

When Chondroma Happens in an Unexpected Location: A Case Report of Intra-Axial Intracranial Chondroma

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Abstract: Chondroma is a benign tumor of mature hyaline cartilage that is often found in the long bones and may be rarely diagnosed in other parts of the body. Here, we present a young patient with the definitive diagnosis of intra-axial intracranial chondroma and without dural connection. The presenting symptoms of the patient were headache and impaired vision. The brain magnetic resonance imaging (MRI) revealed a huge enhancing parasagittal brain mass. The diagnosis was confirmed by immunohistochemistry, which was positive for S100.

Keywords: case report, intracranial chondroma, rare malignant tumors

Introduction

Chondroma is a benign tumor of mature hyaline cartilage, which is often found in the long bones of the upper and lower limbs or pelvis.^{1,2} Intracranial chondromas are rare and account for only 0.2% of intracranial primary tumors.³ Most of the reported cases of intracranial chondromas are sporadic, although they have also been reported in the settings of Maffucci's and Ollier's syndromes.⁴ The first case of intracranial chondroma was reported by Hirschfeld in 1851, who at the time believed that the tumor originates from the remnants of embryonic chondrogenic cells along the basement membrane of synchondrosis. So far, several hypotheses describe the pathogenesis of this lesion. Intracranial chondromas reported so far have often been extradural and were located in the sphenoid-occipital area at the skull base and with dural connection, although less frequent cases of intracranial chondroma in the paranasal sinuses, choroid plexus, or dura have also been reported.⁵⁻⁸ Interestingly, a very limited number of reported intracranial chondromas have been intraparenchymal without dural connection.^{9,10} Here, we present a young patient with the definite diagnosis of intra-axial intracranial chondroma without dural connection.

Case Presentation

A 36-year-old woman presented with the complaint of occasional headaches since one year ago that has exacerbated since one week before the presentation and accompanied by blurred vision. She had no significant past medical or drug history. As the physical examination was unremarkable, brain MRI without gadolinium-based contrast agents was requested, which revealed a large, low-signal, heterogeneous mass in T2 and T1 MRI sequences in the right frontal lobe, extending to the right parietal lobe, and with

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significant midline shift to the left. To determine the nature of the mass, MRI with contrast injection was performed, showing a peripheral, punctuated enhancing lesion. Given the imaging findings, the differential diagnoses of low-grade glioma and brain lymphoma were rendered (Figure 1). The patient underwent craniotomy and the tissue sample was sent for histologic examination. Macroscopic examination showed multiple fragmented, pale-blue tissue fragments with elastic consistency, resembling cartilage, and totally measuring 8 * 8 * 3 cm. Primary microscopic examination described a neoplastic lesion with cartilaginous lobules and the appearance of whorls and gave the final diagnosis of metaplastic meningioma, WHO grade I. As radiology did not correlate with pathology, the specimen was sent to a neuroradiologist and a neuropathologist for second opinion. The neuroradiologist described a huge intra-axial mass in the

frontoparietal area with compression and midline shifts that was non-contrast enhancing. The diagnosis of meningioma and hemangiopericytoma was excluded radiologically as they are contrast-enhancing and the definite radiologic diagnosis was not given. Examination of the specimen by the neuropathologist described a mesenchymal neoplasm, consisting of chondroid lobules without atypia, or necrosis. There was no evidence of brain parenchyma or dura (Figure 2A). As there was no brain tissue or dura in the specimen, the possibility of specimen displacement in the operating room was suggested. However, there was no related surgery in that day. To determine the exact nature of the mass, immunohistochemical staining for EMA, CK, GFAP, and S100 markers was performed, which was positive for S100 and negative for the other three markers (Figure 2B). The final diagnosis was intracranial chondroma.

