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CASE REPORT

Intracranial Parenchymal Mesenchymal Chondrosarcoma: a Case Report

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INTRODUCTION

Primary intracranial mesenchymal chondrosarcoma is a rare entity, with most cases extra-axial. We present an unusual case of parenchymal mesenchymal chondrosarcoma where reaching a radiological diagnosis is a challenge.

CASE REPORT

In December 2015, a 19-year-old man with unremarkable past health presented with insidious onset of right-side facial twitching but no focal neurological deficit. He attended our department for routine magnetic resonance imaging (MRI) of the brain. He experienced a brief episode of generalised convulsion during the examination. The MRI was aborted, and he was immediately admitted for in-patient care.

Urgent plain computed tomography scan of the brain revealed a large intra-axial tumour with extensive calcification centring over the left insula, with a smaller eccentric soft tissue component (Figure 1a and b). On MRI, the mass consisted of a small eccentric component at its medial aspect, predominantly hypointense on T1-weighted images, and mildly hyperintense on T2-weighted images; the rest of the mass showed irregular areas of T1 and T2 hypodensities with blooming on susceptibility imaging, corresponding to the calcification on computed tomography scan. A fair amount of perilesional oedema with mild rightward mid-line shift was noted (Figure 1c to e). The non-calcified component demonstrated avid heterogenous enhancement, with an increased relative blood volume on perfusion study (Figure 1f to g). There was an elevated choline (Cho) peak at 3.2 ppm on spectroscopy, with elevated Cho:creatine and Cho:N-acetylaspartate ratios (Figure 1h). No restricted diffusion was observed (Figure 1i to j). For the calcified part, there was only very low signal and the perfusion and spectroscopy pattern were not interpretable. The initial suspicion was that of a high-grade glioma, such as an astrocytoma or oligodendroglioma, less likely a germ cell tumour.

The patient underwent surgical excision 2 days later. Intra-operatively, a largely calcified left insula tumour

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Ethics Approval: The patient was treated in accordance with the Declaration of Helsinki. Written informed consent was obtained for all treatment and procedures.

