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Volumetric analysis of a rhabdoid meningioma during preoperative follow-up
A case report

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Abstract

The authors describe a case of rhabdoid meningioma (RM) in a 17-year-old boy that was determined by measuring the tumor volume during preoperative follow-up. The volume of the tumor, located in the left occipital lobe, was measured every 1-5 months, using an image analysis software. The tumor volume doubling time (Td) ranged from 1.0 to 4.9 years in the first 11 months, but became 0.3 years in the last two months. The tumor grew rapidly in the last two months at which time surgery was performed. Pathological examination of the surgical specimen showed that the tumor contained rhabdoid cells (RCs). RCs were heterogeneously distributed in the tumor, admixed with spindle-shaped cells. The areas where RCs were predominant had malignant histological features, with necrosis and high proliferation indices, whereas the areas with few RCs lacked the malignant features. The tumor grew slowly in the initial phase, possibly because components with low proliferation rates occupied most of the tumor. The tumor began to grow rapidly when the malignant component containing abundant RCs became predominant. To the authors' knowledge, this is the first report monitoring the volumetric change of RM periodically. Our investigation indicated that volumetric analysis is useful to decide surgical intervention of the meningiomas with potential malignancy.

Key words : Rhabdoid meningioma ; the volumetric analysis ; immunohistochemistry ; ultrastructure.

Introduction

Meningiomas are generally benign slowly growing tumors, composed of neoplastic meningothelial cells attached to the dura mater. Fifteen histological variants of meningiomas have been defined by the revised version of the new World Health Organization (WHO) classifications (2007), some of which are associated with aggressive biological behavior (16). Rhabdoid meningioma (RM) is a rare variant of meningioma composed of rhabdoid cells, large round cells with abundant eosinophilic cytoplasm and eccentric nuclei. RM corresponds to WHO grade III, and usually exhibits malignant histological features, such as high mitotic rates or necrosis. However, few cases of RM have been reported, so the clinical course is still unclear (1, 3-6, 8, 10, 12-15, 17-19, 22, 24-26). We present a case of RM that was determined by measuring the tumor volume during preoperative follow-up. The tumor had an unusual multinodular shape and contained heterogeneously disseminated rhabdoid cells (RCs), which seemed to be associated with the unusual progression of the tumor. To the authors' knowledge, this is the first report in which the clinical course of RM was monitored by measuring changes in tumor volume.

Case report

A 17-year-old boy suffered from generalized convulsions for over 4 years. The first attack occurred when he was 14 years old, the second when he was 16. Following consultation with a doctor, he was treated with an anticonvulsant. At that time, no brain abnormalities were detected by magnetic resonance (MR) imaging. When the patient was referred to our hospital, he had no neurological deficits. MR imaging revealed a well-enhanced mass in the left occipital lobe and a small mass deep in the left temporal lobe (Fig. 1). No other abnormities were seen by chest X-ray or whole body gallium scintigraphy. The tentative diagnosis was meningioma ; differential diagnoses were sarcoma or malignant lymphoma. A conservative observation schedule was chosen and MR imaging was performed every 1 to 5 months. Tumor volume was measured by Zed View software (LEXI inc. Tokyo, Japan) (2) ; 4.15 cm^3 on day 0, 4.44 cm^3 on day 35 (Fig. 2A), 4.69 cm^3 on day 104, 4.81 cm^3 on day 171 (Fig. 2B), 5.28 cm^3 on day 321, and 7.72 cm^3 on day 385 (Fig. 2C). Tumor volume doubling time (Td) was calculated according to the formula : $Td = \log 2/(\log V_1 - \log V_0) \times t$, where V_1 is the tumor volume measured on the last follow-up, V_0 is on the previous follow-up, and t is the time interval expressed in years (20). The Td calculated between

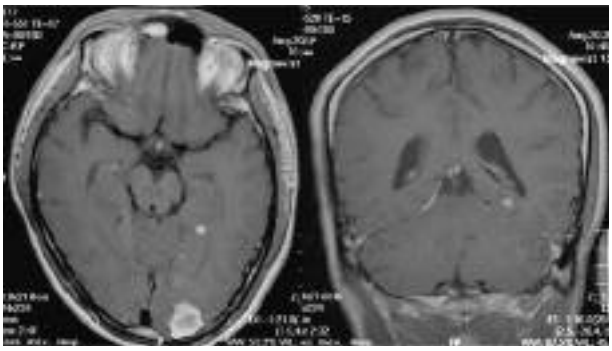

FIG. 1. — Axial and coronal T1-weighted magnetic resonance (MR) imaging with contrast medium showed two well-enhanced masses in the left occipital and deep left temporal lobes.

