Mesenchymal Chondrosarcoma at the Falx Cerebri

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Intracranial mesenchymal chondrosarcoma is one of the rare tumors of the central nervous system, which often presents as an aggressive-appearing extra-axial hypervascular tumor at the falx cerebri. The tumor presents mostly in young adults, while meningioma or metastatic intracranial tumor most commonly in aged people. However, its imaging findings are non-specific. In this article, we present a 13-year-old girl with this primary intracranial tumor and its pellucid magnetic resonance imaging findings. We also review the other subtypes of chondrosarcoma and discuss the differential diagnosis of an aggressive-appearing extra-axial hypervascular tumor at the falx cerebri.

Key words: Brain neoplasms, MR; Falx Cerebri; Sarcoma, chondrosarcoma

CASE REPORT

A 13-year-old girl with a history of intermittent headache preceding vomiting and diplopia since one month ago, and then unsteady gait developed gradually. She was referred to our ophthalmological clinic because of papilledema noted by ophthalmoscope. She had been robust until one month before and there was nothing unusual about her development milestones. Ophthalmological examination disclosed strabismus of the left eye, optic disc edema and swelling. The neurological examination revealed paralysis of the left third cranial nerve. The routine laboratory studies were unremarkable and her medical history was noncontributory.

MRI showed a $8 \times 7 \times 5$-cm lobulated extra-axial mass at the parafalcine frontal region. The signal intensity of this tumor was isointense to hypointense on T1-weighted MR image [500/30/2 (repetition time/echo time/excitations)], inhomogeneously hyperintense with peritumor vasogenic edema on T2-weighted image (4000/100/2), and strong enhancement on gadolinium-enhanced T1-weighted image (Fig. 1). There were scattered foci of signal void tumor vessels
within the tumor, indicating hypervascularity. The diagnosis of meningioma or hemangiopericytoma was made before surgery.

At surgery, a well-defined extra-axial mass abutting the falx cerebri with severe brain swelling as predicted by MRI was found. The blood supply of this tumor was rich. The margin between the tumor and normal brain parenchyma was clear. The tumor did not invade the superior and inferior sagittal sinuses. It was removed completely under the help of microscopy and Cavitation Ultrasonic Surgical Aspirator (CUSA). Microscopic examination revealed some islands of low-grade neoplastic chondrocytes and pale-staining hyaline cartilage surrounded by small round cells disposed in a hemangiopericytomatous vascular pattern (Fig. 2). However, the tumor cells did not contain so plentiful reticulin fibers as hemangiopericytoma in the reticulin stain. The immunostaining for S-
100 protein also revealed positive staining on cartilaginous islands. The final diagnosis of mesenchymal chondrosarcoma was made. Following surgery, the patient had no neurological deficits. The patient didn’t receive post-operative radiotherapy. Post-operative follow-up with a series of MR studies revealed no evidence of local recurrence within 2 years after surgery.

**DISCUSSION**

Intracranial chondrosarcomas are rare tumors of the central nervous system. They constitute less than 0.16% of the brain tumors [2]. Seventy-five percent of intracranial chondrosarcomas originate at the skull base. The others arise from the meninges along the falx cerebri, tentorium, and convexity [1-3]. They are classified as classic, mesenchymal and myxoid subtypes based on their cytoarchitecture. According to the review by Korten in 1998, age distribution of these tumors ranges from 3 months to 76 years old, with a mean age of 37 years, and there is also no sex predilection [2].

The first report of non-skeletal intracranial mesenchymal chondrosarcoma was presented in 1973 by Wu et al, in which a rare neoplasm that can mimic a meningioma radiographically was described [4]. It is the most malignant tumor of the three subtypes (classic, mesenchymal and myxoid subtypes), with a tendency of intradural or cerebral growth and occasionally distant metastasis [1,2,5]. The meninges, especially the falx cerebri, are the most common locations of this tumor in the central nervous system [5]. It presents mostly in young adults (i.e., 10-30 years) and there is a slightly female predominance [3]. Reviewing the English-language literature to date, only 26 cases of intracranial mesenchymal chondrosarcomas have been reported [3]. In contrast, the classic subtype of intracranial chondrosarcomas has predominance in the 6th to 7th decades and has a male-to-female ratio of 5:3 [2,6].

