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Normal Pressure Hydrocephalus as an Unusual Presentation of Supratentorial Extraventricular Space-Occupying Processes: Report on Two Cases

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Key Words
Normal pressure hydrocephalus · Meningioma · Glioblastoma multiforme

Abstract
Normal pressure hydrocephalus (NPH) is a clinical and radiographic syndrome characterized by ventriculomegaly, abnormal gait, urinary incontinence, and dementia. The condition may occur due to a variety of secondary causes but may be idiopathic in approximately 50% of patients. Secondary causes may include head injury, subarachnoid hemorrhage, meningitis, and central nervous system tumor. Here, we describe two extremely rare cases of supratentorial extraventricular space-occupying processes: meningioma and glioblastoma multiforme, which initially presented with NPH.

Introduction

Normal pressure hydrocephalus (NPH) can be idiopathic or secondary. The latter may occur in the setting of head trauma, subarachnoid hemorrhage (SAH), meningitis, and central nervous system (CNS) tumor [1, 2]. The present report describes two unusual cases of supratentorial extraventricular space-occupying processes: meningioma and glioblastoma multiforme (GBM), which initially presented with NPH.

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Case Reports

Case 1

A 58-year-old female patient was referred to our clinic with a 6-month history of progressive gait disturbance and urinary incontinence. The neurological exam showed the full clinical picture of NPH syndrome, including remarkable dementia which had been completely ignored by the patient and relatives. The computed tomography (CT) scan demonstrated marked ventriculomegaly as well as a well-delineated left-side temporoparietal lesion, consistent with a convexity meningioma (**fig. 1**a, b). The ophthalmological examination showed no abnormalities. However, lumbar tap was not performed because of the presence of the intracranial space-occupying process. The patient was operated on in March 2010, with total removal of the tumor. Histological examination revealed World Health Organization (WHO) grade I meningioma (**fig. 2**). Transient right-hand paresis appeared in the early postoperative period, with no change in the NPH symptoms. The patient was discharged and scheduled for ventriculoperitoneal shunt placement. However, her family refused any further surgical interventions. Surprisingly, 1 month later, the patient presented with significant improvement of the gait difficulties and urinary incontinence. The control CT examination showed no data for residual tumor as well as a slight decrease of the ventricular size (fig. 1c). No additional dynamics of the patient's neurological status were noted until January 2011.

Case 2

A 68-year-old male patient was referred to the clinic with an 18-month history of involuntary left arm movements and subsequent development of progressive gait disturbances and urinary incontinence. The neuroimaging studies made before the admission demonstrated communicating hydrocephalus without any clinically significant lesions within the brain parenchyma (**fig. 3**a, b). The ophthalmological examination was normal. Just prior to the hospitalization, the patient started to complain of progressive left arm weakness. The control CT scan revealed a heterodense tumor situated in the right frontotemporal region with compression and dislocation of the adjacent brain structures to the left (fig. 3c). Partial excision of the lesion was performed in June 2010, using frameless stereotaxy technique. Histologically, the tumor demonstrated typical features of GBM (WHO grade IV glioma; **fig. 4**). No additional deficit was observed after the intervention. The patient was discharged and scheduled for adjuvant chemoradiation. In spite of the treatment, he died 3 months later from the consequences of GBM.

Discussion

NPH is a clinical and radiographic syndrome characterized by ventriculomegaly, gait disturbances, urinary incontinence, and mental decline. The condition may occur due to a variety of secondary causes including head injury, SAH, meningitis, and CNS tumor [1, 2]. A number of possible mechanisms have been suggested to explain the pathogenesis of adult hydrocephalus that occurs in association with intracranial space-occupying processes [3–8].

The blockage of the CSF outflow through the ventricular system is one of the most common pathways for the development of secondary hydrocephalus in adults. The majorities of cases demonstrate rapidly progressing symptoms of increased intracranial pressure and necessitate urgent intervention [3]. However, in some instances, patients may also develop NPH, as described by Lobato et al. [4] and Sayama et al. [5]. Obviously, this mechanism is not relevant to our cases, as both tumors were extraventricular.

