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Central Neurocytoma: Report of a Case

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Abstract

Central neurocytoma is a rare intraventricular tumor recently accepted as a clinicopathological entity.

A 21-year-old female was admitted with three-year history of episodic headaches and vomiting, and with rapid deterioration of her left vision over two weeks prior to admission. Computed tomography (CT) scan revealed a marked hydrocephalus and an isodense, mildly enhancing mass in the left lateral ventricle. On magnetic resonance imaging (MRI) scan, T_1-weighted images revealed an intraventricular mass of slightly high intensity signal, which contained areas of low intensity signal representing multiple intratumoral cysts. The tumor showed a minimal enhancement with Gd-DTPA. A diagnosis of central neurocytoma was confirmed by an electron-microscopic study of a surgical specimen; there were numerous neuronal cell processes containing microtubules and dense-core vesicles, and a few small intercellular junctions were also identified.

Introduction

Most primary brain tumors are derived from glial cells, and true neuronal cell tumors of the central nervous system are rare. Intraventricular tumors are usually derived from ependymal cells or choroid plexus cells. Central neurocytoma is a rare intraventricular neuronal cell tumor, which was for the first time described in 1982 by Hassoun et al.[2]. Although several additional cases have been reported since that time, there are few reports of patients studied by magnetic resonance imaging (MRI) scan. We report a case of central neurocytoma, with special reference to computed tomography (CT) scan and MRI scan features as well as histopathologic findings. The pertinent literature is also discussed.

Case Report

A 21-year-old woman presented with complaints of left visual disturbance, headaches and vomiting. The initial episode of headaches and vomiting occurred three years before. The

Key words: Central neurocytoma, Intraventricular tumor, Computed tomography (CT), Magnetic resonance imaging (MRI).
索引語：神経細胞腫，脳室内腫瘍，CT, MRI.
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headaches gradually increased in intensity, becoming severe over the four months prior to admission. Two weeks prior to admission, she noticed decreased visual acuity in her left eye, which rapidly worsened. CT scan at a local hospital revealed a large intraventricular tumor and severe hydrocephalus, and she was transfered to our clinic for further examination and treatment.

On admission, she was alert and had minimal headaches but no vomiting. Her left visual acuity was finger counting at 30 cm and her left optic disc was pale. Visual acuity and the disc were normal in her right eye, and ocular movements were bilaterally full.

Plain x-ray films of the skull indicated a sustained elevation of intracranial pressure. A noncontrast CT scan revealed a large iso-density mass with multiple intratumoral cysts occupying the frontal horn and body of the left lateral ventricle and extending into the third ventricle. The lateral and third ventricles were also markedly dilated, and the septum pellucidum was displaced to the right side. A post-contrast CT scan showed only mild enhancement of the tumor (Fig. 1). On MRI scan, the tumor showed slightly high signal intensities in T_1-weighted and proton density-weighted images, and it was minimally enhanced with an intravenous injection of Gd-DTPA. The coronal and sagittal images demonstrated to better advantage the anatomical relationship of the tumor and the ventricles (Fig. 2 & 3). The tumor filled the foramen of Monro, extended into the third ventricle, and seemed to infiltrate the upper brainstem.

A right ventricular drainage was performed on an emergency basis, and her visual acuity in the left eye improved to finger counting at 50 cm within three days. Five days after admission, she underwent a bifrontal craniotomy, and the tumor was removed through a transcallosal approach. A gray, soft and suckable tumor occupying the left lateral ventricle was found to have arisen from the septum pellucidum and extended into the third ventricle through the dilated foramen of Monro. The tumor within the lateral ventricle and the anterior portion of the third ventricle was radically removed but a

