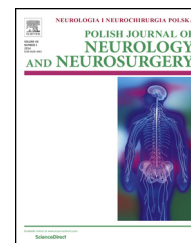


Available online at www.sciencedirect.com

ScienceDirect

journal homepage: <http://www.elsevier.com/locate/pjnns>

Case report

A rare tentorial mesenchymal chondrosarcoma in posterior cranial fossa: Case report

Anqi Xiao^a, Zhenlin Li^b, Xin He^c, Chao You^{a,*}^aDepartment of Neurosurgery, West China Hospital, Sichuan University, Chengdu, PR China^bDepartment of Radiology, West China Hospital, Sichuan University, Chengdu, PR China^cDepartment of Pathology, West China Hospital, Sichuan University, Chengdu, PR China

ARTICLE INFO

Article history:

Received 22 February 2014

Accepted 12 June 2014

Available online 27 June 2014

Keywords:

Posterior cranial fossa

Tentorium cerebella

Mesenchymal chondrosarcoma

Computerized Tomography

Magnetic Resonance Imaging

ABSTRACT

Intracranial extraskeletal mesenchymal chondrosarcoma is a very rare malignant tumor with predilection site of frontoparietal falx cerebri. Only few cases of mesenchymal chondrosarcoma in posterior cranial fossa are reported. Here, we report a 23-year-old young man with a dura-attached mass in left posterior cranial fossa misdiagnosed as a tentorial meningioma preoperatively. According to the following operation, the lesion was confirmed as mesenchymal chondrosarcoma surgically and pathologically. On MRI, the tumor was characterized by lobulated soft-tissue mass with dura-attached base, patchy calcifications and heterogeneous signal intensities. On contrast-enhanced MRI, it was well-defined, with marked enhancement. We consider that these imaging features above might remind us to consider the diagnosis of mesenchymal chondrosarcoma in posterior cranial fossa. The postoperative treatment of radiotherapy is still controversial. As for our case, according to the 24 months follow-up after postoperative γ -knife, our patient shows an optimistic prognosis so far.

© 2014 Polish Neurological Society. Published by Elsevier Urban & Partner Sp. z o.o. All rights reserved.

Introduction

Central nervous system mesenchymal chondrosarcoma is a very rare malignant tumor, which accounts for less than 0.16% of primary intracranial tumors [1]. Generally, the tumor is attached to dura, and occurs in supratentorial region, most common in frontoparietal falx cerebri [1]. The tumors found in

the posterior fossa are rarely reported. Due to its rare incidence and atypical site, the mesenchymal chondrosarcoma of posterior fossa could easily be misdiagnosed as other pathological neoplasm radiologically. Here, we report a 23-year-old young man with dura attached mass in posterior fossa which was misdiagnosed as meningioma preoperatively and confirmed as a tentorial mesenchymal chondrosarcoma through postoperative pathology.

* Corresponding author at: Department of Neurosurgery, West China Hospital, Sichuan University, 37 Guo Xue Xiang Street, Chengdu, Sichuan 610041, PR China. Tel.: +86 28 85422490; fax: +86 28 85164009.

E-mail address: shion_17928@hotmail.com (C. You).

<http://dx.doi.org/10.1016/j.pjnns.2014.06.001>

0028-3843/© 2014 Polish Neurological Society. Published by Elsevier Urban & Partner Sp. z o.o. All rights reserved.