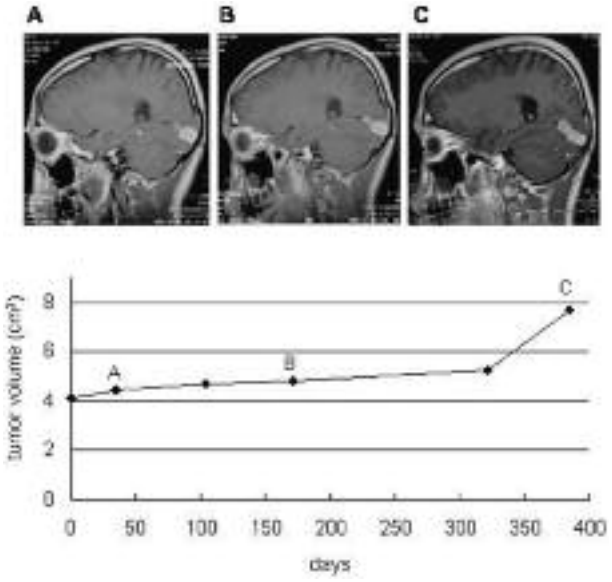

FIG. 2. — Sagittal preoperative MR images of the tumor in the left occipital lobe, with contrast medium, on days 35 (A), 171 (B) and 385 (C) after the initial consultation. Tumor volume in the left occipital lobe measured by Zed view software : 4.4 cm^3 (A), 4.8 cm^3 (B) and 7.7 cm^3 (C).

the day 0 and 35, the day 35 and 104, the day 104 and 171, the day 171 and 321, and the day 321 and 385 was 1.0, 2.4, 4.9, 3.0 and 0.3 years, respectively. The tumor in the temporal lobe did not increase in volume during the same time period. Given the increase in the size of the tumor in the left occipital lobe, the decision was made to perform surgery. No abnormal vascularization from the internal or external carotid arteries was detected by preoperative angiography.

The bone flap above the tumor was noticeably thickened. The dura mater adhered to the tumor, but was easily removed. The tumor was difficult to excise because it was very soft and its margin was unclear. The residual tumor, identified by 5-aminolevulinic acid photodynamic examination, was completely removed. The dural attachment was removed and repaired by a pericrainal flap (Simpson Grade I).

Pathological examination revealed that the tumor contained rhabdoid cells (RCs) with abundant eosinophilic cytoplasm and smaller spindle-shaped cells. The histological appearance varied depending on the ratio between these two types of cells. In the main components, both the RCs and spindle cells were arranged in a fascicular pattern (Fig. 3A), where geographic necrosis was often observed. In some regions, the tumor cells were exclusively RCs. The RCs were loosely cohesive and proliferated in a sheet-like pattern (Fig. 3B). Cellularity was high and necrosis was prominent (Fig. 3D). The components with many RCs invaded the brain parenchyma (Fig. 3E). Part of the tumor was composed mainly of spindle-shaped cells arranged in a fascicular pattern with a proliferation of collagen fibers (Fig. 3C). In this area, there were few RCs, and no necrosis was found. Although this component resembled fibrous meningiomas, nuclear atypia of the tumor cells was evident. Immunohistochemically, most of the tumor cells in the main tumor were positive for vimentin (Fig. 3F). A small number of tumor cells were immunoreactive for epithelial membrane antigen and cytokeratin. The MIB-1/Ki-67 staining index increased to 6-10% in the areas where RCs were abundant, but was 1-2% in the areas mainly composed of spindle-shaped cells. Ultrastructurally, RCs contained abundant intracellular organelles, including mitochondria, lysosomes and rough endoplasmic reticulum (ER) (Fig. 3G). Various amounts of intermediate filaments were arranged among the organelles or surrounded the nucleus (Fig. 3H).

The patient's postoperative course was uneventful and local radiation therapy was performed including the small lesion in temporal lobe (total 56Gy). MR imaging showed thickened dura mater and a small recurrent mass in the right occipital lobe two months after the surgical operation. In contrast, no change in the small deep temporal lesion was observed.

Discussion

The authors evaluated the volume of a RM tumor by image analysis over a 13-months period, from the initial consultation in our hospital until the surgical operation. The volume of the main tumor showed significant growth in the last two months, became multinodular and widened the cerebral sulcus, extending towards the brain parenchyma. The tumor was surgically removed and a pathological diagnosis of RM was made.