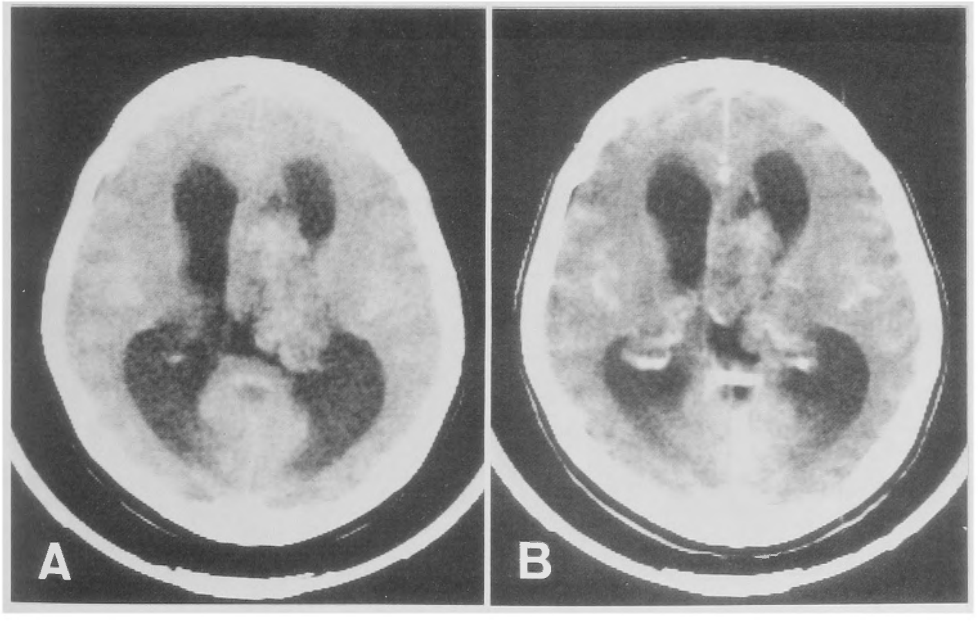

Fig. 1. (A) Plain and (B) contrast-enhanced CT scans, showing a large mass in the left lateral and third ventricles associated with a marked hydrocephalus.

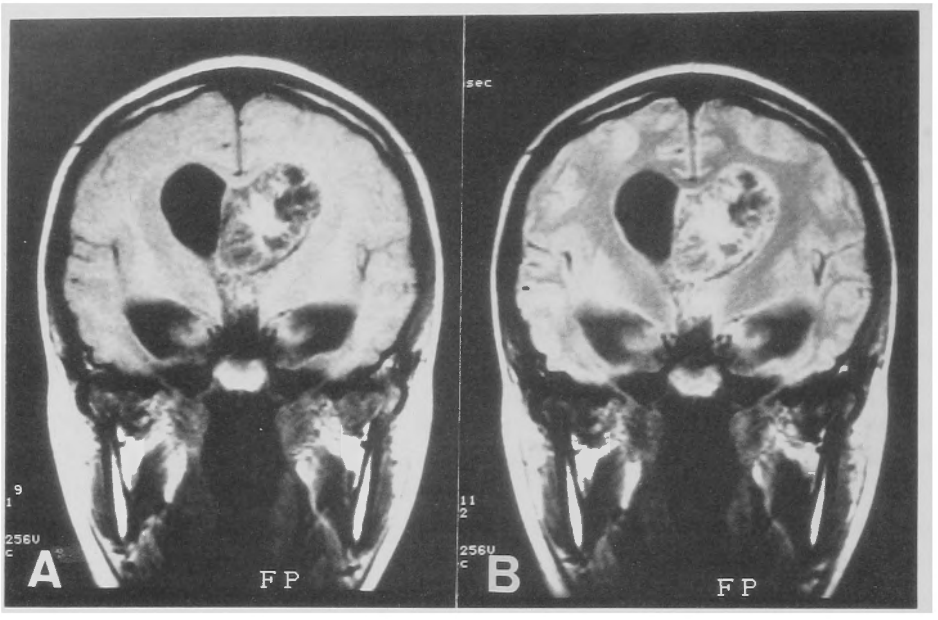

Fig. 2. (A) T_1-weighted (TR/TE＝500/25) and (B) proton density-weighted (TR/TE＝1846/25) MRI scans, showing a mass of heterogeneous signal intensities in the left lateral and third ventricles.