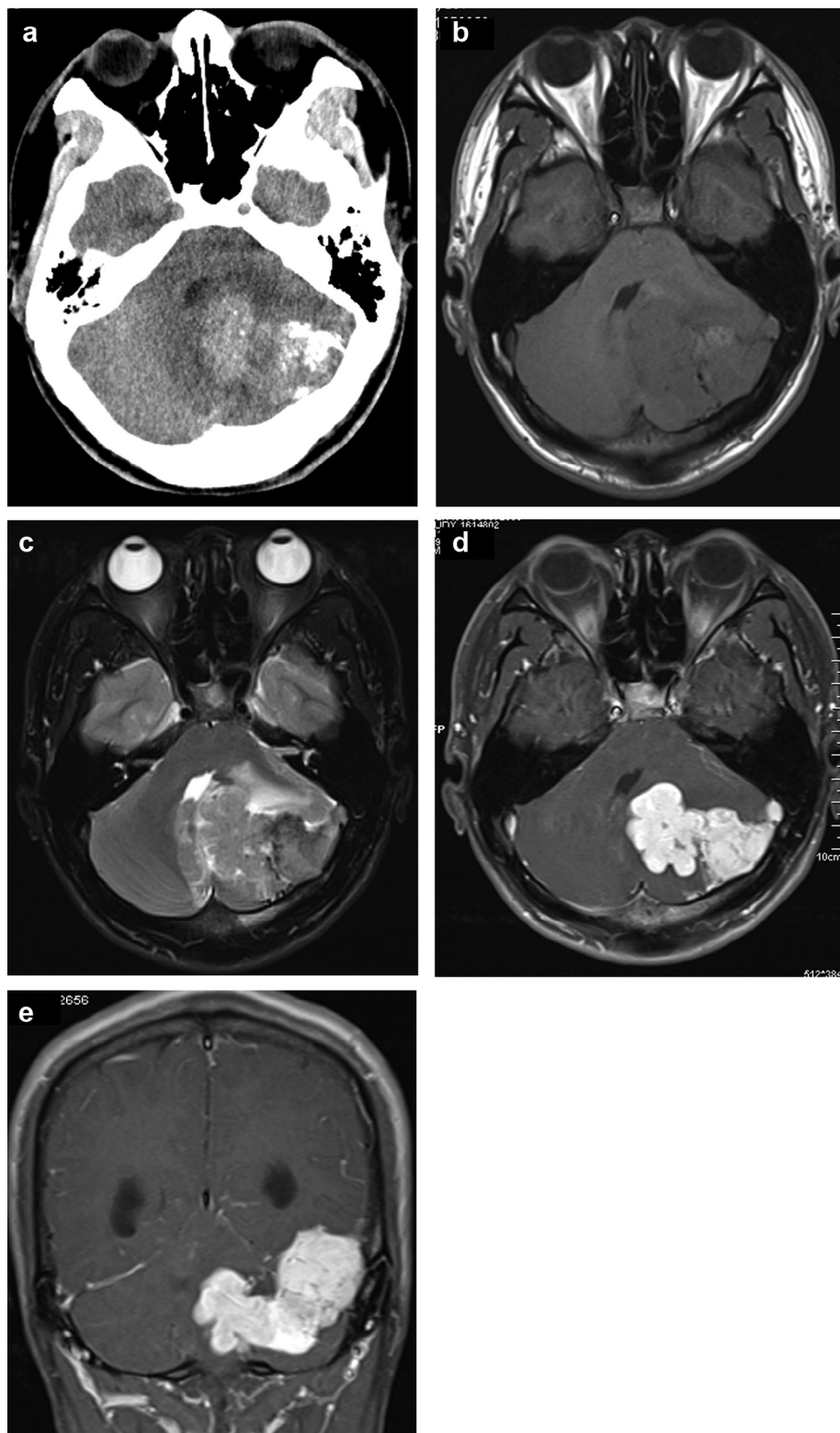


Fig. 1 – Preoperative CT and MR imagings for a 23-year-old young man with tentorial mesenchymal chondrosarcoma in posterior cranial fossa (A–E). CT plain scan showed a iso-/hyper-density mass with indistinct boundary located in the left cerebellar hemisphere. Within the mass, multi-patchy calcification shadows were displayed. (A) The MRI showed a giant lobulated mass in the left side of posterior fossa with heterogeneous iso-/hypointense on T1 weighted imaging (B) and non-uniform hyperintense on T2 weighted imaging (C). Consistent with CT, calcifications in tumor showed as hypointensity. On axial and coronal contrast-enhanced T1 weighted imaging (D and E), the lesion showed vivid enhancement with well-defined margin and wide range of attachment to tentorium, involved both supratentorial and subtentorial cranial cavities, and pressed left cerebellum and occipitotemporal parenchyma. The narrowed 4th ventricle with supratentorial hydrocephalus was clearly shown simultaneously.