Microscopically, mesenchymal subtype has a distinctive appearance which shows a densely cellular stroma of undifferentiated small round cells punctuated by islands of neoplastic chondrocytes and hyaline cartilage. A hemangiopericytomatous vascular pattern is common. However, the classic subtype is composed of atypical chondrocytes with rare anaplastic figures in a well-developed cartilaginous matrix. The myxoid subtype has the characteristic of ribbons chondrocytes in a myxoid matrix and is in association with other disease entities such as Ollier disease and Maffucci syndrome [1,7]. All of these tumors are strongly reactive to S-100 protein and vimentin. They have negative response to epithelial markers like epithelial membrane antigen and cytokeratin that help to differentiate chondrosarcoma from meningioma and chordoma [7,8].

The mesenchymal chondrosarcomas, the most malignant form, are isointense to hypointense on T1-weighted images and hyperintense on T2-weighted images and have strong enhancement after gadolinium enhancement. They are usually hypervascular angiographically and often attach to the meninges in the frontoparietal region, especially in the falx cerebri [3]. According to the review by Bingaman et al., 19 patients had dura invasion in 26 cases with this tumor. However, they did not mention whether venous sinus invasion was present or not in these cases [3]. Classic chondrosarcomas are well-defined slow-growing extra-axial masses, which are usually isodense to hyperattenuated, with variable degrees of heterogeneous enhancement on CT images. Occasionally, they have pressure erosion on the adjacent bone but no hyperostosis or bone destruction. On MR images, they are usually hypointense on T1-weighted images and hyperintense on T2-weighted images. Despite of the absence of gross calcification, there may be intense low signal intensity on the T1-weighted images which reflects the presence of cartilage islands [1]. The myxoid chondrosarcomas, the most rare subtype, are typically avascular angiographically [9].

It might be difficult to differentiate intracranial mesenchymal chondrosarcoma from other extra-axial hypervascular tumors at the falx cerebri, such as meningioma, hemangiopericytoma, metastasis and vascular malformation. However, certain features can be used prospectively to differentiate them. On CT scans, intracranial mesenchymal chondrosarcomas are usually isodense to hyperattenuated, with variable degrees of heterogeneous enhancement [1]. On MR imaging of our case, the tumor is isointense on T1-weighted and hyperintense on T2-weighted images with foci of signal void tumor vessels. Post-gadolinium T1-weighted images show intense enhancement of this tumor. In contrast, meningiomas are usually isointense on T2-weighted image and have homogeneous intense enhancement after gadolinium injection on T1-weighted image. Meningioma and metastasis are usually present in aged people and lack cartilaginous islands. Unlike chondrosarcoma or meningioma, hyperostosis and intratumoral calcification are usually not present in hemangiopericytoma [10]. As the high-grade mesenchymal chondrosarcomas may have a tendency for local recurrence and distant metastasis, accurate diagnosis is important for
optimal management [5]. At present, treatment of intracranial chondrosarcoma includes surgical resection as extensive as possible. Some debates exist regarding various treatment adjuncts, including radiation therapy, stereotactic surgery, and brachytherapy [1-3]; however, a standard protocol has not been established because of the rarity of the tumor.

In conclusion, intracranial mesenchymal chondrosarcoma is an aggressive-appearing extra-axial hypervascular tumor, which mostly presents in young adults and often locates at the falx cerebri. It is isointense on T1-weighted and hyperintense on T2-weighted MR images with foci of signal void tumor vessels. These characteristics of this tumor help us to differentiate it from meningioma, which mostly presents in elderly and is isointense on T2-weighted MR images. We recommend this tumor should be considered in the differential diagnosis of an aggressive-appearing extra-axial hypervascular tumor at the falx cerebri, especially in young adults.

REFERENCES


大腦镰上的間質軟骨肉瘤：病例報告及歷史回顧

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間質軟骨肉瘤是中樞神經系統中罕見腫瘤之一，它通常以大腦镰上血液供應豐富的腫瘤表現。此腫瘤的影像表現雖無專一性，然而它通常發生在年輕人身上，不同於此，腦膜瘤及轉移病通常發生在年紀大的病人。在此，我們提出一個十三歲女孩罹患此罕見腫瘤的病例及清晰的磁振造影影像。我們也回顧其他種類的軟骨肉瘤並討論大腦镰上血液供應豐富腫瘤的鑑別診斷。

關鍵詞：腦腫瘤，磁振造影；大腦镰；間質軟骨肉瘤