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Apparent diffusion coefficient and arterial spin labeling perfusion of conventional chondrosarcoma in the parafalcine region: a case report

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ABSTRACT

Intracranial chondrosarcoma is a very rare malignant tumor of the central nervous system, and is difficult to preoperatively distinguish from other tumors using conventional imaging techniques. Here, we report the case of a 24-year-old woman who presented with mild headache due to chondrosarcoma in the frontal lobe. Preoperative conventional images showed findings typical of an oligodendroglial tumor. However, high apparent diffusion coefficient (ADC) value and extreme hypoperfusion on arterial spin labeling (ASL) were inconsistent with oligodendroglial tumor characteristics. The tumor was completely removed using a standard surgical procedure. Histologic diagnosis was a conventional (classic) chondrosarcoma. High ADC and hypoperfusion on ASL represented low cellularity and low vascularity within conventional chondrosarcoma, respectively. We discuss the utility of ADC and ASL for the preoperative diagnosis of conventional chondrosarcoma.

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Introduction

Intracranial chondrosarcoma is thought to be derived from persistent embryonic cartilage tissues at cartilaginous joints [1]. Intracranial cartilage tumors can be classified as histologically chordoma, chondroma, and chondrosarcoma. There are 2 types of chondrosarcoma variants: conventional (classic) type and mesenchymal type, according to the World Health Organization classification reedited in 2016 [2]. The incidence of all intracranial chondrosarcomas is reportedly less than 0.15% of all intracranial neoplasms [3,4]. Neurosurgeons may see very few cases of this disease during their years in practice, making it difficult to differentially diagnose it from other brain tumors. Most of intracranial chondrosarcomas have been reported to arise from the extra-axial structures such as skull base, the convex dura, falx, and intraventricle [3,5–8]. Thus, its localization often leads to misdiagnosis of meningioma [9,10]. Therefore, selection and interpretation of preoperative neuroimaging are crucial to precisely diagnose intracranial chondrosarcoma. We encountered a case of conventional chondrosarcoma in the left frontal lobe. In addition to conventional magnetic resonance imaging (MRI)...
imaging (MRI), we performed optional techniques including measurement of apparent diffusion coefficient (ADC) values and arterial spin labeling (ASL) perfusion imaging to preoperatively distinguish chondrosarcoma from other brain tumors. We discuss the utility of using ADC and ASL to preoperatively diagnose intracranial chondrosarcoma.

Case report

Preoperative images

The patient in our case study was a 24-year-old woman who presented with a slight headache for 1 month before admission to our hospital. She had no remarkable medical or family history. Conventional computed tomography (CT) showed multiple calcified lesions within a tumor located in the left frontal lobe (Fig. 1A), and conventional MRI revealed a mass lesion of 4.7 cm in diameter involving the cortex of this lobe. The mass lesion was depicted as hypointense on T1-weighted imaging, hyperintense on T2-weighted imaging, and heterogeneous enhancement on gadolinium-enhanced T1-weighted imaging (Fig. 1B–D). A part of the internal edge of the tumor seemed to perforate the falx. However, both the dura mater and falx adjacent to the tumor only showed slight enhancement, suggesting reactive changes. Based on the age of the patient and the CT and MRI findings such as calcified foci, heterogeneous enhancement, and lack of dural tail, a preoperative diagnosis was an oligodendrogial tumor rather than a meningioma. Then we assessed water molecule diffusion and blood flow within the tumor by ADC and ASL, respectively, using a 3T MR imager (Discovery 750, GE Healthcare, Milwaukee, WI). Diffusion-weighted MR scans to obtain ADC values were performed with the following sequences: repetition time (TR), 8000 ms; echo time (TE), 60 ms; matrix 128 × 80; field of view, 220 × 220 mm; 4 mm thickness with 1.5 mm gap; 3 motion-probing gradient directions; and b value, 1000 s/mm². We subsequently performed ASL using the same MRI machine as that used to determine ADC values with the following sequences: 3-dimensional fast spin echo, pseudo-continuous ASL (pCASL); TR/TE, 4347/10.5 ms; field of view, 240 × 240 mm²; postlabeling delay, 1525 ms. The ADC value was 2.4 × 10⁻³ mm²/s in the lesion, which was 3-fold higher than the value of 0.8 × 10⁻³ mm²/s measured in the normal region (Fig. 2, left). ASL showed extreme hypoperfusion with 0.2-fold more blood flow than that in the normal brain tissue (Fig. 2, right). These findings were highly inconsistent with the characteristics of oligodendrogial tumors and meningioma.

Surgery, histologic diagnosis, and outcome

A left frontal craniotomy was performed under general anesthesia, and we easily dissected the tumor bulk from the convex dura and falx. We were able to easily remove the tumor edge from the perforating hole at the falx. While dissecting from the

![Fig. 1 – Demonstration of conventional imaging. (A) Plain CT; (B) T1WI; (C) T2WI; (D) Gd-T1WI. CT, computed tomography; T1WI, T1-weighted imaging; T2WI, T2-weighted imaging; Gd-T1WI, gadolinium-enhanced T1-weighted imaging.](image-url)
brain surface, we found firm adhesion between the bottom of the tumor and the underlying cortex. Finally, an elastic hard tumor of 4.7 cm in diameter was completely removed.

