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Case Report

Giant Occipital Chondroma; An Uncommon Presentation of A Rare Entity

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Abstract

Intracranial chondroma is a rare slow-growing benign tumour typically developed from cartilaginous rests of skull base synchondroses. The clinical symptoms are variable depending on the location and the size of the lesion. We report here a case of an 18 -year-old male who presented with headaches and worsening confusion. Computed tomography scan and magnetic resonance imaging of the brain revealed a giant extra-axial mass in parieto-occipital region, with heterogeneous coarse calcification and chondroid matrix, resulting in significant mass effect, displacement of adjacent structures, and associated obstructive hydrocephalus. Chondroid tumor was evoked and differential diagnosis such as meningioma was considered less likely due to hypo vascular appearance on cerebral angiogram. Adjacent bone remodelling and development of collateral venous outflow suggested a slow-growing tumor. A gross total resection was achieved and the pathologic evaluation confirmed the diagnosis of chondroma. We describe here the different steps and imaging features which could be helpful to evoke the diagnosis of this rare entity, and also discuss the relevant differential diagnosis.

Keywords: Intracranial Chondroma; Occipital Falx; Extra-Axial; Chondroid Tumor; Obstructive Hydrocephalus

Abbreviations

CT-Scan: Computed Tomography Scan; MRI: Magnetic Resonance Imaging; CSF: Cerebrospinal Fluid; DTI: Diffusion Tensor Imaging; CK: Cytokeratin; EMA: Epithelial Membrane Antigen

Introduction

Intracranial chondromas are rare benign tumors accounting for 0.3% [1] of intracranial tumors, mostly seen during the third decade of life [2].

Even if chondromas can arise from different intra cranial structures (ventricle and choroid plexus, dura mater, brain parenchyma), the majority arises from the base of the skull [3]. It can be explained by the most commonly accepted physiopathology based on rests of cartilaginous cells along the skull base which bones are embryologically derived from cartilage [4]. Falx origin are a rare presentation with usually involvement of the fronto-parietal lobe [5].

In our case, a giant heterogeneous extra-axial chondroma was extended in the parieto-occipital lobe with mass effect on the posterior horns of lateral ventricles. Here we discuss the steps for the imaging evaluation through CT-scan, MRI and cerebral angiogram findings which led to diagnosis of this case.

Case Report

An 18 -year-old male presented with two months worsening headaches, confusion and abnormal sensation in the back of the head. He had a history of occipital headaches over the past three years on almost daily basis which are triggered by lifting heavy weights. The headaches had been increasing in frequency and severity over time. He had no other past medical history. The neurological examination was intact.

A CT-scan revealed a large lobulated mass in the parieto-occipital region with displacement of the surrounding brain structures. The lesion is well defined with hyperdense rim, appeared hetero-

geneous with dense rings and arcs calcifications, and fluid contents. There is no significant surrounding edema. The mass results in inferior displacement of the tentorium and effacement of the forth ventricle. There was obstructive hydrocephalus at the initial CT-scan (Figure 1). Mass appears to be partially attached to the falx, suggesting the extra axial origin. There was subtle thinning of the adjacent occipital bone without destruction, suggesting a slow growing process (Figure 2).

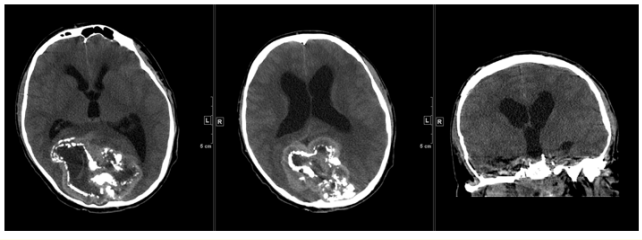

Figure 1: Axial and coronal non-contrast CT showing the obstructive hydrocephalus due to the lesion.

