



The typical and atypical MR imaging findings of central neurocytomas: Report on eighteen cases and review of the literature



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ABSTRACT

There were few studies have documented the MRI features of typical and atypical CNCs for diagnosis and therapeutic modalities. Here, 18 histopathologically confirmed cases of intracranial CNCs (8 men and 10 women with a mean age of 28.3 years, range 10–64 years) were retrospectively analyzed. The histopathological and immunohistochemical features were also assessed. On MR imaging, the 14 typical cases of CNCs showed relatively round, lobulated tumor masses in the body of the right lateral ventricle (5 cases), left lateral ventricle (4 cases), third ventricles (2 cases), and midline (3 cases). These typical CNCs masses contained clusters of cysts of varying sizes and “soap bubble” appearance on T2WI; they showed mild to moderate heterogeneously enhancement on T1WI. The 4 atypical cases of CNCs showed as strongly contrast enhancement of the tumors with the attachment or infiltrate of the wall of the ventricle than the typical benign cases. These atypical CNCs were in the right lateral ventricle (2 cases), left lateral ventricle (1 case), and third ventricle (1 case). Microscopically, the typical CNCs were well-differentiated tumors with benign histological features. The typical and atypical CNCs were composed of uniform, small to medium-sized cells with rounded nuclei and scant cytoplasm. Immunohistochemically, the typical CNCs were strong in Syn immunopositive (14/14) and neuron-specific enolase (12/14). The atypical CNC tumor cells showed malignant behavior and more positive expression of Ki67 than the benign cases. Surgery is the first choice of treatment, and radiotherapy may be beneficial to postoperative patients.

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1. Introduction

Central neurocytomas (CNCs) are rare, benign tumors of the central nervous system (CNS). The majority of CNCs are composed of uniform, round cells with neuronal differentiation. The incidence of CNCs is approximately 0.25–0.5% of all intracranial

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tumors [1,2]. Increasing numbers of reports have suggested that these tumors are most common within the ventricles, affect young adults, and are associated with favorable outcomes, without malignant transformation. Approximately one-fourth of these benign tumors has unusually aggressive behavior and are termed “atypical central neurocytomas.” These atypical CNCs exhibit malignant behavior and have an increased tendency to spread through the cerebrospinal fluid, where they cause craniospinal axis dissemination. However, only a few studies have documented the MRI features of typical and atypical CNCs for diagnosis and therapeutic modalities. Here, a retrospective review of the MRI findings of 18 cases with typical and atypical CNCs was conducted and findings were correlated with histopathological features.

2. Materials and methods

The records of patients with CNCs from our hospital's pathology department database from January 2007 to May 2013 were screened. In all, 18 patients (8 men and 10 women with a mean age of 28.3 years, range 10–64 years) with CNCs who underwent preoperative MRI were included in this study. An institutional review board exemption and a waiver of the requirement for written informed consent were obtained for this retrospective study.

All MR studies were performed on a 3.0T MRI (Siemens Trio 3.0T) or a 1.5T MRI (Gyrosan Intera; Philips Medical Systems, Best, the Netherlands) using standard head coils. The MRI protocol included axial T1-weighted sequences; axial and sagittal T2-weighted sequences; and contrast-enhanced axial, sagittal, and coronal T1-weighted sequences. The sequence parameters were as follows: T1-weighted fast field echo (FFE) sequence (TR/TE, 174–291/4.6 ms; slice thickness, 5.0 mm; field of view, 280–512 mm; matrix scan, 256 × 256). T2-weighted turbo-spin echo sequence (TR/TE, 1600–3500/90 ms; slice thickness, 5.0 mm; field of view, 280–380 mm; matrix scan, 256 × 256). An intravenous dose of 0.1–0.2 mmol/kg of contrast agent (Gadolinium-DTPA, Magnevist, Schering) had been administered to the patients undergoing contrast-enhanced MR scanning. Cerebral angiography and MRS was not performed in these patients.

All the medical records of 18 patients were reviewed and patient age, sex, initial symptoms, treatment, follow-up imaging, and outcome were recorded.

The MRI images were reviewed independently by two experienced radiologists. Disagreements over imaging findings between reader 1 and reader 2 were resolved by consensus in a joint reading session to construct the final decision.

