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Neuroradiology

Communicating hydrocephalus and coexisting nonenhancing tumor: An ominous sign for patients with neurofibromatosis type 1?

Ryo Miyahara MD^a, Satoshi Tsutsumi MD^{a,*}, Satoshi Adachi MD^b, Hisato Ishii MD^a, Akihide Kondo MD^b, Yukimasa Yasumoto MD^a

^a Department of Neurological Surgery, Juntendo University Urayasu Hospital, 2-1-1 Tomioka, Urayasu, Chiba, 279-0021, Japan

^b Department of Neurological Surgery, Juntendo University School of Medicine, Tokyo, Japan

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ABSTRACT

A 26-year-old woman with familial neurofibromatosis type 1 sustained headache that worsened for 1 month. Neuroimaging revealed a mild ventriculomegaly and nonenhancing lesion in the pons. In spite of repeated cerebrospinal fluid examinations and magnetic resonance imaging, the etiology was not determined. The affected pons markedly enlarged in the following 2 months, with extensive leptomeningeal dissemination. Biopsy through hemilaminectomy of the T9 was diagnosed as glioblastoma multiforme. Prompt histologic examination should be performed when patients with familial neurofibromatosis type 1 manifest communicating hydrocephalus coexistent with a nonenhancing tumor.

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Introduction

Glioblastoma multiforme (GBM) is one of the most challenging malignancies to treat, with a 5-year survival rate of 5% [1,2]. It infrequently presents leptomeningeal dissemination as a secondary manifestation [3–5]. In extremely rare instances, it presents with primary leptomeningeal GBM [6,7]. Patients with neurofibromatosis type 1 (NF1) are documented as predisposing to GBM and leptomeningeal gliomatosis [8,9]. Hydrocephalus

in these patients is commonly obstructive around the aqueduct [10]. Here we present a young adult with NF1 who sustained communicating hydrocephalus and nonenhancing pontine tumor and was eventually diagnosed with GBM.

Case presentation

A 26-year-old woman with familial NF1 sustained headache that worsened for 1 month. Central nervous system tumors

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* Corresponding author.

E-mail address: shotaro@juntendo-urayasu.jp (S. Tsutsumi).

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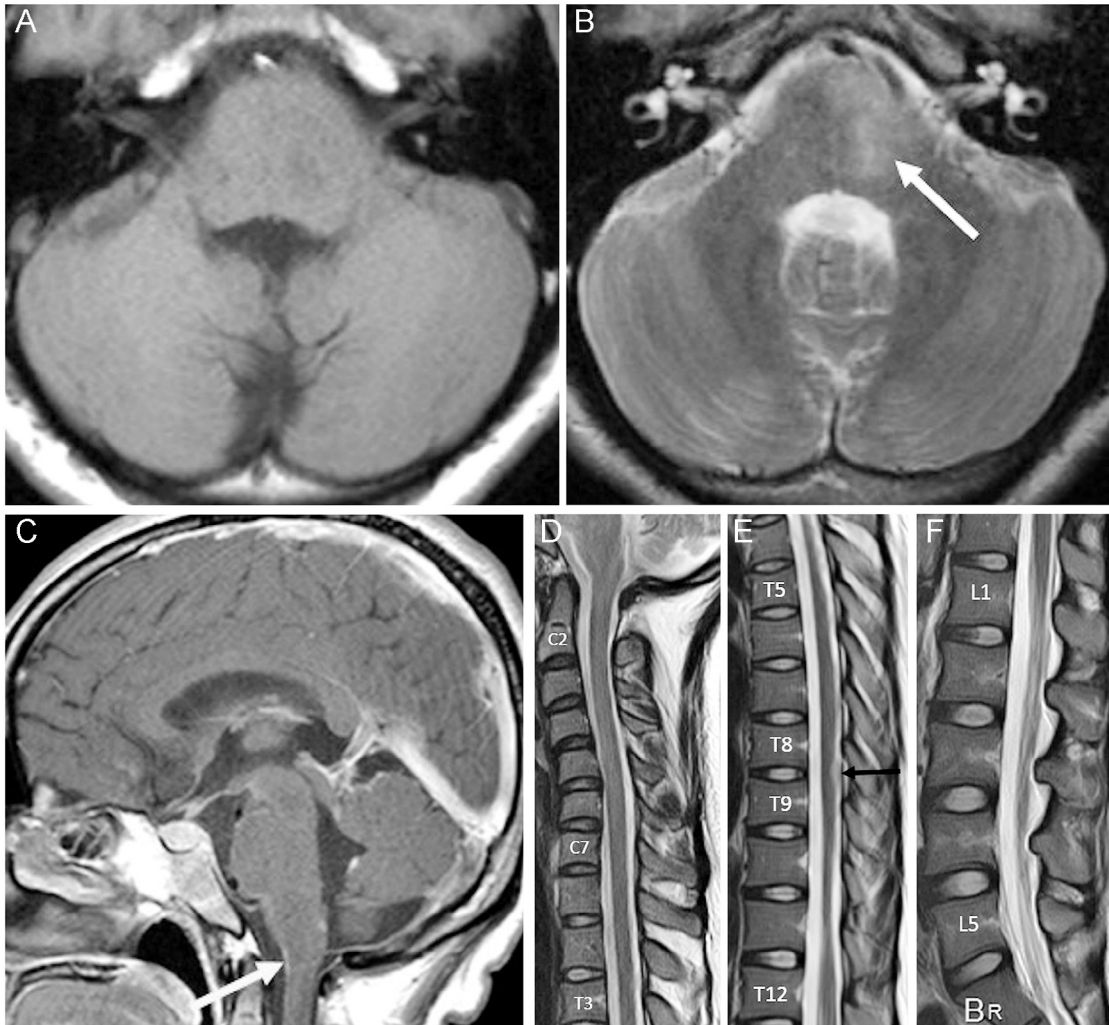