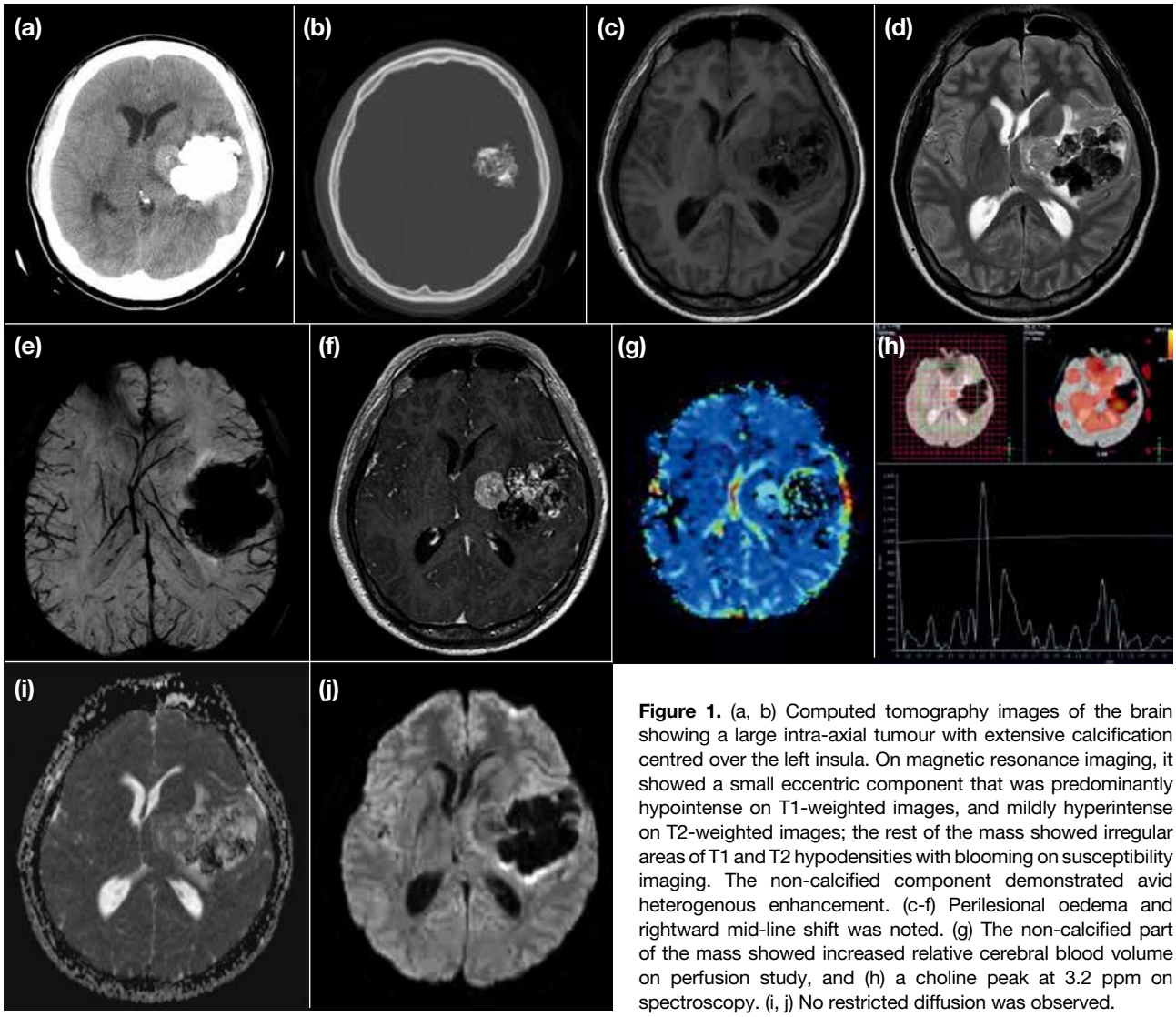

Figure 1. (a, b) Computed tomography images of the brain showing a large intra-axial tumour with extensive calcification centred over the left insula. On magnetic resonance imaging, it showed a small eccentric component that was predominantly hypointense on T1-weighted images, and mildly hyperintense on T2-weighted images; the rest of the mass showed irregular areas of T1 and T2 hypodensities with blooming on susceptibility imaging. The non-calcified component demonstrated avid heterogenous enhancement. (c-f) Perilesional oedema and rightward mid-line shift was noted. (g) The non-calcified part of the mass showed increased relative cerebral blood volume on perfusion study, and (h) a choline peak at 3.2 ppm on spectroscopy. (i, j) No restricted diffusion was observed.

with some soft tissue and fibrous component was identified. Histology showed a partially encapsulated tumour with high cellularity and components of hyaline cartilage and calcification. Mitotic figures were readily seen (Figure 2). Reverse transcription polymerase chain reaction confirmed the presence of fusion gene product of mesenchymal chondrosarcoma.

The recovery period was unremarkable. He underwent a course of radiotherapy and remained free of recurrence with no gross neurological deficits at 5-year follow-up examination.

DISCUSSION

Chondrosarcoma is a malignant bone tumour characterised by the production of chondroid matrix. There are four pathological subtypes: conventional chondrosarcoma, mesenchymal chondrosarcoma, clear cell chondrosarcoma and de-differentiated chondrosarcoma, with the latter two subtypes being exceedingly rare as intracranial tumour.[1]

Intracranial chondrosarcoma usually affect individuals 45 to 49 years of age, with no gender preference.[2] Nonetheless the mesenchymal subtype, as in our case, tends to affect younger patients in their 20s.[3]

The majority of intracranial mesenchymal chondrosarcoma, in contrast to the classic subtypes, are less frequently found at the skull base.[1,4,5] Instead, the