The following imaging findings of each scan were recorded: lesion location, shape, size, numbers, and intensity of the unenhanced and contrast-enhanced lesions (classified as hypo-, iso-, or hyperintense with respect to the adjacent tissues).

All 18 cases had undergone surgery. All pathologic specimens were reviewed and described by two experienced pathologists. The histological techniques included routine hematoxylin eosin staining (HE) and immunohistochemical evaluation specific for neuron enolase (Neu-N) and synaptophysin (Syn). The immunohistochemical analysis also included staining for CgA, CD56, S100, EMA, GFAP, Olig-2, CD34, and the specific for detecting Ki-67 antigen present in proliferative cells.

Numerical data such as the size of tumor at CT or MRI measured at the contrast-enhanced images of each transaxial section were averaged for each patient and descriptively compared among patients. All descriptive and statistical analyses were performed using the Statistical Package for the Social Sciences (SPSS version 16.0; Chicago, IL). Variables are presented with standard errors (SE) and high quality authentic CT and MRI scans and histopathological findings are presented.

3. Results

3.1. Clinical data

This study group consisted of 8 men and 10 women with a mean age of 28.3 years (range 10–64 years). All the patients were diagnosed with central neurocytoma by histopathological examinations. Among the 18 CNCs patients, the clinical symptoms included increased intracranial pressure (ICP), headaches (16/18, 88.9%), and vomiting (13/18, 72.2%). One patient was asymptomatic but MR imaging showed accidental head trauma and another patient reported blurred vision. Surgical procedures were performed on all the patients. Gross total removal of the tumor was performed in 14 cases and subtotal removal was performed in 4 cases. Postoperative fractionated radiotherapy (5000–5500cGy) was administered to the four of subtotal removal patients.

3.2. MR imaging of typical central neurocytoma findings

MR imaging generally indicated typical CNCs in 14 cases. They involved relatively round, lobulated tumor masses in the body of the right lateral ventricle (5 cases), left lateral ventricle (4 cases), third ventricles (2 cases), and midline (3 cases). In the T1 and T2 weighted MR images, the tumor masses showed isointense to hyperintense signal intensity. The solitary tissues of the tumor mass were showed heterogeneous hyperintense appearances on DWI. Here, 12 cases showed clusters of cysts of varying sizes that had a “soap bubble” appearance on T2WI. All cases showed mild to moderate contrast enhancement of the tumors with gadolinium. There was mild dilation of the lateral ventricle in 8 cases and moderate dilatation of the third ventricle in 4 cases (Table 1, Figs. 1–4).

3.3. MR imaging of atypical central neurocytoma findings

There were 4 cases of atypical CNCs. All 4 were heterogeneously contrast enhanced solid masses, 2 in the right lateral ventricle, 1 in the left lateral ventricle, and 1 in the third ventricle. These tumors measured from 3.6–4.8 cm in size and more pronounced contrast attachment to the wall of the ventricle than the typical benign cases. The solitary tissues of the tumor mass were showed heterogeneous hyperintense aggressiveness behavior on T2WI and DWI. The tumor mass showed unusual aggressiveness and malignant behavior, such as recurred after sub-total resection in 1 patient, in both the right lateral ventricle and the third ventricle (Table 1, Fig. 5).

3.4. Pathological and immunohistochemistry findings

CNCs tumor tissue is gray in color and friable with necrosis and cystic degeneration areas. Intratumoral hemorrhage is not typical. Microscopically, CNCs were well-differentiated tumors with benign histological features. They may display varied histological architecture even within the same specimen. All the CNCs studied here were composed of uniform, small to medium-sized cells with rounded nuclei, finely stippled chromatin, inconspicuous nucleoli, and scant cytoplasm (Fig. 6A). Immunohistochemically, the CNCs were strong in Syn immunopositive (18/18, Fig. 6B). Neuron-specific enolase (Neu-N) immunostaining was positive in 16 cases (16/18, Fig. 6C), and Ki67 showed more positive expression (> 10%) in 4 atypical CNCs patients (4/18). Glial fibrillary acidic protein (GFAP), CD56, CD34, Olig-2, and S-100 immunostaining were positive and ranged from 30 to 50%. CgA was immunonegative in all cases (Table 2).

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