CASE REPORT

INTRACRANIAL CHONDROSARCOMA IN RIGHT FRONTAL REGION, MIMICKING MENINGIOMA: A CASE REPORT

Ashok Kumar,[1] Bushra Rehan,[1] Imtiaz Ali Panhwar,[1] Javeria Anees,[1] Naveen Faridi[2]

[1] Department of Radiology, Liaquat National Hospital, Karachi, Pakistan.
[2] Department of Histopathology, Liaquat National Hospital, Karachi, Pakistan.

PJR April - June 2020; 30(2): 141-144

ABSTRACT

Intracranial chondrosarcoma is a rare malignant cartilage-forming tumor. Small number of cases in the posterior cranial fossa has been reported previously. We describe a case of 17 year old male patient who presented with history of seizures from 2 years. The preoperative diagnosis was cystic meningioma as MRI of the patient revealed large multilobulated extra axial solid cum cystic mass in right frontal region with mild punctuate enhancement of solid component with enhancement of walls and septae. This was associated with widening of diploic space of frontal bone with inhomogeneous signal and mild post contrast enhancement. There was mild Dural thickening in this region with indistinct inner table. However, the tumor was histologically composed of cartilaginous tissue without meningiomatous differentiation and the final diagnosis was a well differentiated chondrosarcoma based on its histological appearance.

Keywords: intracranial tumor, chondrosarcoma, classical chondrosarcoma

Introduction

Primary intracranial chondrosarcoma is an extremely rare malignant tumor of the central nervous system, which accounts for <0.16% of all primary intracranial tumors.[1] The tumor most commonly arises from the skull base, however, cases originating from the choroid plexus, dura matter and brain parenchyma have also been reported.[1] Clinical presentation includes headaches, seizures, dizziness, tinnitus or decreased visual acuity.

The imaging findings are nonspecific, and can mimic other neoplasms, particularly metastases if the patient has a known primary neoplasm elsewhere and if the tumor is Dural based then the differential may include meningioma.[2]

Careful attention is required to raise the suspicion of an intracranial chondrosarcoma on the basis of radiological findings. Few radiological key features support the diagnosis. As intracranial chondrosarcomas are slow-growing in nature, there may be erosive changes on the adjacent bone, in comparison to hyperostosis caused by meningiomas or bone destruction seen with metastatic disease.[2]

Histo pathological diagnosis is the gold-standard. Here we present a case of a patient with well differentiated classic intracranial chondrosarcoma in the right front-parietal region, which was radiologically reported as meningioma and the final diagnosis was a well differentiated chondrosarcoma based on its histological appearance.

Case Report

We present a case of 17 year old male patient presented to accident and emergency department of Liaquat Nation hospital Karachi with complain of 14 to 15 episodes of fits. He underwent an MRI with contrast which revealed large multilobulated extra

Correspondence : Dr. Ashok Kumar
Department of Radiology,
Liaquat National Hospital,
Karachi, Pakistan.
Email: ashok.jeythwani@gmail.com

Submitted 6 May 2020, Accepted 18 May 2020

axial solid cum cystic mass in right frontal region causing significant mass effect on right frontal parenchyma, midline shift of approximately 2.4 cm with compression of left frontal lobe. It appeared to be hypo to iso intense on T1 and intermediate to high on T2 weighted images. On post contrast study, it showed mild punctuate enhancement of solid component with enhancement of walls and septae.

There was widening of diploic space of frontal bone with inhomogeneous signals and mild dural thickening in this region with indistinct inner table. It approximately measures 7.5 x 7.2 x 4.8cm (AP x TS x CC).

The mass was abutting and compressing anterior one third of superior sagittal sinus. Possibility of atypical cystic meningioma was raised.

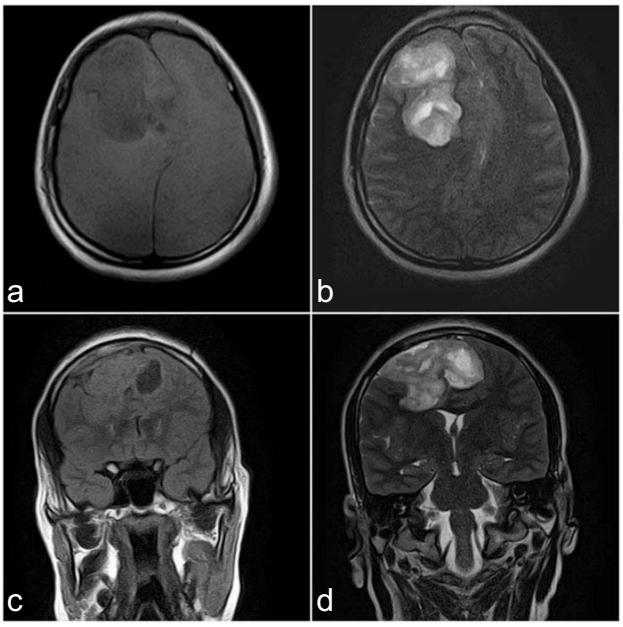

Figure 1: Shows multilobulated extra axial solid cum cystic mass in right frontal region. It is hypo to iso intense on T1 **(a)** and intermediate to high on T2 weighted images **(b,d)** There is widening of diploic space of frontal bone **(c)**.