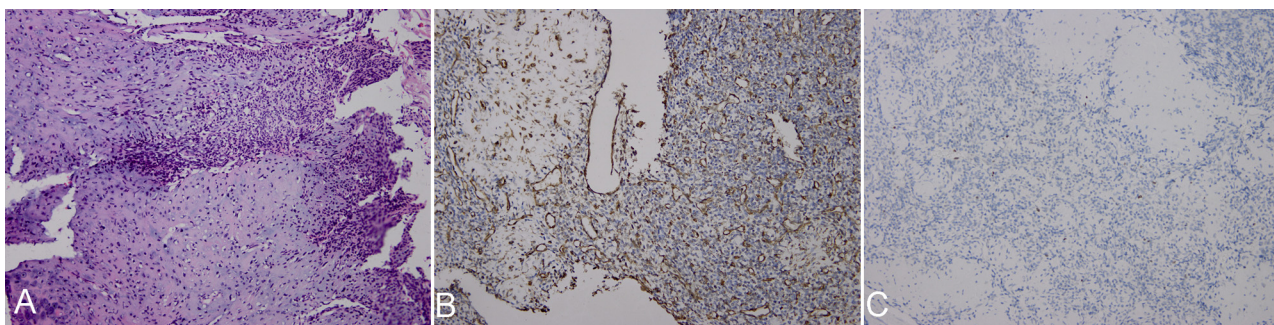


Fig. 2 – (A) Light microscopy of the tumor shows it consists of undifferentiated round or spindle-shaped cells and mature cartilaginous tissue (hematoxylin-eosin staining 200×). (B) The undifferentiated round cells were positive for vimentin staining. (C) Only scattered proliferating cells were positive for the proliferative marker Ki67 (labeling index lower than 2%).

Case report

A 23-year-old male, with a 5-month history of persistent headache and dizzy, was admitted to our hospital. The neurological examinations were normal and pathological signs were all negative. The CT revealed an iso-/hyper-density mass with indistinct boundary located in the left cerebellar hemisphere. Within the lesion, multi-patchy calcification shadows were shown (Fig. 1A). The preoperative MRI showed a giant lobulated mass, about 5.7 cm × 4.7 cm × 5.2 cm, widely attached to tentorium and rear dura, protruding into the left side of posterior fossa and supratentorial area, pressing ipsilateral cerebellum and occipitotemporal parenchyma. The lesion appeared as heterogeneous iso-/hypointense on T1 weighted imaging and non-uniform hyperintense on T2 weighted imaging. The patchy calcification shadows on CT were shown as hypointense on both sequences above (Fig. 1B and C). On contrast-enhanced T1 weighted imaging, the mass



Fig. 3 – Postoperative MRI at the 3rd day after surgery. On coronal contrast-enhanced T1 weighted imaging, the main part of tumor was resected with remission of the left cerebellar compression, though a small amount of residual tumor in left occipitotemporal lobe was remained.

had vivid enhancement with clear boundary (Fig. 1D and E). Due to its compression, the 4th ventricle was narrowed with supratentorial hydrocephalus, moreover, the left cerebellar tonsil had downward shift to about 1.5 cm below the foramen magnum plane. Based on the preoperative MRI, the tentorial meningioma was considered and the operation was arranged immediately. The tumor resection through posterior fossa paramedian approach was performed. In gross, the multi-lobular lesion was reddish-brown with hard texture, attached to the tentorium and adjacent dura. The main part of tumor extended into left cerebellum with partial extension to the occipitotemporal lobes. The mass and involved dura were grossly resected. The postoperative pathological finding revealed the tumor was composed of undifferentiated round or spindle-shaped cells and mature cartilaginous tissue. Immunohistochemical examination showed that the undifferentiated round cells were positive for vimentin and only scattered proliferating cells were positive for the proliferative marker Ki67 (labeling index lower than 2%) (Fig. 2). Thus, the pathological diagnosis of extraskeletal mesenchymal chondrosarcoma was confirmed. On the 3rd day after surgery, MRI revealed that the main part of tumor was resected, with the disappearance of cerebellar tonsillar herniation and the remission of the compressed the 4th ventricle, though a small amount of residual tumor was remained (Fig. 3). After operation, the patient's symptoms were alleviated and he was discharged on 8th-day after surgery. Six months later, this patient was given γ -knife treatment. We gave him a long-term follow-up, and up to now, 30 months after surgery, he is still alive and back to work without obvious symptoms.