Fig. 1 – Axial T1- (A) and T2-weighted images, (B) postcontrast sagittal T1-weighted image of the brain (C), and sagittal T2-weighted images of the spine (D-F) at presentation showing nonenhancing pontine lesion (B arrow), linear enhancement along the ventral medulla (C arrow), and a nodular lesion between the T8 and T9 levels, on the dorsal surface of the cord (E arrow). The aqueduct is patent (C).

had not been identified in her. At presentation, the patient showed right abducens nerve palsy but did not show other clinical symptoms including meningeal irritation. Cerebral magnetic resonance imaging (MRI) revealed a mild tetraventriculomegaly with patent aqueduct, linear enhancement along the ventral medulla, and a coexisting nonenhancing mass in the pons (Fig. 1A-C). Spinal MRI found a small nodular lesion between the T8 and T9 levels on the dorsal surface of the cord (Fig. 1D-F). At the time, contrast examination was not performed for an unknown reason. Lumbar cerebrospinal fluid (CSF) tap confirmed a markedly elevated intracranial pressure of 65 cmH₂O. The cell count of CSF was 11 per microliter, whereas the protein and glucose levels were 400 mg/dL and 50 mg/dL, respectively. CSF cytology identified only a few lymphocytes with atypia.

In spite of repetitive CSF examinations and MRIs, the etiology was not determined. However, MRI performed 2 months later revealed an enlargement of the pons (Fig. 2A-C) and extensive leptomeningeal dissemination over the cerebrospinal axis (Fig. 2C-F). Biopsy through a hemilaminectomy of the T9

identified a subdural tumor that was grayish in color, elastic hard, and moderately vascular. Microscopically, the tumor comprised highly atypical cells with prominent pleomorphism (Fig. 3). Immunohistochemically, the tumor cells were diffusely positive for glial fibrillary acidic protein. The MIB-1 index was 30%. O⁶-methylguanine-DNA methyltransferase promoter methylation was present, whereas mutations in isocitrate dehydrogenase 1 and isocitrate dehydrogenase 2 were not identified. These were consistent with GBM. The patient underwent chemoradiation therapy postoperatively.

Discussion

Based on the clinical and pathologic findings, the present case was thought as leptomeningeal GBM sustaining communicating hydrocephalus and nonenhancing pontine tumor. Such nonenhancing parenchymal tumors are a rare entity, charac-

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