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Case Report

Extraskeletal Intracranial Mesenchymal Chondrosarcoma: A Case Report

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Abstract

Mesenchymal chondrosarcoma (MCS) is a rare high-grade malignant tumor that affect young adults. The intracranial location is extremely exceptional. It is characterized by an undifferentiated mesenchymal cell, with islands of hyaline cartilage. This is a case of a 23-year-old young patient suffering from a generalized tonic-clonic seizures. The cerebral scanner objectified a hemorrhagic frontotemporal pass of 6x5x5 cm. The immunohistochemical and anatomopathological of the surgical specimen was in favor extraskeletal intracranial mesenchymal. The patient's progress was unfavorable due to tumor recurrence postoperatively. In this article, we discuss the clinical presentation, the diagnostic assessment, the therapeutic strategy as well as the prognosis of this rare pathology.

Keywords: Extraskeletal; Mesenchymal chondrosarcoma; Intracranial; Surgery; Prognosis.

Introduction

Firstly, described by Liechtenstein and Bernstein in 1959 [1], chondrosarcoma is a rare malignant cartilage tumor probably developed from the primary mesenchyme. MCS represents a small subgroup of intracranial sarcomas and constitutes less than 0.16% of primary intracranial tumors [2]. The origin of intracranial mesenchymal chondrosarcoma can be from the calvarium, the meninges, or the brain parenchyma. It can be classified as intra-cranial intra-axial, intra-cranial extra-axial, or both.

The definitive diagnosis is histological. Surgery is the only curative treatment guaranteeing the best cancer results. The

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prognosis is always poor due to the high rates of local recurrences and distant metastases, mainly pulmonary

In this article, a case of a 23-year- old women is described who had generalized tonic clonic seizures, the diagnosis of MCS was confirmed by histological examination of surgical specimen. This is a rare localization not reported before at National Institute of Oncology.

Case presentation

This is a 23-year-old young patient with no particular medical or surgical history, who consults in neurological emergencies for generalized tonic-clonic seizures, associated with headaches and vomiting.

At the initial examination, the patient's consciousness was clear. The patient did not present any neurological deficit. Ophthalmologic examination does not reveal papillary edema. The cranial nerves were intact. Results of the rest of physical examination were normal.

A cerebral CT scan was made to objectify a very vascularized left frontotemporal hemorrhagic process of 6x4x5 cm suggesting a high-grade glial tumor.

After stabilization and treatment of the intracranial hypertension syndrome, the patient was operated. A large left frontotemporal flap was performed. The opening of the dura revealed a frontal lobe infiltrated by a very hemorrhagic greyish process, pushing back the wall of the ventricle, which remains intact. Tumor resection was gradual and macroscopically complete.

The immunohistochemical and anatomopathological examination objective an undifferentiated process composed of round cells of small size, with low abundance basophilic cytoplasm, and atypical hyperchromatic nuclei, arranged in bundles within a hemangiopericyte-type stroma. This proliferation by lobulated place is dissociated by chondroid foci. CD-99 was positive in round cells, and S100 was positive on all tumor cells. On these histological results, the diagnosis of an extra-skeletal MCS was retained.

Postoperatively, and 7 days after the surgical procedure, the patient presented a deterioration of the neurological state with drowsiness, speech disturbance and psychomotor slowdown. A brain Magnetic resonance imaging (MRI) performed showed a residual left frontotemporal lesion process, intra-parenchymal enhanced in an intense and heterogeneous way after injection of contrast product, measuring 3.8x3.6 cm (Figure 1).

The patient was a candidate for radiotherapy but died before the start of treatment.