Discussion

Intracranial mesenchymal chondrosarcoma is a rare malignant tumor which incidence is less than 0.16% of the primary intracranial tumors [1–3]. The intracranial extraskeletal mesenchymal chondrosarcoma was first reported by Dahlin and Henderson in 1962 [1]. After more central nervous system mesenchymal chondrosarcomas have been reported, its predilection age in second or third decade of life with female preponderance is noticed [3]. Typically, the tumor is supratentorial, more often located in frontoparietal region and attached to the dura mater. Few posterior cranial fossa

mesenchymal chondrosarcomas deriving from tentorium were reported.

Due to its invasive property, the tentorial mesenchymal chondrosarcoma could involve both supratentorial and sub-tentorial cranial cavities. When the mass extends into the posterior cranial fossa, it could press the cerebellum causing cerebellar tonsillar hernia. These abnormal signs are all found in our case. The intracranial extraskeletal mesenchymal chondrosarcoma has a growth characteristic of dura attachment, quite similar to the imaging findings of meningioma and hemangiopericytoma, therefore it could easily be misdiagnosed radiologically [1].

About the pathogenesis of the intracranial extraskeletal mesenchymal chondrosarcoma is still unclear. Some scholars consider it may derive from embryonic cartilaginous remnants in meninges, or from dural fibroblasts, or from the meningeal multipotent mesenchymal cells. Some researchers believe that mesenchymal chondrosarcoma is a sort of differentiating pre-mesenchymal neoplasm of chondroprogenitor cells, which could be found in whole body not limited in bony structures [2].

On pathologic examination, mesenchymal chondrosarcoma has dual morphological features under the microscope: besides of diffuse undifferentiated round or spindle-shaped mesenchymal cells, islands of differentiated chondrocytes, cartilaginous matrix, or local calcifications could be found among, without obvious transition zone between [4]. While on immunohistochemical examination, the undifferentiated mesenchymal cells are positive for vimentin staining, but negative for glial fibrillary acidic protein and neuron-specific enolase stainings, that makes identification in differentiation with meningioma or hemangiopericytoma.

On CT, intracranial mesenchymal chondrosarcoma is showed as heterogeneous soft tissue densities with patchy calcification shadows. On MRI, the tumor appears as well-defined lobulated soft tissue mass with heterogeneous hypointensity on T1WI, and non-uniform hyperintensity on T2WI and FLAIR. On contrast-enhanced MRI, the tumor displays significant enhancement indicating its abundant blood supply [1]. The local non-enhancement patchy or stripe shadows inside could be seen frequently, indicating its calcifications. These imaging features are all found in our case.

Intracranial extraskeletal mesenchymal chondrosarcoma is extra-axial tumor appearing vivid enhancement and dura attachment on contrast-enhanced MRI, quite similar to the imaging characteristics of meningioma. In spite of that, based on their different pathological features, there are still some differences between on MRI. Firstly, though both having a wide dural attachment on contrast-enhanced MRI, the mesenchymal chondrosarcoma does not appear an obvious dural tail sign as the meningioma does [3]. Secondly, relatively, the typical benign meningioma grows slowly and often has semi-circular or oval shape with wide base attached to dura, while the mesenchymal chondrosarcoma has rapid and uneven growth toward different directions forming lobulated shapes as malignant development, and commonly invades bilateral brain tissues across falx or tentorium. Thirdly, the features of calcification of these tumors are different. Generally, the concomitant calcifications in meningioma are spot-like, but in the mesenchymal chondrosarcoma are patchy-like and more frequently found, based on the abundant intratumoral chondrocytes and cartilage matrixes

where the calcium is apt to deposit. Hemangiopericytoma is also a meningeal-based tumor, with similar imaging appearance to mesenchymal chondrosarcoma. However, in mesenchymal chondrosarcoma, it is easier to find patchy-like calcifications, which are rarely seen in hemangiopericytoma [5]. PNET are most seen as extra-axial, dural-based masses with a tendency to affect a younger population, with the peak incidence in the second decade [6-8]. It is really challenging to distinguish extraskeletal mesenchymal chondrosarcoma from PNET based on the traditional MRI or CT. However, PNET has a characteristic taurine peak at 3.4 ppm on ^1H MRS (magnetic resonance spectroscopy), which is a unique feature in differentiation [9]. Combined the review of literatures with the imaging data of our case, these features above should be noticed and perhaps have significances in radiological identification.

