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Radiological features of the intracranial extra-skeletal mesenchymal chondrosarcoma. A report of two cases insight in the literature review

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ABSTRACT

Background: Mesenchymal chondrosarcomas are the most malignant form of chondrosarcomas. They have mostly affected bones. Rarely, these tumors can be intracranial extraskeletal (IEMC) that originates from the meninges or parenchyma.

Methods and Materials: We presented two IEMC patients who were treated at our institutions and followed up for the long-term. To understand the radiological features of IEMC, we conducted a systematic literature review for previously reported series and cases of IEMCs.

Results: We surgically treated two young males with IEMC initially diagnosed at their age of 18 and 20 years. The patients initially treated with gross total resection (GTR) and GTR followed by radiotherapy, and followed-up for 218 and 73 months, respectively. With both patients, we obtained 83 reported IEMC patients from the literature. Among them, only 30 cases were reported with their radiological MRI details. The mean age of the reported cases was 24.5 ± 16.0 years (2 months–71 years). Female predominance was 54.2%. The mean progression-free and overall survivals were 27.9 and 39.0 months, respectively. Most IEMCs showed a partially calcified mass on roentgenography and a highly vascular mass on angiography. On T1WIs, IEMCs almost show hypo- to isointensity and intense heterogeneous enhancement after administering a contrast substance. On T2WIs, IEMCs show iso- to hyperintensity.

Conclusions: IEMCs usually show dural attachment without a net dural tail sign and a well-identified brimmed vascular nodule on TOF-MRA. This nodule appears as a prominent blooming on SWI. TOF-MRA and SWI images can help in the radiological diagnosis of IEMCs.

1. BACKGROUND

Intracranial extraskeletal mesenchymal chondrosarcomas (IEMCs) are rare malignant neoplasms. They almost occurred among pediatric and young people. Primary intracranial chondrosarcomas constitute about 0.15% of all brain tumors.¹ After including our two patients and an exhaustive literature review, we find only 83 reported patients with IEMCs.

Microscopically, IEMC characterizes by two distinct components with a frequently abrupt transition between these two components.

Keywords

intracranial extraskeletal
mesenchymal
chondrosarcoma,
TOF-MRA,
SWI,
vasogenic edema,
diagnosis



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The first component is comprised of highly cellular and undifferentiated mesenchymal cells. The second component is comprised of islands of well-differentiated and benign-appearing cartilage.^{1,2}

The recommended appropriate management is gross total resection (GTR) followed by radiotherapy; however, IEMCs have a high propensity for neural axis dissemination.¹⁻⁴ IEMCs are reported to be aggressive, quick-growing tumors with high mitotic index properties.^{1,2} Therefore, preoperative radiological distinguishing the subtype of chondrosarcoma is essential in managing these patients. In this study, we discuss the diagnostic radiological features that can help in distinguishing these rare malignant lesions by evaluation of the data from our two patients and followed with a brief review.

2. METHODS AND MATERIALS

We presented two IEMC patients who were treated at our institutions and followed up for the long-term. Both patients gave written consent to applied surgeries and for publications. To understand the radiological features of IEMC, we conducted a systematic literature review for previously reported series and cases of IEMCs.

To understand the radiological features of IEMCs, we identified the publications reported series or cases of IEMC. The selection criteria were; full-text published in English and accessed by MEDLINE, GOOGLE SCHOLAR, and PUBMED databases irrespective of the setting, study design, or details. Then, we analyzed the pooled sample after adding our two patients (n = 83). The search in the mentioned databases was up to Oct 1, 2020. We excluded the cases reported bone destructions, epidural location, tumors originate from skull bones, or reports did not identify non-skeletal situations.

Statistical analysis

Since the cases reported with sufficient data were less than 50 (cases with MRI and CT details 30 and 25 cases, respectively), most reported cases were case reports, and the early reported IEMC cases miss the important details regarding radiological examinations, we could not achieve meta-analysis.