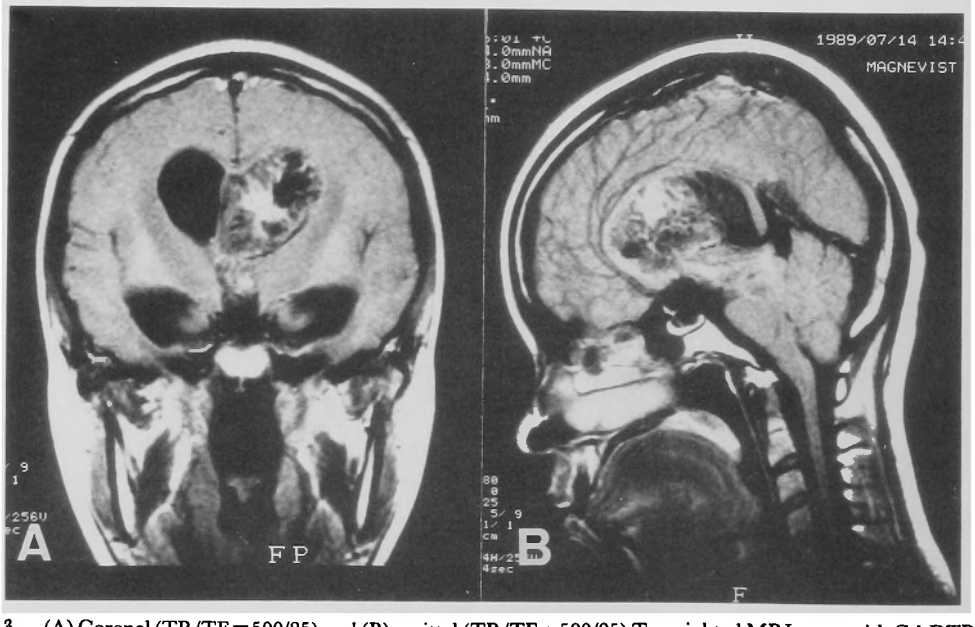

Fig. 3. (A) Coronal (TR/TE＝500/25) and (B) sagittal (TR/TE＝580/25) T_1-weighted MRI scans with Gd-DTPA injection, showing a mild enhancement of the tumor. The caudal portion of the tumor in the third ventricle seems to have infiltrated the upper brainstem.

small portion of the tumor in the posterior portion of the third ventricle was left untouched, as the infiltration into the parechyma had been suggested on MRI scan.

Postoperatively the patient suffered from mutism for three days, but she recovered without neurologic deficits. As the ventricular dilatation progressed, a ventriculo-peritoneal shunting operation was performed two weeks after the craniotomy. She received local irradiation therapy (50 Gy), and was discharged with left visual disturbance (finger counting at 50 cm). At the follow-up visit nine months after discharge, her neurologic condition was stable.

Pathological Findings

Sections of the tumor specimen were stained with hematoxylin and eosin. The neoplasm was composed of monotonous cells. The tumor cells had ovoid nuclei of uniform size. The tumor cell bodies were palely eosinophilic and showed perinuclear haloes. In part, fine fibrillary processes could be detected. Mitoses were rare (Fig. 4).

Immunohistochemical staining was performed for neuron-specific enolase (NSE), glial fibrillary acidic proteins (GFAP) and synaptophysin. The neoplastic cells were reactive for NSE and had some positive sites for synaptophysin. There were few cells positive for GFAP.

Electron microscopy revealed numerous cell processes between neoplastic cell bodies. There were microtubules and dense core vesicles in the cell processes. Some small intercellular junctions were found, but typical synaptic structures were not identified (Fig. 5).