The peritumoral edema of abaxial tumor might arise from obstacle of venous return by mass compression or tumor invasion surrounding brain tissue. Generally, the intracranial extraskeletal mesenchymal chondrosarcoma has more serious peritumoral edema compared with meningioma due to its invasiveness. However, the malignant meningioma may have similar imaging characteristics that are really hard to distinguish.

Due to its invasiveness, it is very difficult to totally resect the tumor only through the surgical therapy. The combination of surgery and radiotherapy is considered to reduce the risk of recurrence, however, the potential effect is still controversial [1]. Till now, the therapy data of γ -knife treating intracranial extraskeletal mesenchymal chondrosarcoma is scarce. Though only several cases reported, the therapeutic effect of γ -knife showed a better result during postoperative follow-up periods [10]. As for our patient, he experienced no obvious symptoms or neurological deficits, and no evidence of recurrence for 24 months after γ -knife and 30 months after craniotomy. We think that the efficacy of combination of surgery and radiotherapy is worthy of recognition.

Conclusion

Intracranial extraskeletal mesenchymal chondrosarcoma is a rare malignant tumor with poor prognosis. Typically it occurs in supratentorial region with dural attachment. The tentorial mesenchymal chondrosarcoma of posterior cranial fossa is very rare. Due to its rarity and similar imaging findings with meningioma, it often is misdiagnosed radiologically. We suggest that when a lobulated mass in young adults with patchy calcifications and dural attachment appears significant enhancement and serious peritumor edema on MRI, apart from meningioma, the diagnosis of mesenchymal chondrosarcoma should be considered. Currently, radical surgical excision and close follow-up monitoring are thought to be the optimal therapy [4]. The potential effect of postoperative radiotherapy and chemotherapy is under study and controversial. To our knowledge, the clinical cases of γ -knife treating postoperative extraskeletal mesenchymal chondrosarcoma are scarce, but showed a better result during their follow-up periods. Perhaps, γ -knife could provide a good therapeutic method in treating postoperative residual tumor of intracranial extraskeletal mesenchymal chondrosarcoma, and worth further clinical researches.

Conflict of interest

None declared.

Acknowledgment and financial support

None declared.

Ethics

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; Uniform Requirements for manuscripts submitted to Biomedical journals.

REFERENCES

- [1] Bingaman KD, Alleyne CJ, Olson JJ. Intracranial extraskeletal mesenchymal chondrosarcoma: case report. *Neurosurgery* 2000;46(1):207–11.
- [2] Harsh IV GR, Wilson CB. Central nervous system mesenchymal chondrosarcoma. Case report. *J Neurosurg* 1984;61:375–81.
- [3] Kan Z, Li H, Zhang J, You C. Intracranial mesenchymal chondrosarcoma: case report and literature review. *Br J Neurosurg* 2012;26(6):912–4.
- [4] Guccion JG, Font RL, Enzinger FM, Zimmerman LE. Extraskeletal mesenchymal chondrosarcoma. *Arch Pathol* 1973;95:336–40.
- [5] Chiechi MV, Smirniotopoulos JG, Mena H. Intracranial hemangiopericytomas: MR and CT features. *AJNR Am J Neuroradiol* 1996;17(7):1365–71.
- [6] Lin L, Varikatt W, Dexter M, Ng T. Diagnostic pitfall in the diagnosis of mesenchymal chondrosarcoma arising in the central nervous system. *Neuropathology* 2012;32(1):82–90.
- [7] Louis DN, Ohgaki H, Wiestler OD, Cavenee WK, editors. WHO classification of tumours of the central nervous system. 4th ed. Lyon: IARC Press; 2007. p. 176.
- [8] Theeler BJ, Keylock J, Yoest S, Forouhar M. Ewing's sarcoma family tumors mimicking primary central nervous system neoplasms. *J Neurol Sci* 2009;284:186–9.
- [9] Majós C, Alonso J, Aguilera C, Serrallonga M, Acebes JJ, Arús C, et al. Adult primitive neuroectodermal tumor: proton MR spectroscopic findings with possible application for differential diagnosis. *Radiology* 2002;225(2):556–66.
- [10] Förander P, Rähn T, Kihlström L, Ulfarsson E, Mathiesen T. Combination of microsurgery and Gamma knife surgery for the treatment of intracranial chondrosarcomas. *J Neurosurg* 2006;105(Suppl.):18–25.