According to many authors, the hyperproteinorrachia, which can be observed in some patients with CNS tumors, may cause NPH by impairing CSF reabsorption at the level of the arachnoid granulations [6]. However, in our cases, this mechanism can only be suspected, because no lumbar puncture was performed before surgery.

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The dissemination of CNS tumor into the subarachnoid space may also lead to NPH. Interestingly, the systematic review of the literature revealed only one such case: a 69-year-old female patient with leptomeningeal gliomatosis, who initially presented with confusion, progressive memory decline, and urinary incontinence. The repeated CT scan demonstrated slight dilatation of the lateral ventricles, which together with the authors' clinical description supports the diagnosis of NPH, although the latter was not specifically proposed [7]. Even possible in our case with GBM, this mechanism remains unproved, because no autopsy was done.

The intracranial tumors may also be associated with recurrent SAHs. The latter may provoke adult hydrocephalus on the basis of a decreased rate of CSF absorption at the arachnoid villi. For example, Chang et al. [8] described a 51-year-old male patient with spinal leptomeningeal metastases from a giant-cell GBM, who consequently developed SAH and NPH. In our cases, however, this mechanism is probably of no significance, because in neither of them subarachnoid bleeding was observed.

Conclusion

The association of supratentorial extraventricular space-occupying processes with NPH is rather uncommon. The exact mechanism of this phenomenon is unclear and requires further investigation.

Acknowledgement

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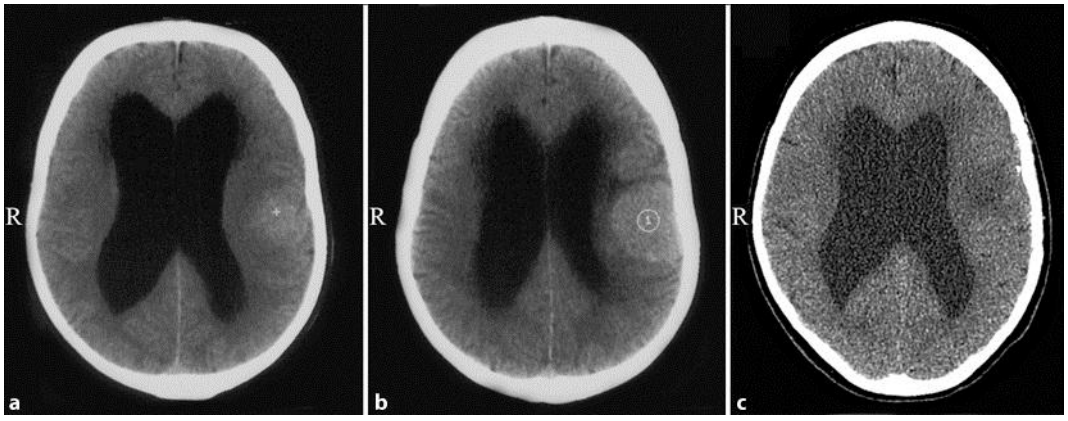

Fig. 1. Neuroimaging of case 1. CT scans before the surgical intervention (**a**, **b**) and 1 month later (**c**).

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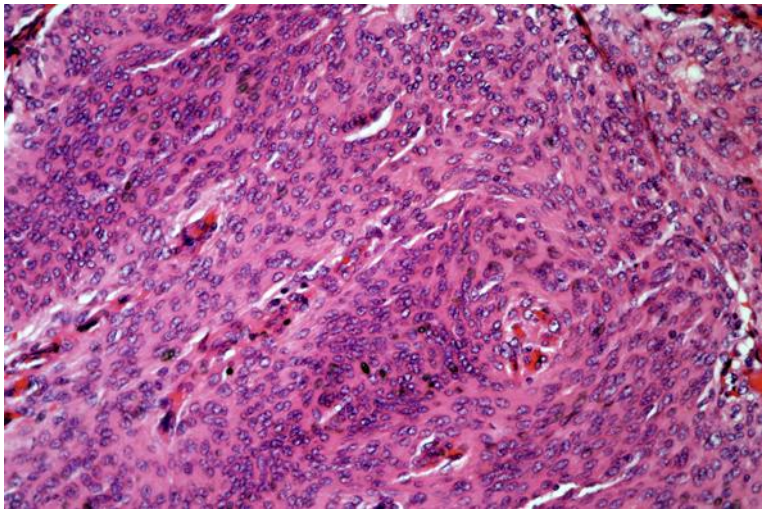