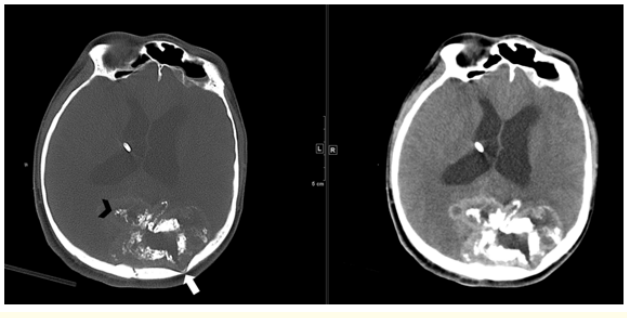

Figure 2: Non-contrast CT of the brain shows a large amorphous mass with areas of coarse calcification including "ring and arc" calcifications (black arrowhead). There is focal area of bone remodeling in the left calvarium (white arrow). The central region of the mass shows fluid density material

Regarding the findings and the poor airway reflexes, the patient was intubated for airway protection and a shunt catheter was placed. Intracranial pressure monitoring was performed. Subtle improvement of the clinical status and the intracranial pressure were noted after shunt placement. Laboratory testing including parasital, fungal, bacterial and viral examinations were unremarkable. CSF review was unremarkable without pleocytosis or malignant cells. A body CT-scan and testicular ultrasound were also made to rule out a metastatic process.

MRI confirmed extra-axial origin with demonstration of a clear CSF cleft. The internal content of the mass was heterogeneous as seen in the CT-scan. Small and predominantly peripheral enhancement was seen without significant enhancement within the internal content of the mass (Figure 3). The major contents of the mass are calcifications (Figure 4), proteinaceous material, and small areas of evolving blood products (Figure 5). No surrounding edema was found. The MRI also revealed optic nerve sheaths dilatation and CSF compression of the pituitary gland, suggesting increased intracranial pressure. The DTI showed an anterior displacement of the different tracts, but no evidence for an intra parenchymal mass.

Figure 3: Non-contrast T1(a) and post-contrast T1(b) weighted images show a large amorphous extra-axial mass with peripheral rim enhancement (arrow), without significant enhancement in the internal mass.

Figure 4: Axial T2 weighted image (a) shows cystic component of the mass (white arrowhead). Gradient echo image (b) shows calcifications (black arrowhead).

Figure 5: Non-contrast T1(a), T2(b), T2*(c), diffusion(d) and apparent diffusion coefficient(e) images show blood containing material.

The angiogram showed no tumoral stain in association with the mass. Portions of superior sagittal sinus and transverse sinuses were occluded. Several prominent collateral veins were noted suggesting a slow growing process (Figure 6).

Figure 6: Angiograms showing no tumor staining in either external carotid artery(a) or internal carotid artery(b) injections. On venous phase(c), lack of opacification by compression of the superior sagittal sinus (black arrow) and collateral vein development (white arrows). Note the inferior displacement of the straight sinus by the mass (star).

Based on the imaging findings, the diagnosis of a mesenchymal origin tumor was suspected including chondroid tumor and a surgical resection was proposed.

The patient underwent a left parieto-occipital craniotomy for gross total resection. The intra-operative findings were a calcified cartilaginous extra-axial mass crossing over midline with compression along the posterior sagittal sinus and transverse sinus, and remodeling of surrounding bone. The post-operative MRI confirmed gross total resection.

Until one month after surgery the ventricular caliber were unchanged, however the inferior descend of cerebellar tonsils and the CSF signal surrounding the craniocervical junction were significantly improved. Pathology review showed bland appearing hyaline cartilage with calcification with no malignancy identified. The final diagnosis of giant intracranial chondroma was made. No further treatment than the initial complete resection was so required. Two weeks after surgery, the patient was discharged. No pain, fever or any new neurological problem during the interval were noted at the first post-operative visit two months after discharge, except a residual numbness to right lower extremity in improvement. The follow-up MRI showed significant improvement in the mass effect and hydrocephalus (Figure 7). To date, with five months follow-up, there has been no new neurological or other symptoms.

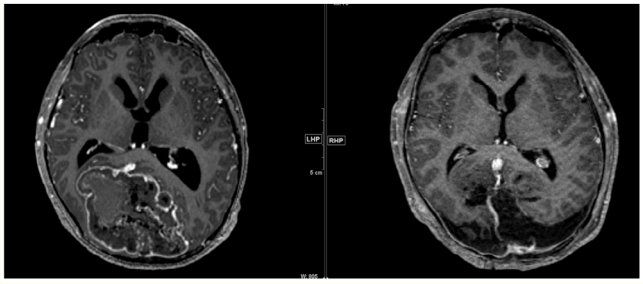

Figure 7: Axial post-contrast T1 MRI showing gross total resection and improvement after surgery (right) of the hydrocephalus.

