CHORDOID MENINGIOMA: A case report.

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ABSTRACT

"Chordoid meningioma" is an uncommon histopathological variant of meningioma. I report a case of chordoid meningioma occurring in a 53 year-old female patient. Nonenhanced CT scan of the brain showed a low density mass(25HU) with rimlike calcification and surrounding edema in left temporal lobe.The enhancement was thick-irregular ring configuration.This mass presented as intra-axial location.Its lateral margin was close to left sylvian fissure,but no border revealed dural attachment.

Surgical excision was performed and the pathologic result established the definit diagnosis.

INTRODUCTION

Meningiomas are some of the most common intracranial tumors.The 1993 World Health Organization classification of CNS tumors described 14 distinct histopathological variants.[1] One of these variants is the chordoid meningioma,a very rare subtype,that comprises approximately 0.5% or less of all meningiomas.[2]

The term "chordoid meningiomas" was first used by Kepes et al in 1988 to describe meningiomas that were composed of spindle or epithelial cells forming chordoma-like clusters and cords,in a myxoid matrix.Prominent lymphoplasmacellular infiltration was also a feature in these tumors.[3]

A literature survey revealed that the majority of case reports were discussed mainly on the clinical manifestations and histopathology. In this report,I describe a case of chordoid meningioma, highlighting the strange CT features.

CASE REPORT

A 53 year-old female patient presented with a four-year history of psychosis.Seven days before admission,she had many episodes of focal seizures with loss of consciousness.Physical and neurological examination was unremarkable.

In a routine laboratory test,the hemoglobin, mean corpuscular volume and mean corpuscular hemoglobin were 11.0g/dL,85.8fL and 27.7pg, respectively.Normocytic hypochromic anemia was noted.The total protein,albumin and globulin were 8.7g/dL,3.1g/dL and 5.6g/dL, respectively.

Axial computed tomography(CT) of the brain revealed a hypodense lesion(25HU) with rimlike calcification and moderate degree of surrounding edema in left temporal lobe(Figure 1). Intense thick irregular ring enhancement was seen on post contrast study(Figure 2).Lateral margin of the mass was close to left sylvian fissure,but no dural attachment was seen.There was no bone destruction or hyperostosis.

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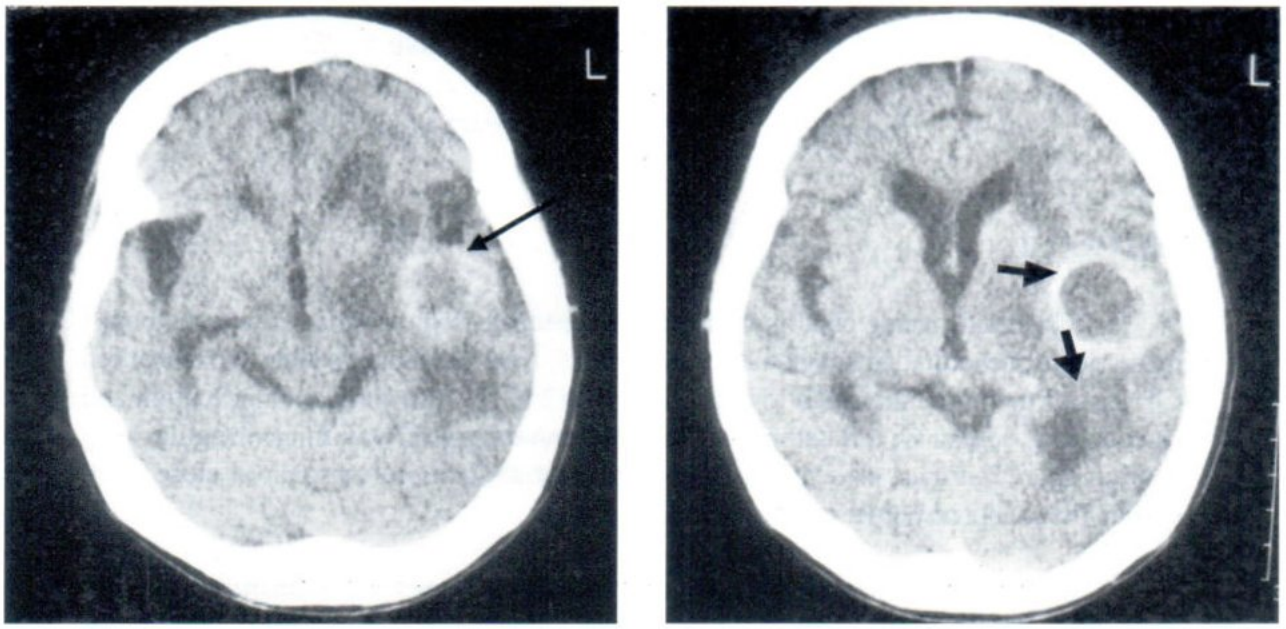