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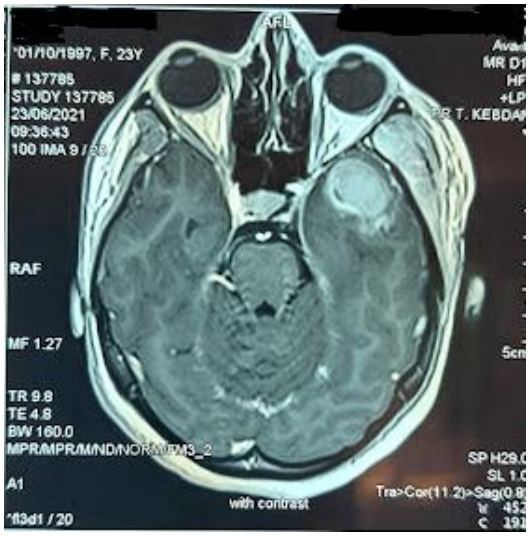

Figure 1: Brain MRI showed the residual left frontotemporal lesion process.

Discussion

MCS is a rare affection, presenting 20% of all bone tumors. Involvement of extra-skeletal tissue is exceptional, and cerebral localization is uncommon [3]. They are high-grade tumors of malignancy, with a tendency for local relapse and metastasis [4]. With a slight female preponderance, extra-skeletal intracranial MCS affects patients during their second or third decade of life [5,6].

In case of intra-cerebral localization, the involvement of the craniospinal meninges is the most common. Tumors are most frequently found in the frontoparietal region and are attached to the meninges [7]. In our case, the lesion was frontotemporal.

Symptoms depend on the location and size of the tumor. Headaches are common and indicate the aggressive nature of the tumor. They reflect either the increase in intracranial pressure, or the compression of intracranial structures. Focal neurological deficit, seizures, speech disturbance or sensory dysfunction may also be observed. Our patient presented with seizures and symptoms of intracranial hypertension.

MRI is the most relevant radiological means for the diagnosis of intra-cranial tumors. Extraskeletal intracranial MCS are typically hypo or normointense on T1-weighted, with an intense contrast enhancement. Extraskeletal intracranial MCS are highly vascularized tumors; therefore, a differential diagnosis with arteriovenous malformations and meningioma can be difficult radiologically.

The definitive diagnosis is essentially histological. Extraskeletal intracranial MCS have a biphasic architecture, with an undifferentiated contingent of small immature mesenchymal cells, and a well-differentiated contingent of mature cartilage cells [8,9]. Immunohistochemically, mesenchymal cells are positive for vimentin, and negative for glial fibrillary acidic protein and neuro-specific enolase.

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MSC metastases primarily affect the lungs, bones, and lymph nodes [10], and are exceptional in intracranial locations [11]. Cerebrospinal fluid seeding is rare [12]. Extraskeletal intracranial MCS are tumors with a very poor prognosis. It is the most malignant of all the subtypes of chondrosarcomas [2]. It is associated with a higher potential for relapse and local recurrence, even 20 years after treatment. The 5-year survival rate ranges from 42% [2] to 68% [13] and the 10-year survival rate is 28% [2] to 32% [13].

Radical surgery is the first-line treatment of choice [14,15]. A large carcinologic resection of the tumor, if possible, is associated with better results. However, these tumors are highly recurrent even after macroscopically complete resection. This is the case of our patient, who presented a local recurrence postoperatively after an almost complete resection. This rather rapid recurrence after surgery puts into question the completeness of surgical resection.

Radiotherapy is used to prevent local recurrence, but its role is still controversial. Radiotherapy should be discussed in case of unresectable tumors, inadequate surgical margins, or subtotal resection [4]. However, due to the poor prognosis of these tumors, and irrespective to the treatment of the primary site, adjuvant systemic therapy should be considered [4]. The chemotherapy protocol must take into account the histological subtype [16]. Nevertheless, the absolute benefits of radiotherapy and chemotherapy still unclear because of the rarity of the tumor.