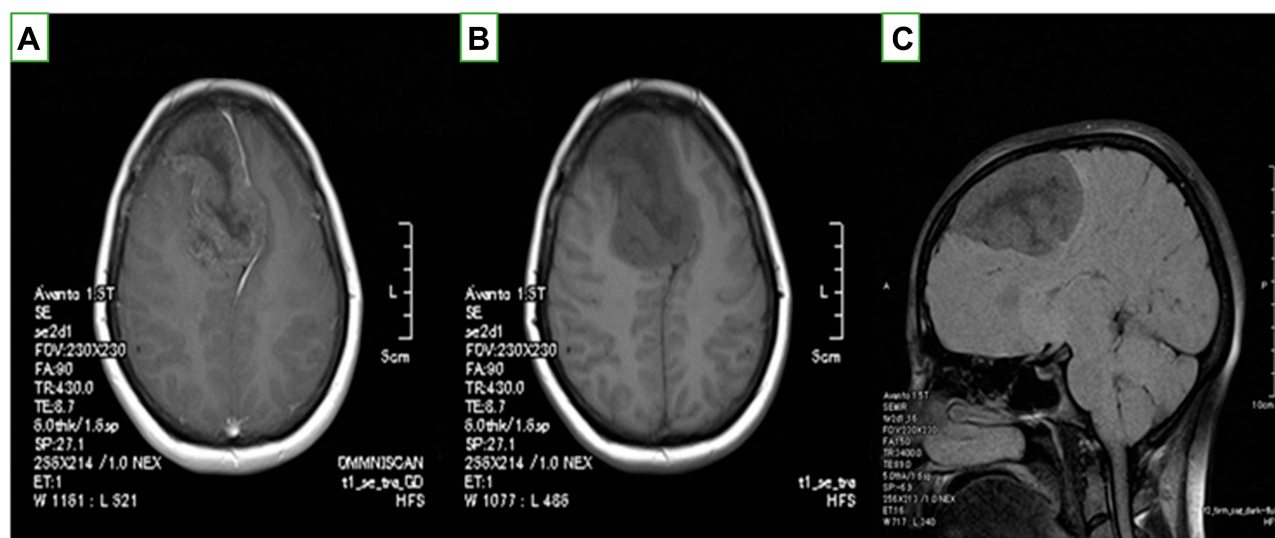


Figure 1 Brain MRI of the patient: axial plane of T1-weighted brain MRI (A) with and (B) without contrast. (C) Sagittal plane of T1-weighted brain MRI. MRI of the brain shows a low-signal, heterogeneous mass in T2 and T1 in the right frontal lobe area, extending to the right parietal area and with significant midline shift to the left. After contrast injection, a punctuate enhancement was reported.

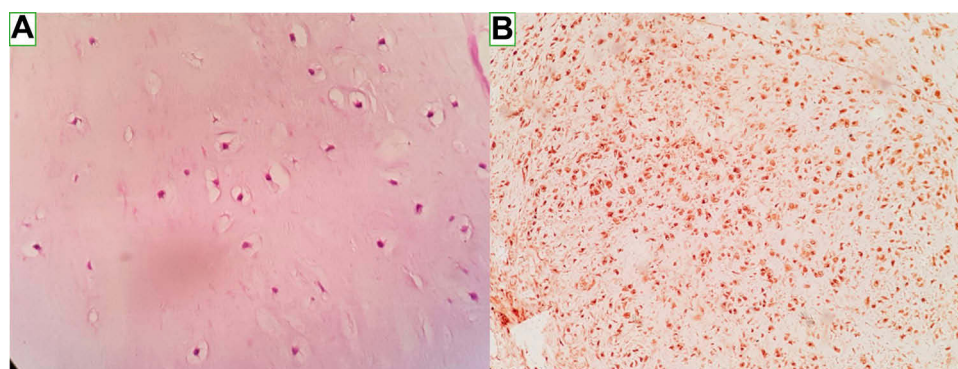


Figure 2 (A) Hematoxylin and eosin staining, 400x, showing cartilaginous lobules without atypia, mitosis, or necrosis and (B) positive IHC staining of S100 marker.