3. RESULTS

Our Illustrative Cases

Case 1: An 18-year-old boy presented to our

outpatient clinic with a one-month history of headaches. A contrast-enhanced MRI revealed a well-capsulated heterogeneously enhanced tumoral mass measuring 4.9x4.5x4.2 cm. It was located in the right temporoparietal region. The intense contrast well-enhanced tumor was iso-hyperintense on T2WIs and iso-hypointense on T1WIs (Fig. 1). Presumptive diagnosis indicated that it is an atypical meningioma. At the time of surgery, we observed that the tumor had attached to the dura. GTR was applied to the mass. Histopathologically, the tumor was confirmed to be an MC. We discharged him on POD3 with no neurological deficits. No adjuvant treatment was applied. The postoperative metastatic work-up (PET-CT) demonstrated no extracranial involvement. Up to the POM22, his postoperative course was uneventful. At his yearly control visit, he had no symptoms. A contrast-enhanced MRI revealed a lobulated heterogeneously enhanced tumoral mass measuring 5.2x3.8x3.2 cm. It was located in the same first operation field. The tumor was resected completely. The tumor was confirmed histopathologically to be a recurrence of the first tumor. The patient received RT. On his POM72, he had been free of tumor recurrence or extracranial metastatic lesions. The patient had done his attended school. He works as an accountant for more than two years.

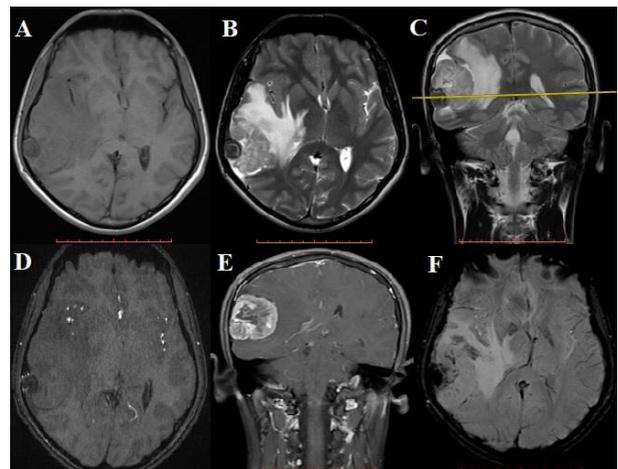


Figure 1. An 18-year-old boy presented to our outpatient clinic with a one-month history of headaches. His neurological examination was intact except for increases in DTRs. A contrast-enhanced MRI revealed a well-capsulated heterogeneously enhancing tumor measuring 4.9x4.5x4.2 cm. The mass was located in the right temporoparietal region and surrounded by diffuse edema. The lesion made a 7-8 mm shift to the left side and extending into the underlying brain

parenchyma. **[A]:** T1WIs demonstrated the lesion with two compartments. The lateral (small) one was hypointense and surrounded by the larger medial compartment. The medial compartment showed slight hypointensity compared to grey matter. **[B]:** T2WI-axial images demonstrated the lesion with separate two components. A small hypointense one was surrounded by the larger one that was hyperintense. **[C]:** T2WI-coronal images showed diffuse edema. **[D]:** TOF-MRA images showed a lateral hypointense nodule resembles the same as those that are seen in high vascularized lesions. The medial component was a slight inhomogeneous iso-hypointense. **[E]:** Contrast-enhanced T1WIs showed intense heterogeneously enhanced mass with two compartments. **[F]:** Brimmed prominent blooming nodule surrounded by the larger mild hyperintense component.

Case 2: A 38-year-old man referred to us with three days' duration of difficulties in speech, changes in consciousness, and severe headaches. A contrast-enhanced MRI revealed a multilobulated and well-capsulated mass. It was a heterogeneously enhanced tumor and measuring 8.2x6.2x5.6 cm. The tumor was located in the left frontotemporal region and surrounded by diffuse vasogenic edema. It was hyperintense on T2WIs and hypointense on T1WIs (Fig. 2). The tumor was diagnosed as intra-axial metastasis of MC. The patient received gross total resection followed by RT for right parietal IEMC 18 years ago. Over 18 years, he had received multiple mandibular and maxilla surgeries and twice adjuvant RT for several metastases. In his last presentation, he was lethargic and had not full responses to alters. NTR was achieved. After the surgical intervention, the patient required ICU care and died of disease on POD93.