A considerable amount of literature on growth analysis of meningiomas has been published (7, 12, 20, 21, 23, 27), and yet this is the first report monitoring the volumetric change of RM. The growth rate of meningiomas may vary considerably. It was

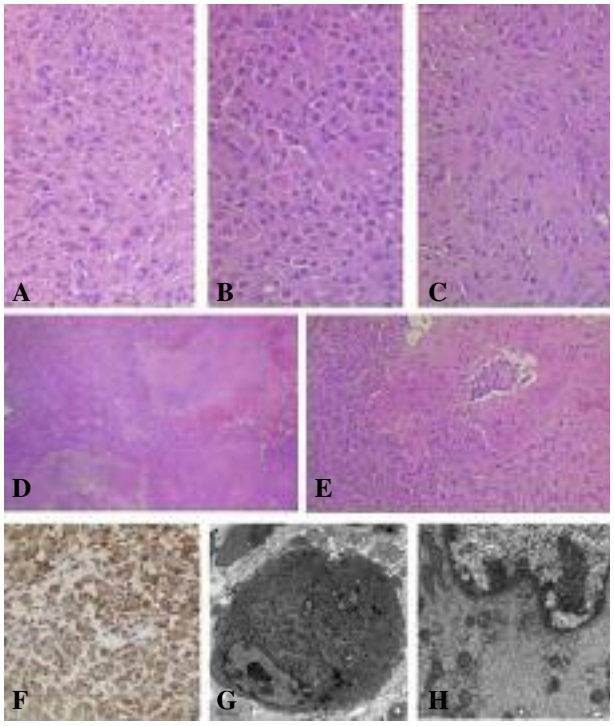

FIG. 3. — Photomicrographs of hematoxylin and eosin (HE) stained sections cut from different regions of the main tumor. Both the rhabdoid cells (RCs) and the spindle cells were arranged in a fascicular pattern (A). The RCs were loosely cohesive and proliferated in a sheet-like pattern (B). Part of the tumor contained mainly spindle- shaped cells arranged in a fascicular pattern with a proliferation of collagen fibers (C). Geographic necrosis was prominent in the area with the highest cellularity (D), and the tumor invaded the brain parenchyma (E). Most of the tumor cells contained vimentin immunoreactivity, which was diffuse at the periphery of the cytoplasm or around the nucleus (F). Ultrastructurally, RCs contained abundant intracellurar organelles, including mitochondria, lysosomes and rough ER (G). Varying amounts of intermediate filaments were arranged among the organelles or surrounded the nucleus (H). HE stain, A : × 200, B : × 200, C : × 200, D : × 200, E : × 200. vimentin : F : × 200. G : Bar = 2 µm, H : Bar = 0.5 µm.

previously reported that the Td of the asymptomatic meningiomas ranged from 1.27 to 143.5 years (mean, 21.6 years) (20). Jääskeläinen *et al.* reported that faster growth rate of the meningiomas related to high histological grading, and proposed the following limits to classify growth rate according to the Td ; very fast, Td < 50 days (0.14 years) ; fast, Td 50-200 days (0.14-0.55 years) ; moderate, Td 200-500 days (0.55-1.37 years) ; slow, Td > 500 days (1.37 years) (9). The Tds of the present case ranged from 1.0 to 4.9 years before the day 321, corresponding to the moderate to slow Td. The Td calculated between the day 321 and 385 was 0.3 years, suggesting that the tumor began to grow faster than before in the last two months.

In the surgical specimens, the tumor was seen to be composed of RCs and spindle cells. The ratio of RCs to spindle cells varied throughout the tumor. Malignant histological features, necrosis and high proliferation indices, were noted in the area where RCs were predominant. The heterogeneous distribution of the RCs might explain the change in growth rate of the tumor. The tumor grew slowly in the initial phase, possibly because components with low proliferation rates, predominantly the spindle shaped cells, occupied most of the tumor. The tumor began to grow rapidly when the malignant component containing abundant RCs became predominant. Our investigation showed that RMs can change their clinical course and become more aggressive over time, depending on their pathological features. In the present case, the decision was made to perform surgery based on the rapid increase of the tumor volume. At this point, the volumetric analysis of the tumor was useful to evaluate for the time of surgical intervention.

As previously described, RMs often contain conventional components similar to meningothelial or fibrous meningiomas, and rhabdoid transformation of meningiomas can occur during tumor progression. Perry *et al.* reported 15 cases of RM ; 9 of the cases (60%) had RCs when surgery was initial performed, but 6 (40%) had RCs only after recurrence (25). Since revision of the WHO classifications in 2000, 18 cases of RM have been reported in the literature, including our case (1, 3-6, 8, 10, 13-15, 17-19, 22, 24, 26). Fourteen cases (77.8%) were diagnosed as RMs when surgery was initially performed, including 3 cases (16.7%) in which the RCs were detected retrospectively (8, 18, 22). These results suggest that careful examination of the pathological specimens will increase the diagnosis of RCs at the initial surgery, although RMs will sometimes contain only a small number of RCs.

As observed in our case, rhabdoid transformation of tumor cells in meningiomas may be associated with aggressive biological and clinical behavior (12). On the other hand, RMs lacking malignant features have been reported (5, 18, 19, 25). These tumors contained various amount of conventional meningioma components, but the proportion of RCs was not always low. Most importantly, necrosis was not observed in these cases, and the proliferative indices were low, even when the RCs predominated. Thus, further analysis will be needed to conclude whether the appearance of RCs is always associated with malignant behavior in meningiomas.

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