimen was too small to perform a molecular examination.

Discussion

Primary Diffuse Leptomeningeal Glioneuronal Tumor (DLGNT) is a rare tumor of the central nervous system that has been recently included to 2016 revision of WHO classification of central nervous system tumors in the category of neuronal and mixed glial-neuronal tumors. Magnetic Resonance Imaging (MRI) of the tumor usually reveals diffuse leptomeningeal contrast enhancement and hydrocephalus [1]. In addition to the above-mentioned conventional MRI findings, our case had diffuse edema affecting a long segment in cervical spinal cord without an intrinsic mass lesion. Increased intracranial pressure due to communicating hydrocephalus is the major clinical finding in DLGNT [2]. MRI findings are important for diagnosis. Conventional MRI findings of DLGNT are leptomeningeal contrast enhancement especially in the basal cisterns and throughout the spinal cord, which can be confused with tuberculous and other types of meningitis, and accompanying hydrocephalus [3,4]. The only specific radiological finding described for this tumor is subpial cysts which can only be seen in some of the patients [5]. It is of interest that various spinal cord lesions are described in DLGNT tumor patients in the literature whereas brain lesions are somewhat the same limited number of lesions. Many radiological findings such as intramedullary predominantly cystic lesion without spinal leptomeningeal contrast enhancement, enhancing intramedullary lobulated mass, small enhancing intramedullary lesions, subpial cysts, and extramedullary nodular lesions were described in the spinal cord [5-7]. Our case showed diffuse leptomeningeal contrast enhancement which was most prominent in the anterior median fissure and two tiny enhancing extramedullary nodular lesions. In addition to these classical findings, the most striking finding in our case was long segment spinal cord edema exceeding two vertebral bodies although there was no intramedullary tumoral lesion. We suggest that the most appropriate mechanism for the formation of long segment T2-hyperintense lesion is that diffuse leptomeningeal involvement prevents venous return and edema occurs in the spinal cord as a result of increased capillary pressure. In diffuse leptomeningeal involvement, parenchymal infarcts can also be expected due to alteration of the arterial supply. In our patient, diffusion restriction was not observed in the cervical spinal cord in DWI. On the contrary, increased diffusion was observed, so acute and subacute infarctions were ruled out. Another possible mechanism is that DLGNT may have made a diffuse microinvasion into the spinal cord through the perivascular spaces which is difficult to detect with MRI. This could be demonstrated by postmortem pathological examination, but this was not possible in our case. Lesions can be observed in the brain and spinal cord as a result of paraneoplastic effect, however, in this case antibody-mediated damage in myelin and neuronal elements and associated clinical findings are expected. Our patient did not have transverse myelitis findings, which ruled out the aforementioned mechanism.

Conclusion

Our case showed that increased T2 signals can be seen in the spinal cord in DLGNT. Although MR images are suggestive of transverse myelitis, they might be misleading. As in our case, the absence of clinical findings of transverse myelitis in patients with myelitis findings on MRI and diffuse leptomeningeal contrast enhancement should suggest DLGNT.

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