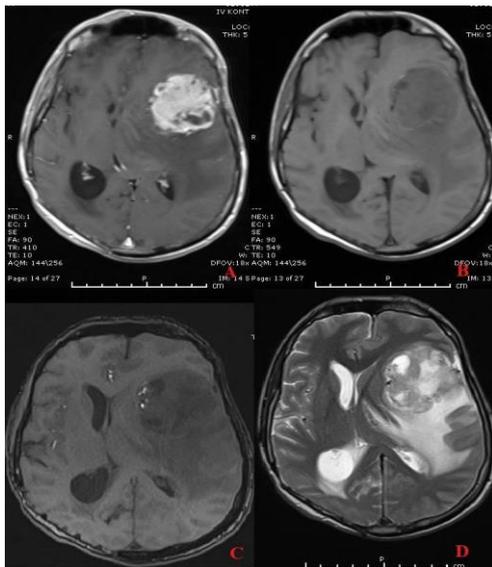


Figure 2. A 38-year-old man referred to us with three days' duration of difficulties in speech, changes in consciousness, and severe headaches. A contrast-enhanced MRI revealed a multilobulated heterogeneously intense enhanced tumor measuring 8.2x6.2x5.6 cm. The mass was located in the left frontotemporal region and surrounded by diffuse edema. The patient received gross total resection followed by RT for right parietal IEMC 18 years ago. Over 18 years, he had received multiple mandibular and maxilla surgeries and twice adjuvant RT for several metastases. **[A]:** Contrast-enhanced T1WIs revealed a multilobulated, heterogeneous, and intense enhanced tumor with distributed hypointense areas. **[B]:** T1WIs showed a mild hypointense lesion with a brimmed hypointense nodule. **[C]:** TOF-MRA images showed a hypointense lateral compartment. It resembles the nodule is seen in a high vascularized lesion. The medial component was a slight inhomogeneous iso-hypointense. **[D]:** T2WI showed heterogeneous hyperintense surrounded by diffuse vasogenic edema while the medial nodule was hyperintense.

Literature Review

We surgically treated two young males with IEMC initially diagnosed at their age of 18 and 20 years. The patients initially treated with gross total resection (GTR) and GTR followed by radiotherapy, and followed-up for 218 and 73 months, respectively. With both patients, we obtained 83 reported IEMC patients from the literature. Among them, only 30 cases were reported with their radiological MRI details. Thirty-three patients were reported without any radiological details. The mean age of the reported cases was 24.5 ± 16.0 years (2 months–71 years). Female predominance was 54.2%. The mean progression-free and overall survivals were 27.9 and 39.0 months, respectively. The most commonly affected intracranial region was frontal that was seen in 22 patients (26.5%). The mean preoperative maximal diameter of 31 reported tumors was 6.0 ± 2.3 cm (1.8–11.0 cm). The parenchymal origin was reported in 13 cases while falx and tentorial originate were reported in 15 and 8 cases, respectively.

The early reported cases were reported with details regarding roentgenography and angiography. Early reports mentioned that IEMCs demonstrate several intracerebral flecks of calcification, destructive lesions, mottled calcification, or dense flocculent calcification on roentgenography. Some IEMCs showed a highly vascular mass on angiography. Twenty-five cases were reported with CT details. Most IEMCs showed a lesion with two compartments; one compartment had density higher than calcium and the other had

density lower than calcium on CT scan. Sometimes they show bone destructions. On T1WIs, IEMCs almost show hypo- to isointensity and intense heterogeneous enhancement after administering a contrast substance. On T2WIs, IEMCs show iso- to hyperintensity.

4. DISCUSSION

Histopathologically, IEMC is a biomorphic tumor that comprises of undifferentiated cell mesenchymal mixed with differentiated cartilaginous tissue. Therefore, the tumor in most cases showed heterogeneous intensity in most sequences. Contrast-enhanced MRI remains the gold standard to demonstrate these lesions. However, the radiological findings are not pathognomonic for IEMCs. Calcifications that may appear incidentally on CT could be aware of the radiologist regarding IEMC. IEMCs almost are extra-axial, well-capsulated, and lobulated in shape. However, 13 cases were reported to be intra-axial (parenchymal in origin) lesions.⁵

TOF-MRA and SWI techniques help in the radiological diagnosis of IEMCs. The apparent dural tail is one of the most characteristic radiological features of meningioma. In our two cases, the dural tail sign wasn't net while we identified the brimmed vascular nodule without vessel dilatations on TOF-MRA. This nodule appears as a prominent blooming on SWI. This nodule is the same as those seen in vascular lesions. Although it is not pathognomonic, vasogenic peritumoral edema on FLAIR and T2WIs usually is prominent. The same features are seen in angiomatous meningiomas too. However, calcification can be distinguishable features for IEMCs. Additionally, we can recognize vascular lesions such as AVM and aneurysms by TOF-MRA.