Fig. 2. Histological findings of case 1. The tumor is composed of uniform cells with syncytial growth in sheets and lobules surrounded by thin stroma (WHO grade I meningothelial meningioma). HE, ×200.

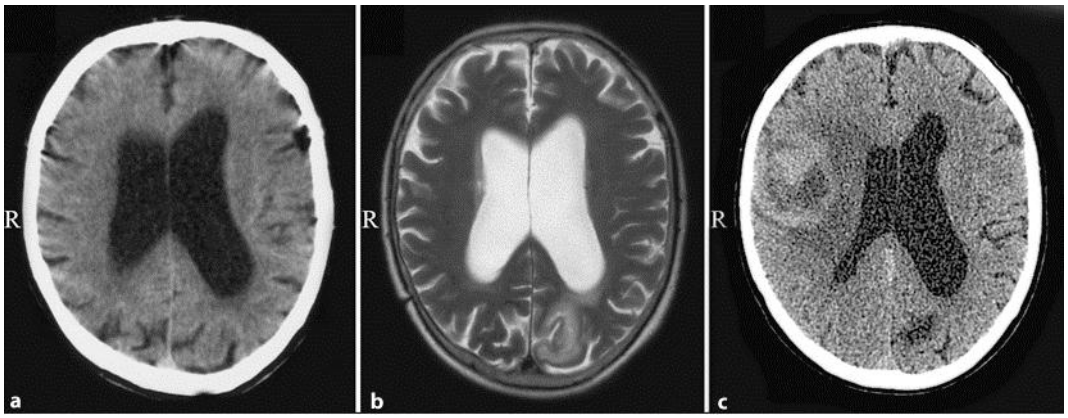

Fig. 3. Neuroimaging of case 2. CT and MRI scans 2 months before the admission (**a**, **b**), and CT scan just prior to surgical intervention (**c**).

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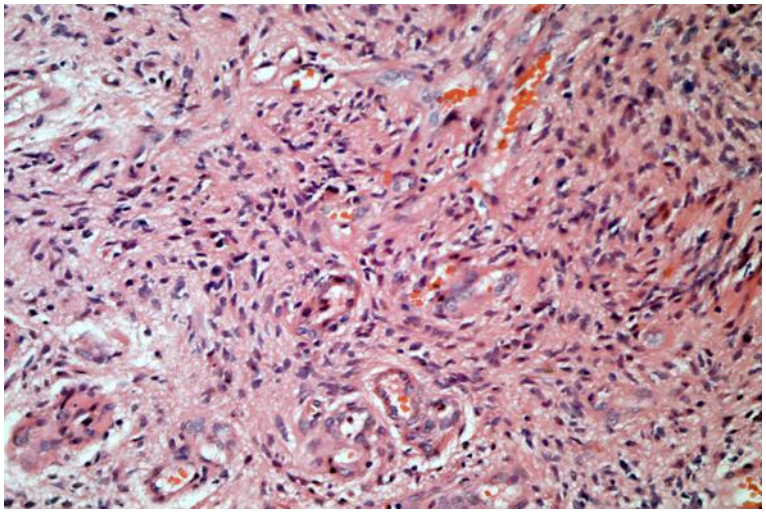

Fig. 4. Histological findings of case 2. The tumor shows prominent cellular polymorphism, microvascular proliferation, and pseudopalisading necroses (WHO grade IV glioma, GBM). HE, ×200.

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