The tumor was myxoid matrix-rich with mildly differentiated tumor cells and poor vascular structures on hematoxylin and eosin staining (Fig. 3A). Binucleated tumor cells were also seen (Fig. 3B), but the majority of tumor cells had a low nuclear-to-cytoplasmic (N/C) ratio and overall low cell density. The percentage of MIB-1-positive cells was greater than 10% (Fig. 3C) and the tumor size was relatively large with a diameter of 4.7 cm. Together, these findings led us to diagnose the tumor as a conventional chondrosarcoma. Immunohistochemical findings, positive for S-100 and negative for keratin and epithelial membrane antigen, supported the diagnosis of conventional chondrosarcoma [2,11]. No tumor cells were found on hematoxylin and eosin-stained preparations obtained from the rim of the hole on the falx. Irradiation and chemotherapy were not performed postoperatively, because the tumor was completely removed. The patient had no evidence of recurrence and metastasis for more than a year after surgery.

**Discussion**

Based on conventional CT and MRI, we initially diagnosed the tumor as an oligodendroglial tumor. However, the results of ADC and ASL were inconsistent with oligodendroglial tumor characteristics. The ADC values of oligodendroglial tumors are usually low. A previous report showed that the mean ADC value of an oligodendroglial tumor was less than $1.4 \times 10^{-3}$ mm$^2$/s [12]. Besides, meningioma might be raised as a tumor which should be preoperatively distinguished from chondrosarcoma [10]. Mean ADC values in meningiomas was reported to range from $0.80 \times 10^{-3}$ to $0.96 \times 10^{-3}$ mm$^2$/s [13]. In contrast, previous reports showed that the ADC value of intracranial chondrosarcoma was more than $2.0 \times 10^{-3}$ mm$^2$/s on the whole [14,15]. There have been a few reports regarding ADC value in intracranial chondrosarcoma. Two reports showed the mean ADC value of chondrosarcomas of the skull base: $2.05 \pm 0.26 \times 10^{-3}$ mm$^2$/s [14] and $2.02 \pm 0.14 \times 10^{-3}$ mm$^2$/s [15], while they did not refer to histologic variants of chondrosarcoma. The ADC value of $2.4 \times 10^{-3}$ mm$^2$/s in our case was higher than the mean value plus a standard deviation in 2 previous reports. Since ADC can be used to quantitatively assess the degree of water diffusion within a region, values depend upon factors that define the diffusional motion of water molecules such as cell density, N/C ratio, and amount of extracellular matrix [14]. Indeed, correlations between malignancies, ADC values, and cell density in skull tumors including chondrosarcoma have been reported [16]. We considered that the high ADC value of our case might result from extremely low cell density, a low N/C ratio, and high levels of collagen within conventional chondrosarcoma.

![Fig. 2 – Demonstration of additional imaging. Left, ADC map; right, ASL. Circles show regions of interest within the tumor and apparent normal white matter of contralateral side. ADC, apparent diffusion coefficient; ASL, arterial spin labeling.](image)

![Fig. 3 – Microscopic features of the tumor. H&E-stained preparations of weak magnification (10×) (A) and strong magnification (40×) (B). (C) Immunohistochemical staining of MIB-1. Arrows indicate the binucleated cells. H&E, hematoxylin and eosin.](image)
However, high ADC might not be necessarily adapted to the mesenchymal type as the other variant in intracranial chondrosarcoma, of which cell component often resembles the Ewing sarcoma-like small cell malignancy [2]. Because the prognosis is quite different between conventional and mesenchymal types [17], it would be better to enable differentiating between 2 types using appropriate neuroimaging prior to surgical removal of tumor. In cartilage tumors of the extremity bones, ADC has been already considered as a predictor for differentiating malignancy grading of chondrosarcoma [18]. Further reports are aspired to clarify whether ADC differs between conventional and mesenchymal types of intracranial chondrosarcoma.

A finding of ASL perfusion imaging in this case showed extreme hypoperfusion (0.2-fold more than blood flow in the normal region) reflecting poor vascular structures within conventional chondrosarcoma. In contrast, chondrosarcomas in extremity bones generally show hyperperfusion on ASL [19] because of their specific histologic features of a fine capillary network [20]. Meningiomas also show hyperperfusion as much as gliomas [21]. Perfusion status of intracranial conventional chondrosarcoma has been also assessed using perfusion MRI [5] and single-photon emission computed tomography with N-isopropyl-[123I]-p-iodoamphetamine [22]. These reports showed that conventional chondrosarcomas demonstrated hyperperfusion compared with other brain tumors. Extreme hypoperfusion is likely to be the most important hallmark of the conventional chondrosarcoma [5]. A majority of conventional chondrosarcomas and even 50% of mesenchymal chondrosarcomas show avascular finding on angiography [5]. Regardless of hypoperfusion, conventional chondrosarcoma is enhanced mildly or moderately on gadolinium-enhanced T1-weighted imaging [5]. This finding suggests that conventional chondrosarcoma leads to more or less disruption of the blood-brain barrier. ASL takes advantage of arterial blood as an endogenous tracer, which can reveal a more accurate perfusion status without being influenced by the condition of the blood-brain barrier. Therefore, ASL could accurately demonstrate hypoperfusion within tumor tissue in this case. To the best of our knowledge, this is the first report that assessed perfusion of intracranial conventional chondrosarcoma using ASL.

In conclusion, we think that extreme hypoperfusion on ASL must play a key role for differentiating the intracranial chondrosarcoma from other brain tumors. In addition, high ADC value cannot only distinguish intracranial chondrosarcoma from others, but might also differentiate between the conventional type and the mesenchymal type of the intracranial chondrosarcoma.

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