Discussion

Intracranial chondromas are rare slow growing tumors with relatively large size at presentation. After an asymptomatic phase, headaches [3] or cranial nerve deficit can be the presenting symptoms, depending on the location. Intracranial chondromas can be found in Ollier's multiple chondromatosis [6] or Maffucci syndrome [7]. Basal chondromas arise mainly from the synchondroses covering the foramen lacerum [8] but other intracranial location such

as convexity can be found, wherever there is aberrant embryonal cartilage rests. The imaging features have to be known to eliminate the differential diagnosis especially chondrosarcoma, but also chordoma and meningioma.

A well-delimitated, lobular lesion with irregular as well as "ring and arc" calcifications, associated with adjacent bone remodeling [9,10] with rim enhancement are the most typical characteristics of chondroma. Speckled and irregular calcifications are commonly seen (about 60%) but there are cases of poorly calcified chondromas where calcifications increase through years, without tumor enlargement [11].

Some authors proposed high-dose contrast medium injection showing delayed enhancement to differentiate chondromas from meningiomas [12]. In the difficult cases, to eliminate the differential diagnosis of a meningioma, an arteriogram is useful. Chondromas are avascular and so no tumor stain is found, whereas a late-capillary and early-venous tumor blush from the meningeal arterial supply is typically found in meningiomas [13]. No vascular abnormality is found in chondroma cases but a displacement of vessels due to mass effect can be seen.

Chordoma is another differential diagnosis which can be evoke through location but also immunohistochemical criteria. A midline location in the clivus and positive staining for CK, EMA and S - 100 protein suggest a chordoma [14].

In our case, the partially attachment to the falx and the presence of CSF between the mass and the adjacent displaced brain tissue confirmed the extra-axial origin of the lesion. The adjacent bone remodeling and development of additional collateral venous outflow were consistent with a slow growing lesion. Imaging features were not consistent with typical meningioma, especially the absence of mass enhancement confirmed with the arteriogram. The mass showed no aneurysms or abnormally high-flow, no early draining veins and no regions of abnormal hypervascularity.

Based on the imaging, the most consistent diagnosis evoked was a mesenchymal origin for the tumor, especially chondroid tumor. The differential diagnosis with chondrosarcoma was evoked and pathology after surgical resection was waited for final diagnosis.

Chondrosarcoma, which represents 0.15% of intracranial tumors [15] has to be rule out since its malignant implication and therapy. Intracranial chondrosarcomas arise preferentially in the skull base in approximately 75% [16], due to its development through rest of endochondral cartilage too. Different histological subtypes of chondrosarcomas exist but the only two types occurring intracranially are the conventional and mesenchymal subtypes [17]. If no specific imaging features can formally differentiate chondromas from chondrosarcomas, some histopathological findings can make the diagnosis. The presence of binucleated chondrocytes is one of the most important feature to make the diagnosis of well-differentiated (grade I) chondrosarcomas instead of chondroma [18]. Also, irregular lobules with cellular fibrous tissue around the tumor are signs for chondrosarcomas. A multi-parameters criteria can also help to differentiate these two entities [19].

Even if the preliminary result of pathology here was concerning for chondrosarcoma, the final diagnosis was made through the lack of evidence of malignancy.

The complete resection of the chondroma is the definitive treatment of choice with no need of adjuvant therapy [20]. The diagnosis of chondrosarcoma has to be rule out since even if the complete resection is also the mainstay treatment, a combined adjuvant radiation and chemotherapy are required to improve recurrence rate and overall survival [17].

Conclusion

Intracranial extra-axial chondroma is a rare benign cartilaginous lesion which can be responsible of various symptoms because of the mass effect. Some imaging features should suggest the diagnosis of a slow-growing chondroid tumor. An angiogram could be useful to eliminate some differential diagnosis such as meningioma. The treatment of choice is complete surgical resection. The pathology should confirm the diagnosis and especially eliminate a chondrosarcoma which would require adjuvant therapies.

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Conflict of Interest

None.

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