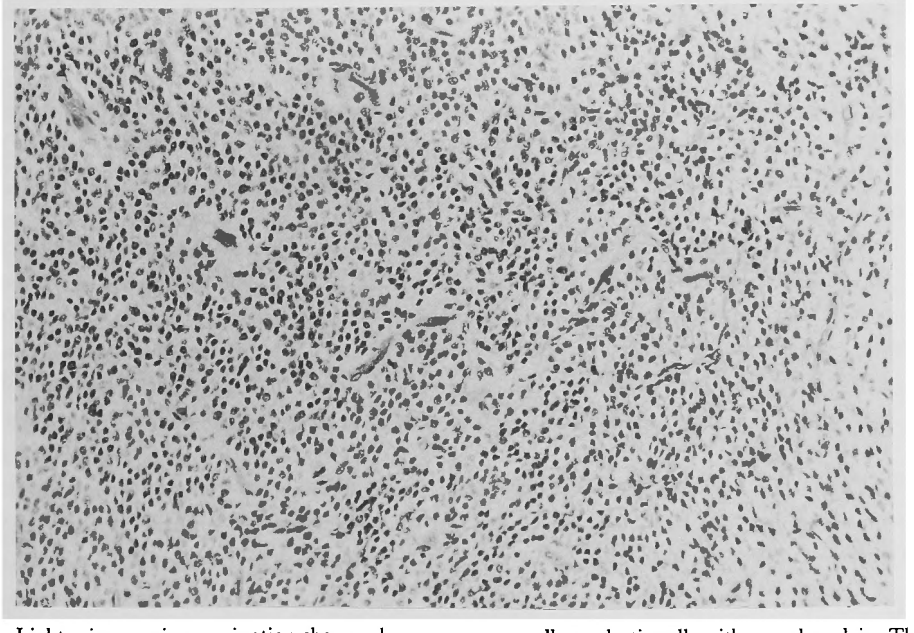

Fig. 4. Light microscopic examination shows a homogeneous small neoplastic cells with round nuclei. The tumor tissue is lobulated by fine fibrillary processes (HE stain, original magnification ×200).

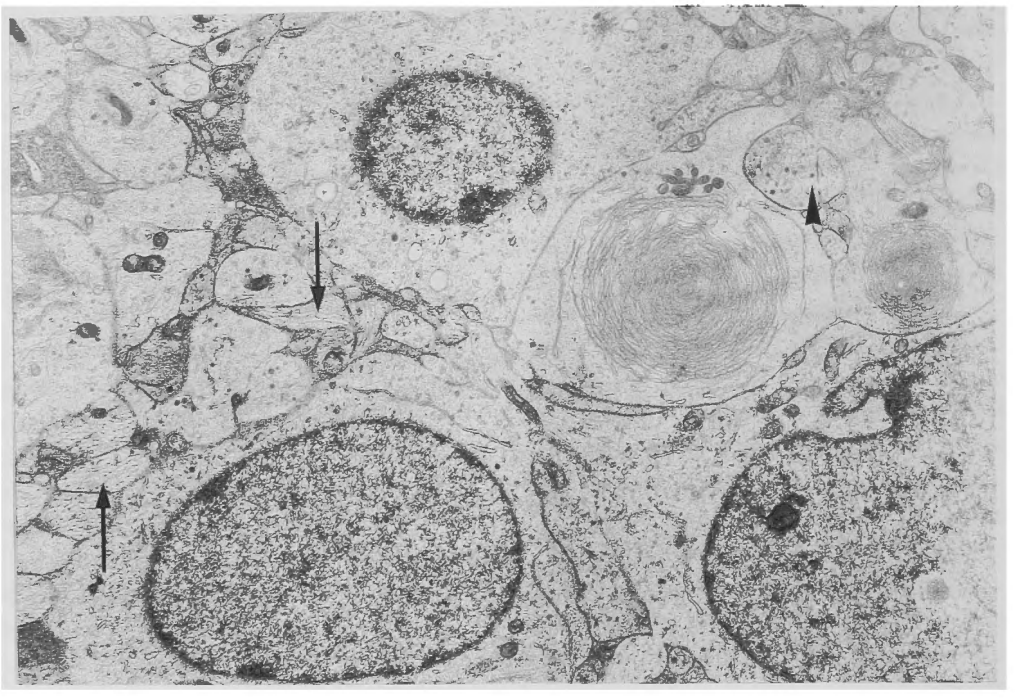

Fig. 5. Electron microphotograph showing numerous cell processes between the neoplastic cell bodies. Microtubules (arrow), dense-core vesicles and clear vesicles (arrowhead) are seen in the cell processes. (Original magnification ×7000).

Discussion

Intraventricular tumors are usually derived from ependymal or choroid plexus cells, and neuronal tumor in this location had been rarely reported. It was 1982 that HASSOUN et al.[2] for the first time reported two cases of neurocytoma in the lateral ventricle and called the tumor "central neurocytoma"

Since that time, several authors have reported additional cases of similar tumors under various names. In 1983, JERDAN et al.[4] reported two adult cases as "differentiated cerebral neuroblastoma" within the ventricles, which showed similar pathological findings as those described by HASSOUN et al.[2]. In 1985, PEARL et al.[10] described three cases of "intraventricular primary cerebral neuroblastoma" in the lateral ventricles and foramen of MONRO. In 1986, OJEDA et al.[8] reported six cases of "primary cerebral neuroblastoma" Three of them occurred in the lateral or the third ventricle.