Discussion

Primary intracranial chondroma is quite rare and consists of only 0.2% of primary intracranial tumors.³ To the best of our knowledge, 163 cases of intracranial chondroma have been reported, which were mostly originated from the skull base, falx, and dura. The skull and later the fossa and dura area originated (Table 1). Only a few of these cases were similar to this case and there was no association between the tumor and the dura. There was no significant gender predilection in intracranial chondromas.^{11–14} The reported cases mostly presented in the third decade of life,¹⁵ although the age range was 15 months to 60 years.¹⁶ Due to the slow-growing nature of the tumor, patients with intracranial chondromas often present with a large mass and a long history of symptoms at the time of presentation, which may vary depending on the location of the tumor.¹⁷ The most commonly reported symptoms are headache and seizures (Table 2). However, focal neurological deficits have also been reported as a primary symptom, as well as Parkinson-like symptoms.¹⁸

Imaging characteristics of intracranial chondromas can be hypo or hyperdense, based on its degree of calcification. On MRI, chondromas appear iso to hypointense in T1 and as a mixture of iso to hyperintense in T2 images.¹⁹ Sullivan et al showed that most of the lesions were hypointense in T1 weighted brain MRI with a heterogeneous appearance in T2 weighted brain MRI.¹⁶

Table 1 Origin of Intracranial Chondromas in the Literature

Origin	Number (%)
Skull base	123(74.5)
Falx	22(13.3)
Dura	11(6.7)
Unknown	6(3.6)
No dural involvement	2(1.2)
Tentorium	1(0.6)

Note: Data from Sullivan et al.¹⁶

Table 2 Common Symptoms of Intracranial Chondromas in the Literature¹⁶

Common Symptoms	Number (%)
Headache	31(52.5)
Diplopia	7(11.9)
Seizure	7(11.9)
Papilledema	5(8.5)
Weakness	5(8.5)
Dizziness	4(0.4)

As most cases of intracranial chondromas are dura-attached (unlike this patient), one of the important differential diagnoses is meningioma. Intracranial chondroma, like meningioma, presents as a well-circumscribed, extra-axial, and dura-based mass, although in very rare cases, chondromas have no dural connection and have heterogeneous enhancement, which differentiates these lesions radiologically from meningiomas.⁵ The differentiation of chondromas from other soft-tissue tumors that can involve the CNS and have similar imaging findings (such as chordoma, chondrosarcoma, and hemangiopericytoma) is very important. Chordoma originates from notochord remnants and is often located in the midline. Chondrosarcoma also tends to be situated in the midline, although it is sometimes found in the lateral hemisphere, and has malignant histological features (hypercellularity, cellular atypia, and increased mitotic activity) with progressive and destructive borders in imaging and histology.²⁰

In all reported cases, intracranial chondromas were white- gray, multilobulated masses with calcification, similar to this case. Microscopic examination reveals a well-circumscribed mass with a fibrotic capsule, consisting of single or binary chondrocytes within the lacunae in a chondroid matrix. In the reported cases, which used immunohistochemical staining as well, tumoral cells were positive for S100 and vimentin, and negative for EMA, and CK.¹⁶ The important point in the pathologic study of these lesions is to consider its important differential diagnoses, including meningiomas, chordoma, osteochondroma, and well-differentiated chondrosarcoma.²⁰ There have also been reports of simultaneous occurrence of two different types of tumors in one patient and in one anatomic location (collision tumor), which included the simultaneous occurrence of choroid plexus papilloma and chondroma.²¹

Intracranial chondromas may occur singly or in association with systemic chondromatosis such as Maffucci's syndrome or Ollier's disease.^{22–25} Both Maffucci's syndrome and Ollier's disease are caused by a somatic mutation in the IDH-1 and IDH-2 genes, encoding the isocitrate dehydrogenase 2 enzymes. However, according to the previous studies, in sporadic intracranial chondromas, no mutation in the IDH genes was reported.^{26,27} Multiple theories have been presented about the origin of intracranial chondromas, most of which consider its origin from the embryonic chondrogenic cells remnants along the basement membrane of synchondrosis.²⁸ Although the theory of metaplasia of meningeal fibroblasts or perivascular mesenchymal tissue fits for dura-attached cases, other theories such as ectopic embryologic rest of

cartilaginous cells or the inappropriate replacement of cartilaginous elements during trauma are also given.^{11,28}

Conclusion

Intracranial chondroma should be considered as a differential diagnosis in the brain parenchyma and tissue sampling should be performed for histopathologic confirmation of these rare entities.

Ethical Consideration

Approval of institutional review board from the Ethics Committee of Mashhad University of Medical Sciences was obtained proactively and a written informed consent document was obtained from the patient for the details and images to be published.

Disclosure

The authors report no conflicts of interest in this work.

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