A B

Fig. 1 (A and B) Axial nonenhanced CT scan of the brain showed a low density mass (25HU) at left temporal region. Lateral margin of the mass was close to left sylvian fissure (black arrow), but no dural attachment was seen. Rimlike calcification (arrowhead) and perilesional edema were noted.

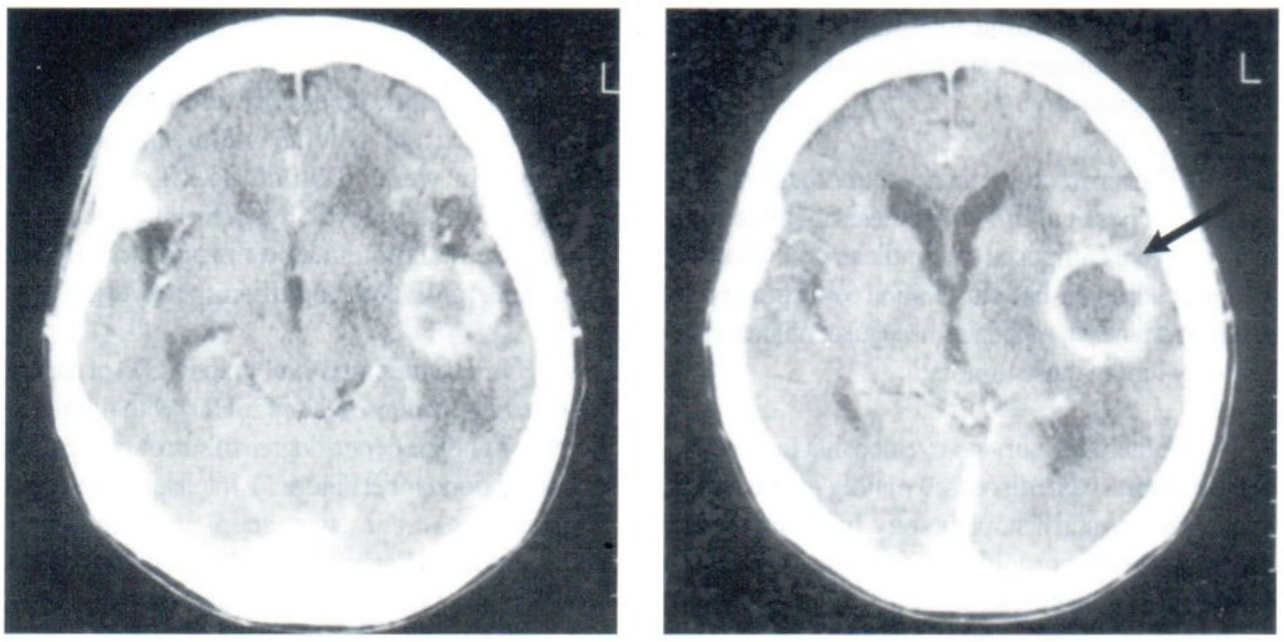

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Fig. 2(A and B) Axial contrast-enhanced CT scan of the brain showed intense thick irregular ring enhancement (black arrow) of the left temporal mass.

No precise diagnosis could be made because of its findings were not typical for any type of brain lesion.The differential diagnosis for calcified brain mass was considered among tuberculoma,astrocytoma, oligodendroglioma,ependymoma and meningioma.