Up-to-date, no study could differentiate IEMC from meningioma by radiological scanning studies. IEMCs are misdiagnosed as atypical meningioma,^{1,2} hemangiopericytoma,³ schwannomas,⁴ dural-based metastasis,⁵ gliomas, or oligodendroglioma.⁶ IEMCs vary from hypo- to isointense on T1-WIs with intense heterogeneous enhancement after administering a contrast substance. On T2WIs, IEMCs demonstrate iso- to hyperintensity. These lesions show an iso- to hyperintensity on MRA images that mimic arteriovenous malformations. Sometimes IEMCs demonstrate extremely hypervascular on angiographic images.^{3,4} Included in our two cases, the radiological features were mentioned in 30

patients. Six among them were reported to have cystic components, four were highly vascular, and three were hemorrhagic.

TOF-MRA can guide the surgeon in understanding the vascularity of the tumor, as TOF-MRA demonstrates the main vessels that passing through the tumors and is useful to identify tumor involvement with the cavernous sinus and main vascular structures. SWI technique is beneficial for detecting smaller vascular lesions that otherwise are missed by other sequences.

The craniospinal meninges are the most commonly seen location of IEMCs in CNS.^{1,3-6} We can divide IEMCs into dural or parenchymal. The most commonly affected region was the frontal with a dural attachment that was seen in 23 patients. Most of the reported lesions with a dural or meningeal attachment are usually supratentorial (71 patients); 49 were reported to be lateral lesions while 22 were midline lesions. Parenchymal origin was reported in 13 tumors. One of the most challenging in diagnosis the lesion is taken small biopsy or specimens from one component without pieces from the second one when obtained pieces without the cartilaginous elements.^{1,3-6} The lesion is generally demonstrated well-circumscribed, the rubbery firm solid, multilobulated, gray or reddish-brown colored, and almost invasive. Prominent vascularity and focal calcification are the main features of the lesion's cut surface.

The present study suffered from a few limitations, most of the early reported IEMC cases miss the important details needed to define the natural history and applied treatment, the recently reported cases miss sufficient data regarding the radiological examinations, and the pooled data were not sufficient to achieve meta-analysis review.

5. CONCLUSIONS

In our patients, we utilized TOF-MRA and SWI images to distinguish these malignant lesions from other benign lesions. Most reported IEMCs show hypo- to isointense and iso- to hyperintensity on T1-WIs and T2-WIs, respectively. They showed intense heterogeneous enhancement with contrast. On T2WIs, IEMCs show iso- to hyperintensity. Although we can't emphasize this due to their rarity, a brimmed vascular nodule on TOF-MRA and a prominent blooming (nodule) sign on SWI may distinguish these highly vascularized solid firm

lesions. They uncommonly have cystic, highly vascular, or hemorrhagic components.

DECLARATIONS

Consent for publication:

Informed written consent was obtained from both patients and their relatives for publication.

Competing interests:

None

Funding:

None

Authors' contributions:

AA: Conceptualization, Methodology, Software, Supervision, Formal analysis, Statistical analysis, Literature review, Visualization, Investigation, Writing – Original draft, Writing – Review, and Validation. **İÇ:** Validation, Writing, Reviewing, Literature Review, and Supervision.

Both authors deserve the first name.

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ABBREVIATIONS

CNS = Central nervous system, DTRs = Deep tendon reflexes, ER = Emergency department, GTR = Gross-total resection, IEMC

= Intracranial extraskeletal mesenchymal chondrosarcoma, MC = Mesenchymal chondrosarcoma, NTR = Near-total resection, POD = Postoperative day, POM = Postoperative month, RT = Radiotherapy, T1WIs = T1-weighted MR images, T2WIs = T2-weighted MR images, TOF-MRA = Time-of-Flight magnetic resonance angiography.

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