# **Chondroma of Cerebral Falx: A Rare Intracranial Diagnosis**

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## ABSTRACT

Chondromas are benign tumors which mostly occur in extremities but also sometimes in the cranium. Intracerebral chondroma is rare condition. Most intracranial chondromas arise from skull base, but chondroma of falx origin is a rare entity and mostly occurs in relation with syndromic disorders such as Mafucci's syndrome or Ollier's syndrome. Here, we report a rare case of falcine intracranial chondroma in a young man who presented with headaches and weakness of lower extremities and no signs of any syndromic disorder. The purpose of this case report was to raise awareness about intracranial chondroma should be considered in the differential diagnosis of calcified masses arising from the falx.

Key Words: Chondroma. Falx. Neoplasms. Benign. Calcification.

## INTRODUCTION

Chondroma is a common cartilaginous tumor which is found at several sites of the body, most frequently in the small bones of the hands and feet but rarely as intracranial tumors.<sup>1</sup> The estimated incidence is 0.2 - 0.3% of all intracranial tumors.<sup>2</sup> Most intracranial chondromas arise from skull base, but chondroma of falx origin is a rare incidence. Indeed, the intracranial chondromas rise from falx is mostly in relation with syndromic disorders such as Mafucci's syndrome or Ollier's syndrome.<sup>3</sup>

Here, we describe a non-syndrome falcine chondroma in a young man.

## **CASE REPORT**

A 20-year male patient was admitted with 6 months history of intermittent headache. He was used to take painkiller (Aspirin or Ibuprofen) to relieve the headache, but recently these had not been as effective as before. On physical examination, except for mild weakness of lower extremities, more on left side than right, no positive neurological signs were found. Head CT scanning revealed an oval lesion between frontal lobes, with homogenous hypo-density containing a large region of hyper-density in its rim (Figure 1). Its size was 12.7 x  $6.5 \times 5.0 \text{ cm}$  and it was located above the corpus callosum, between frontal lobes. Its capsule showed little enhancement on contrast images. During surgery, inter-hemispheric approach was used to reach the lesion. The lesion was enveloped by a thick white

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Figure 1: Pre-operative CT scan showing an oval heterogeneous lesion hypodense core and hyperdense/calcified wall.

capsule, which was subsequently proved to be a part of the cerebral falx (Figure 2). Firstly, the capsule was cut at the top end. It was very hard bone like material all around a cartilaginous core. After debulking the center, the tumor was removed *enblock*. Patho-histological examination of the removed tissues demonstrated a tumor comprising of lobules of mature cartilage with scattered thin walled blood vessels and areas of calcification. Some areas showed haemorrhage and necrosis. Postoperative recovery was uneventful and patient was discharged one week later. At 3 weeks follow-up, no neurological deficits were found. Patient is being followed for future recurrence.

Gross appearance showed central haemorrhagic and friable areas surrounded by thick cartilaginous tissue measuring. Micro-sections showed a tumor comprising of lobules of mature cartilage with scattered thin walled blood vessels and areas of calcification. Some areas showed haemorrhage and necrosis. There was no evidence of malignancy.



Figure 2: Gross appearance of removed tissue.



Figure 3: Histopathology of the resected lesion showing lobules of mature cartilage.

### DISCUSSION

Intracranial chondromas are rare tumors that account for approximately 0.2 - 0.3% of all intracranial neoplasms. They usually occur in young adults with peak prevalence in the third decade. This patient was 20 years of age. However, patients ranging from 15 months to 60 years of age have been reported.4,5 There is no gender predilection.<sup>4</sup> They can grow as solitary tumors, or as components of Ollier disease and Maffucci's syndrome.6 Some cases have been reported to arise at sites of previous trauma.<sup>4</sup> The disease-related chondromas have a tendency to be hypercellular with atypical cytology. It is also well known that patients with Maffucci's syndrome have increased risk of malignant change.4,5 Very rarely, chondromas presenting concomitantly with other glial tumors have been reported in the literature.1

The majority of intracranial chondromas develop from the cartilage found in the basilar synchondroses at the skull base. Tumors arising in the dural portions, leptomeninges, brain parenchyma, ventricles or choroid plexus are less common.<sup>7,8</sup> Several theories have been proposed in the literature explaining the pathogenetic mechanism of development of convexity or falcine chondromas. These include heterotopic chondrocytes, cartilaginous metaplasia of meningeal fibroblasts, abnormal multipotential mesenchymal dural cells or their differentiated cellular descendants, and traumatic displacement of cartilage.<sup>9</sup> However, the exact pathogenetic mechanism is still uncertain.

Due to the non-invasive and slow-growing nature of chondromas, patients often present with a long-standing history of headache and symptoms of increased intracranial pressure. Patients may have signs and symptoms related to compression of adjacent structure, such as seizure, personality changes, and hemiparesis.<sup>10</sup> It has been reported that the mean diameter of these tumors is 6 cm, and their mean weight is 170 gram.<sup>1</sup> Recently, Patel et al. have described a giant cystic intracranial chondroma of the falx measuring 12 x 10 x 7 cm and weighing 480 grams.<sup>4</sup> In this case size of the lesion was 12.7 x 6.5 x 5.0 cm that was comparable to case reported by Patel et al. According to Lacerte et al. intradural chondromas have two distinct CT scan presentations.<sup>9</sup> Type 1 (classical) is more common and reveals mixed density with minimal or moderate enhancement, whereas type 2 is less frequent and has a central hypodense area, which is composed of cystic degeneration or of a very loose-texture of connective tissue without necrosis in pathological evaluation. Neuroimaging of these tumors are non-pathognomonic. The most frequently encountered imaging features include a well-circumscribed mass with calcifications in 60 - 90% of cases, associated hyperostosis, erosion and destruction of surrounding bone in 50 - 60% of cases, and usually mild to moderate, patchy contrast enhancement. Falcine or convexity chondromas typically show no or minimal peritumoral edema, which demonstrates the slow-growing, benign nature of these tumors.<sup>4</sup> In this case on CT scan there was hypodense lesion with surrounding rim of hyperdensity and there was capsular enhancement on contrast images. There was no peritumoral edema. Cerebral angiography has revealed avascularity in nearly all published cases in the literature.1

The differential diagnosis includes craniopharyngioma or meningioma when the tumor is predominantly destructive and only partially calcified; in chordoma the calcified tumor extends near the clivus and cerebellopontine angle; but acoustic neuroma, meningioma, epidermoid or metastasis will be considered in the differential diagnosis when the tumor is non-calcified and causes destruction of the petrous apex.<sup>5</sup> In this case, the most common differential diagnosis was meningioma, epidermoid and teratoma. Complete surgical resection together with the dural attachment is the treatment of choice in these tumors.<sup>11</sup> Microscopic examination in this study was in accordance with most of other reports of cranial chondromas. Histological sections show a tumor comprising of lobules of mature cartilage with scattered thin walled blood vessels and areas of calcification. Some areas show haemorrhage and necrosis. In a benign chondroma, rapid recurrence, invasion, or metastasis, the diagnosis of chondrosarcoma should be suspected and the histology specimen should be reviewed for correct diagnosis. Because chondromas are radioresistant tumors and radiotherapy may increase the risk of malignant transformation, postoperative radiation therapy is not advised in cases of subtotal resection.2,4-7 After total resection, long-term prognosis is excellent with no recurrences reported so far. However, malignant degeneration may occur after subtotal resection, and should always be a matter for consideration especially in recurrent cases.<sup>4,7-9</sup> On the whole, it can be concluded that in tumors of falx cerebri with unusual features, chondroma should also be considered in the differential diagnosis.

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