The patient underwent a left temporal craniotomy for tumor resection. At surgery,the mass was intra-axial,near to left sylvian fissure and not adherent to dura.It appeared well-defined,centrally cystic (containing straw-color,high-proteinaceous fluid) with whitish hard solid wall that suspected to be calcified.The mass was 3 cm in size.Nearly all part of this mass was removed and sent for histopathological examination.Some part of this mass was left due to arterial attachment.Postoperative diagnosis was between cystic glioma and tuberculoma.

At pathology,the specimen consisted of trabeculae of eosinophilic vacuolated cells in the mucoid matrix.There were prominent lymphocytic-plasmacytic infiltrate within the stroma(Figure 3,4). Immunohistochemically,tumor cells showed a typical membranous staining of meningioma for epithelial membrane antigen in focal areas(positive EMA). None of tumor cells expressed glial fibrillary acidic protein(negative GFAP). All these findings were consistent with those of chordoid meningioma.

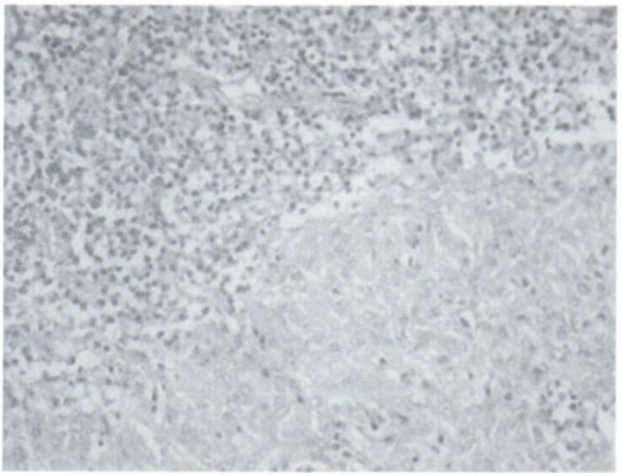

Fig. 3 The stroma (upper left) contains a lymphocytic-plasmacytic infiltrate. The tumor is seen in the lower right consisting of vacuolated cells.

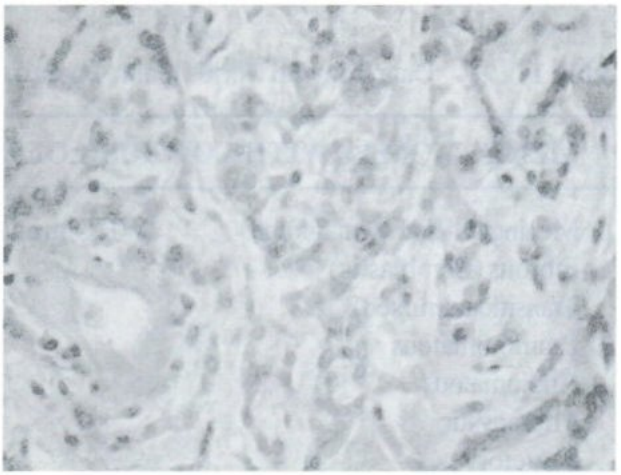

Fig. 4 The tumor is predominantly composed of eosinophilic, vacuolated cells formin trabeculae similar to chordoma.

DISCUSSION

Meningiomas are generally benign tumors comprised of neoplastic meningothelial cells of the arachnoid. They are the most common nonglial primary brain tumor and the most common intracranial extra-axial neoplasm.[4]

Meningiomas are thought to be adult tumors;peak occurrence is between 40 and 60 years of age. Incidence in women outnumbers that in men.[4]

Less than 10% of all meningiomas ever cause symptoms5.A variety of clinical signs and symptoms based on compression of surrounding structures. The patient's symptoms depend entirely on the location of the tumor;although headache,personality change, paresis and seizures are comprise of the classic presentation.[6]

Most meningiomas are extra-axial dural-based lesions.The most common sites of occurrence

included: cerebral convexities ,parasagittal region, sphenoid ridge,parasellar region ,olfactory groove and posterior fossa.[6] Approximately 2% of intracranial meningiomas have no dural attachment.These tumors arise from choroid plexus stromal cells and grow as intraventricular masses.[7]