Patient underwent a craniotomy and excision of mass. Grossly the mass appeared to be grayish white firm to hard tissue.

Histopathology of the excised mass revealed lobules of cartilage separated by fibrous bands. The chondrocytes were atypical showing mild to moderate pleomorphic enlarged, hyperchromatic nuclei. Binucleation was also seen. No mitosis was seen. On immunohistochemical studies performed by DAKO envision method CKAE1/AE3 was negative while S-100 was positive. So on the basis of histopathology the diagnosis of atypical chondroid neoplasm was made.

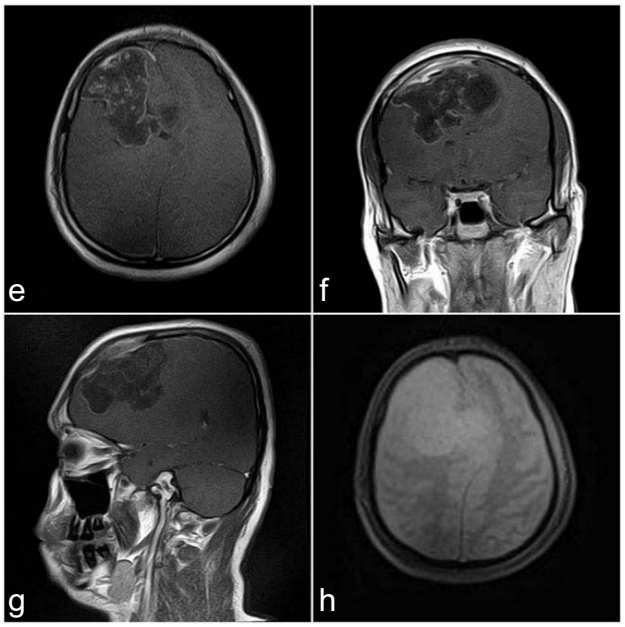

Figure 2e, f & g: Show post contrast images showing mild punctuate enhancement of solid component with enhancement of walls and septae. There is mild dural thickening with indistinct inner table **(f, g)**. No evidence of low signals on GRE images **(h)**.

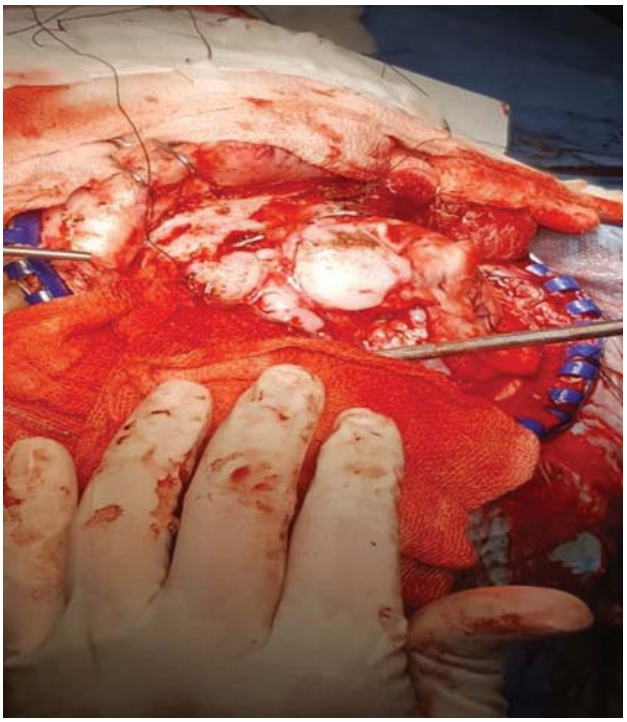

Figure 3: Shows greyish white firm to hard mass.

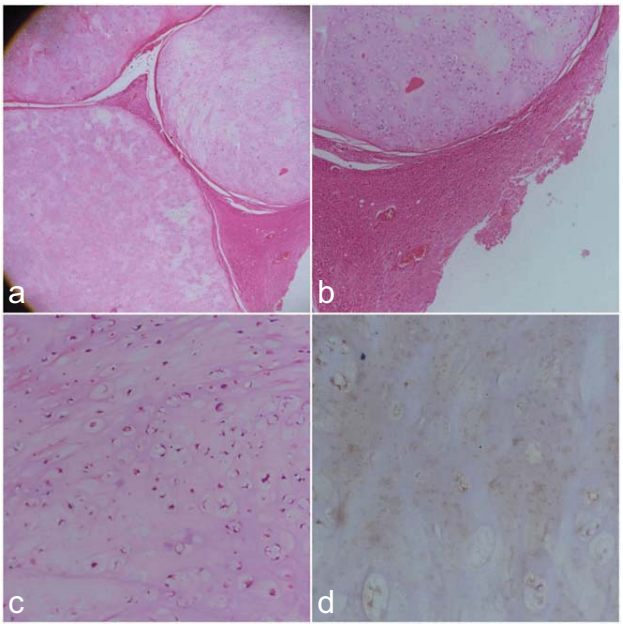

Figure 4: Shows multiple fragments of chondroid neoplastic lesion exhibiting lobular pattern which are composed of chondroid stroma with lacunae containing atypical chondrocytes showing mild to moderate pleomorphic enlarged, hyperchromatic nuclei. Some of the nuclei show bi and multinucleation. No mitosis seen. These chondrocytes are highlighted on immunohistochemical stain S100 while negative for CKAE1/AE3.