Conclusion

Extraskeletal intracranial MCS is a rare and highly aggressive pathology, preferentially affecting young adults. The brain MRI highlight the tumor, most often of frontoparietal localization. Interpretation of radiological images must be careful, to rule out a meningioma or an ateriovenous malformation which may mimic extraskeletal intracranial MCS. The anatomopathology analysis allows confirmation diagnosis based on the biphasic nature of the tumor. Because of its power of local recurrence, surgery is the only therapeutic means that guarantees the best results in terms of recurrence-free survival. However, with aggressive surgery, the prognosis is always pejorative. Radiotherapy and chemotherapy can be discussed in case of tumor surgical margins or unresectable forms, even if their role is not well codified.

Acknowledgment

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Conflicts of interest

The authors declare that they have no conflicts of interest in relation to this article.

Consent

Written informed consent was obtained from our patient for publication of this case report and accompanying images.

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References

1. Lichtenstein L, Bernstein D. Unusual benign and malignant chondroid tumors of bone. A survey of some mesenchymal cartilage tumors and malignant chondroblastic tumors, including a few multicentric ones, as well as many atypical benign chondroblastomas and chondromyxoid fibromas. Cancer. 1959;12(6):1142-57. PubMed | CrossRef
2. Hassounah M, Al-Mefty O, Akhtar M, Jinkins JR, Fox JL. Primary cranial and intracranial chondrosarcoma. Acta neurochirurgica. 1985;78(3):123-32. PubMed | CrossRef
3. Tyagi S, Maheshwari V, Tyagi N. Mesenchymal chondrosarcoma--a case report. J Postgrad Med. 1992;38(1):34. PubMed
4. Harwood AR, Krajbich JI, Fornasier VL. Mesenchymal chondrosarcoma: a report of 17 cases. Clin Orthop Relat Res. 1981;58:144-8. PubMed | CrossRef
5. Salvador AH, Beabout JW, Dahlin DC. Mesenchymal chondrosarcoma—observations on 30 new cases. Cancer. 1971;28(3):605-15. PubMed | CrossRef
6. Kubota T, Hayashi M, Yamamoto S. Primary intracranial mesenchymal chondrosarcoma: case report with review of the literature. Neurosurgery. 1982;10(1):105-10. PubMed | CrossRef
7. Scheithauer BW, Rubinstein LJ. Meningeal mesenchymal chondrosarcoma. Report of 8 cases with review of the literature. Cancer. 1978;42(6):2744-52. PubMed | CrossRef
8. Enzinger FM. Cartilaginous soft tissue tumors. Soft Tissue Tumor. 1995.
9. Dorfman HD, Czerniak B. Bone tumors. St. Louis. Mosby. 1998:879.
10. Nakashima Y, Unni KK, Shives TC, Swee RG, Dahlin DC. Mesenchymal chondrosarcoma of bone and soft tissue. A review of 111 cases. Cancer. 1986;57(12):2444-53. PubMed | CrossRef
11. Bahr AL, Gayler BW. Cranial chondrosarcomas: Report of four cases and review of the literature. Radiology. 1977;124(1):151-6. PubMed | CrossRef
12. Zarabi MC, Parker JC. Intracerebral mesenchymal chondrosarcoma. Ann Clin Lab Sci. 1989;19(6):401-7. PubMed
13. Rollo JL, Green WR, Kahn LB. Primary meningeal mesenchymal chondrosarcoma. Arch Pathol Lab Med. 1979;103(5):239-43. PubMed
14. Guccuion J. Extraskeletal mesenchymal chondrosarcoma. Arch Pathol. 1973;95:336-40. PubMed
15. Harsh GR, Wilson CB. Central nervous system mesenchymal chondrosarcoma: Case report. J Neurosurg. 1984;61(2):375-81. PubMed | CrossRef
16. Huvos AG, Rosen G, Dabska M, Marcove RC. Mesenchymal chondrosarcoma a clinicopathologic analysis of 35 patients with emphasis on treatment. Cancer. 1983;51(7):1230-7. PubMed | CrossRef

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