Plain and contrast-enhanced CT scan detect 85% and 95% of intracranial meningiomas, respectively.[8] NECT scans typically show a sharply circumscribed round or smoothly lobulated mass that abuts a dural surface,usually at an obtuse angle. Approximately 70% to 75% of all meningiomas are homogeneously hyperdense relative to adjacent brain, 25% appear isodense. Hypodense tumors are seen in 1% to 5% of cases and true cystic meningiomas with large intratumoral fluid-filled cysts are also uncommon.[4] Calcification is seen in 20% to 25% and can be diffuse,focal, sandlike,sunburst,globular and even rimlike patterns.[4] Peripheral edema is seen in 60% of cases and may be extensive. CECT scans show intense, relatively uniform enhancement in 90% of all cases. 10% to 15% of meningiomas have an atypical pattern with rimlike tumor enhancement.[9-10]

There are many different histologic subtypes of meningiomas.The WHO classified variable histologic variants based on risk of recurrence and aggressive behavior (Table 1)

Table 1 Data from the world Health Organization classification of Tumors

MENINGIOMAS WITH LOW RISK OF RECURRENCE AND AGGRESSIVE GROWTH	
Meningiothelial meningioma	WHO grade I
Fibrous (fibroblastic)	
Transitional(mixed)	
Psammomatous	
Angiomatous	
Microcystic	
Secretory	
Lymphoplasmacytic-rich	
Metaplastic	
MENINGIOMAS WITH GREATER LIKELIHOOD OF RECURRENCE AND/OR AGGRESSIVE BEHAVIOR	
Atypical	WHO grade II
Clear cell	
Chordoid	
Papillary	WHO grade III
Anaplastic (malignant)	

NECT = Non-enhanced CT., **CECT** = Contrast-enhanced CT.

Kepes et al for the first time, described "Chordoid meningioma"as a distinct meningeal tumor occurring in children and this was associated with systemic manifestations such as refractory microcytic anemia, hypergammaglobulinemia and angiofollicular lymphoid hyperplasia (Castleman disease).[11]

Histopathologically, these tumors were composed of spindle or epithelial cells forming chordoma-like clusters and were scattered in a myxoid matrix. Prominent lymphoplasmacellular infiltration was also a feature in these tumors.[3] In a large series of 42 patients with mean age about 44 years old studied by Couce et al and a 33 year-old case reported by Je-young Yeon et al observed that none of their cases had any systemic manifestations as reported by Kepes et al.They expressed the possibility that the systemic manifestations associated with this tumor are limited to chordoid meningiomas occurring in the childhood.[2,12]

De Tella Jr. et al reported CT findings of a case of chordoid meningioma who showed a large high density mass at right temporal region with dense contrast enhancement and minimal peritumoral edema.[13] Varma et al reported two cases of chordoid meningioma. The first case appeared isodense lesion with heterogeneous enhancement and mild perilesional edema in right frontal parasagittal region. The second case showed a well-defined heterogeneous hypodense mass in right frontal parasagittal region, a broad base towards the anterior third of the falx cerebri with heterogeneous enhancement and moderate perilesional edema causing mass effect and subfalcine herniation.[14]

Similar findings of the CT images of our case and the previously reported cases is evident of perilesional edema, but the mass density, location and pattern of enhancement are different.

In our case,because it appeared intra-axial location,with large intratumoral cyst,rimlike calcification and ring enhancement,which are not classic pattern of meningiomas,leading to differential diagnosis among intra-axial lesions such as astrocytoma, oligodendroglioma, ependymoma and tuberculoma. Even surgical findings did not indicate meningioma. Pathological and immunohistochemical diagnosis was necessary.

Generally,meningioma is considered for a mass adherent to falx or dura, however,we found that meningioma cannot be entirely excluded for an intra-axial mass as well.

Chordoid meningioma is classified as WHO gradeII, by its aggressive behavior and high recurrence rate, particularly after subtotal resection. Periodic follow-up and imaging studies after surgery are indicated.[6]

CONCLUSION

I report unusual or misleading CT features that may not be suggestive of meningioma such as intra-axial location, large intratumoral cyst,rimlike calcification and ring enhancement that proved by histopathology to be a very rare subtype,chordoid meningioma. Because meningiomas are common,the radiologist must be aware of their less frequent and uncharacteristic imaging features in order to suggest the correct diagnosis in cases that are atypical.

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