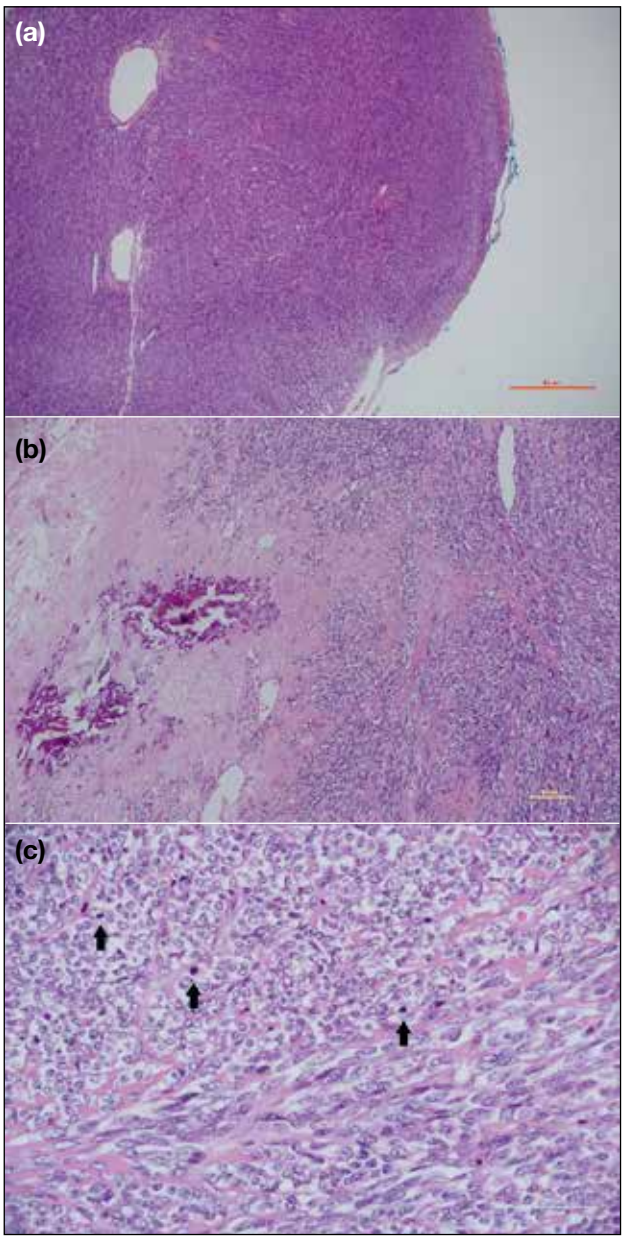

Figure 2. Histological results showing (a) a partially encapsulated highly cellular tumour (H&E stain, ×40) and (b) focal hyaline cartilage and calcification (H&E stain, ×100). (c) The tumour cells possessed elongated vesicular nuclei with indistinct cell borders. Mitotic figures are readily seen (arrows; H&E stain, ×400).

most common location is the craniospinal meninges.[6,7] In one previous study by Wang et al,[8] all included cases had a dural attachment. A sole intra-axial location is rare.

Radiographically the diagnosis can be challenging. On CT, it is often calcified and a characteristic ring and arc configuration may be observed.[9,10] When extra-axial, as in most cases, it can mimic a meningioma or haemangiopericytoma; as an intra-axial mass, the differential includes an oligodendroglioma, ganglioglioma and vascular malformation. On MRI, owing to the calcified matrix, it often displays an internal foci of low T1/2 signal with blooming on susceptibility imaging, while the soft tissue components show a heterogenous enhancement. There are currently limited data on MRI perfusion study and spectroscopy in intracranial mesenchymal chondrosarcoma. Some previous cases suggest a hypovascular pattern for the tumour.[9,11] Nonetheless this was not fully compatible in our case. The presence of Cho peak can be observed in many malignant bone and soft tissue tumours,[12] and is non-specific for the diagnosis. In a rare case of intracranial myxoid chondrosarcoma, an N-acetyl aspartate peak was noted, presumably due to the myxoid component.[13]

Mesenchymal chondrosarcoma is considered a more aggressive subtype, with an increased tendency for local and distant recurrences.[14,15] Unfortunately, due to its infrequent occurrence, there is no well-established treatment protocol, and the use of adjuvant chemo- and radio-therapy remains controversial.[1,6,10] However, a more aggressive and individualised multidisciplinary approach should always be considered in view of the worse prognosis.

CONCLUSION

As an exceedingly rare entity with confusing imaging findings, intracranial parenchymal mesenchymal chondrosarcoma is undoubtedly a challenging radiological diagnosis. It may mimic a high-grade glioma as illustrated in our case.

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