Discussion

Intracranial chondrosarcoma is a rare malignant cartilaginous tumor that was first reported by Mott in 1899.[3] They frequently arise at the skull base from the undifferentiated embryonic cells of the cartilaginous synchondroses.

Intracranial chondrosarcomas of dural origin are very rare, and only 53 cases have been reported. Majority of the skull-base chondrosarcomas are of classic variant (> 80%), Dural-based chondrosarcomas are predominantly of mesenchymal variant (60%). In a study of the 25 falcine and parasagittal chondrosarcomas, only 12 cases were of classic variant. The remaining cases were predominantly mesenchymal with only one myxoid variant.[4]

Histologically, intracranial chondrosarcomas are classified into three subtypes: Well-differentiated (classical type), intermediate (myxoid type) and undifferentiated (mesenchymal type).[1] In a review study of 192 chondrosarcoma cases by Chandler et al,[5] 62% were of the classical subtype, while the mesenchymal and myxoid types were 30 and 8%, respectively. In this study, a case of classical type intracranial chondrosarcoma that occurred in the right frontal region of the skull was presented which was diagnosed as cystic meningioma preoperatively. Mostly classical chondrosarcomas occur in the base of the cranium and affect patients between the fourth and sixth decades of life. In the review of chondrosarcomas by Korten et al,[6] 37% of tumors were located in the petrous bone, while 23% occurred in the occipital bone and clivus, 20% in the sphenoid bone and 14% in the frontal, ethmoidal and parietal bones; the remaining 6% were in dural tissue, which does not typically contain cartilage.

On immunohistochemical study, chondrosarcomas are usually positive for S100, while only scattered proliferating cells are positive for Ki-67. These features differentiate chondrosarcoma from meningioma, hemangiopericytoma, metastasis and vascular malformations.[1]

On imaging, these lesions are often mistaken for meningioma due to their extra-axial location and for metastasis if there is known primary malignancy. As intracranial chondrosarcomas are slow-growing in nature, there may be erosive changes on the adjacent bone, in comparison to hyperostosis caused by meningioma and bone destruction seen with metastatic disease.[2]

Computed tomographic (CT) scan usually reveals an isodense/hyperdense mass with heterogeneous enhancement and varying degrees of calcification in patients with chondrosarcomas. On MRI, they are mostly hyper intense on T2-weighted images and hypo intense on T1-weighted images. Variable shortening of T1 and T2 relaxation times due to scattered mature cartilage cells makes them heterogeneous. On post contrast study, classic variant shows a honeycomb pattern of enhancement whereas mesenchymal variant shows homogenous enhancement.[7]

Conclusion

Chondrosarcomas are rare intracranial cartilaginous tumors and are not limited to the skull base even in their classic variants. As chondrosarcomas have similar imaging findings with meningioma, a confident

radiology based diagnosis is often challenging. Careful analysis of CT and MRI characteristics support the diagnosis. Our case suggests that imaging findings can be confusing, and the diagnosis of a meningioma warrants consideration. Histopathologic diagnosis is always gold standard.

Conflict of Interest: None

References

1. Chi J, Zhang M, Kang J. Classical intracranial chondrosarcoma: A case report. Oncology letters. Nov 2016; **12(5):** 4051-3.

2. Bhatt AA, Campeau N, Black DF. Primary intracranial extraskeletal chondrosarcoma. Applied Radiology. Feb 2017; **46(2):** 32.

3. Mott FW. Chondrosarcoma springing from the sella turcica. Arch Neurol Psychiat. 1899; **1:** 432-3.

4. Naik H, Velho V, Guha A. Parafalcine Chondrosarcoma: Unusual Localization for a Classic Variant. Indian Journal of Neurosurgery. Mar 2019; **8(01):** 81-5.

5. Chandler JP, Yashar P, Laskin WB, Russell EJ. Intracranial chondrosarcoma: A case report and review of the literature. J Neurooncol. 2004; **68:** 33-9.

6. Korten AG, ter Berg HJ, Spincemaille GH, van der Laan RT, Van de Wel AM. Intracranial chondrosarcoma: Review of the literature and report of 15 cases. J Neurol Neurosurg Psychiatry. 1998; **65:** 8892.

7. Krishnan SS, Panigrahi M, Varma D, Madigubba S. Falcine and parasagittal intracranial chondrosarcomas of the classical variant: Report of two cases with review of literature. Neurology India. May 2011; **59(3):** 451