TOWNSEND et al.[13], POON et al.[11], BOLEN et al.[1], and PATIL et al.[9] also described cases of "intraventricular primary cerebral neuroblastoma" or "intraventricular central neurocytoma" In Japan, TSUJITA, et al.[14], KUBOTA et al.[5], SHIMADA et al.[12], ISHIDA et al.[3], NISHIO et al.[7], and NIOKA et al.[6], reported tumors of this category.

Central neurocytoma typically arises in the lateral ventricles, especially from the septum pellucidum or the fornix. It often causes obstructive hydrocephalus. Patients most often complain of visual distur-

bance and show signs and symptoms of increased intracranial pressure. Our case also presented such typical clinical symptoms and signs, and her tumor arose from the septum pellucidum.

Histologically, central neurocytoma looks like oligodendroglioma under a light microscope. The presence of fine fibrillar matrix between the tumor cells helps to distinguish neurocytoma from oligodendroglioma[3,5]. Immunohistochemical examination is also helpful for differential diagnosis. Neurocytoma cells have not been reported to express GFAP, but our case had positive cells, albeit very few. Such cells are probably trapped reactive astrocytes, as PATIL et al.[9] supposed. In the present case, neoplastic cells were positive for NSE and had some positive sites for synaptophysin, findings compatible with the diagnosis of central neurocytoma.

Immunohistochemical study, however, does not necessarily confirm the tumor to be a central neurocytoma, and the ultrastructural examination is indispensable for the definite diagnosis of neurocytoma. The characteristic electron-microscopic features are numerous neurite processes containing microtubules and dense-core vesicles in the cytoplasm. The presence of more mature neuronal tissue with defined synapses and synaptic junctions also have been reported as the characteristic features[3,5].

As has been reported in the previous reports, CT scan showed an isodense tumor in the present case. The tumor in this patient showed only mild enhancement, however, this is by no means characteristic of central neurocytomas. PATIL et al.[9] described that one tumor enhanced only minimally and no enhancement was seen at all in the other tumor, but several other papers have reported moderate contrast enhancement of the tumor[1,2,7,11]. Although BOLEN et al.[1] described that calcification was common in primary cerebral neuroblastoma, it was not seen in our case.

MRI scan findings of central neurocytoma has been reported rarely. PATIL et al.[9] described that neurocytomas were isointense in both T_1- and T_2-weighted and proton density-weighted images and that they had large draining veins. In our patient, the tumor was mildly hyperintense in T_1-weighted and proton density-weighted images, moderately hyperintense in T_2-weighted images, and the draining vein was not identified. It seems that CT scan and MRI scan are apparently most helpful in the detection and delineation of such an intraventricular tumor, but no CT or MRI features are probably specific to central neurocytoma.

As to the treatment, a total surgical removal should be always attempted. The role of radiotherapy following a partial tumor resection is controversial. Only few cases including ours have so far received readiotherapy[2,10], and long follow-up studies are lacking. At the present moment, radiotherapy is generally thought to be of questionable value, as it has been ineffective in controlling the neuroblastomas of the central nervous system.

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和文抄録

Central Neurocytoma の1例

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Central neurocytoma は，最近病理学的に分類されるようになった稀な脳室内腫瘍である．病理組織学的に central neurocytoma と診断した1例を経験したので報告する．

症例は21才の女性．約3年前より時々頭痛，嘔吐があり，入院の2週間前より左眼の急速な視力低下を来した．CT スキャンでは著明な水頭症と，左側脳室から第三脳室，中脳に及ぶ腫瘍を認めた．腫瘍はほぼ等吸収値を示し，わずかに造影効果を認めた．MRI ではT1 強調画像でやや高信号で，内部に多数の嚢胞と思われる低信号域を伴っていた．Gd-DTPA でわずかに造影効果を認めた．

亜全摘術を行い，光顕的には oligodendroglioma に類似した所見を示した．電顕的には多数の neuronal cell process があり，内部には microtubules や dense core vesicle を認めた．また，small intercellular junction も認め，これらの所見より central neurocytoma と診断した．central neurocytoma は光顕的には oligodendroglioma と極めて類似しており，脳室内腫瘍の診断においては電顕による鑑別が重要と考えられる．