Extra Skeletal Intracranial Chondroma of Falcine Origin: Case report and Review of Article

Abstract:
Intracranial chondroma is a benign condition accounting for 0.2-0.3% of all intracranial tumors. Most are found at the skull base near to the sphenop-ethmoidal and sphenop-occipital synchondrosis but chondroma of falc origin is a rare circumstance. We present a patient with a falcechondroma and also review of recent literatures on intracranial chondromas regarding incidence, pathophysiologic origin, clinical symptoms, imaging, histopathology and prognosis.

Key words: Chordoma, extraskeletal, intracranial, falce.


Case Report:
A 30-year-old right handed female patient presented with headache, new onset seizures, urinary incontinence, unsteady gait with drop attack. She had no remarkable past medical history. Her general physical and neurological examinations were normal.

Computed tomography (CT) scanning showed a giant non-enhancing space-occupying mass in left frontal region.

Magnetic resonance imaging showed a relatively large extra-axial mass measuring 6.5x5x5 cm in diameters with small fluid intensity zone at the antero-lateral portion based on anterior falx and located in left frontal region, with no significant enhancement after gadolinium. She underwent craniotomy and complete removal of tumor. The tumor was firm in consistency, relatively avascular and falx based origin.
Table-I

<table>
<thead>
<tr>
<th>Marker</th>
<th>Chondrocyte</th>
<th>Dura (Arachnoid cell)</th>
</tr>
</thead>
<tbody>
<tr>
<td>S100</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>EMA</td>
<td>-</td>
<td>+</td>
</tr>
</tbody>
</table>

Immunohistochemical properties of specimen are shown in table 1.

**Literature review**

Intracranial chondroma is a very rare differential diagnosis of a falx implanted tumor, since it accounts...
for less than 0.5% of primary intracranial tumors.\textsuperscript{7} They are usually solitaire except when associated with Maffucci syndrome or Ollier multiple enchondromatosis and commonly located at the skull base.

**Clinical Features**
The clinical manifestations are nonspecific and depend mainly on its size and location. Thus, it can be a neurologic deficit, psychologic, or personality change, or even intracranial hypertension as any other slow growing tumor.

**Pathogenesis**
Its origin is not clear, with some studies mentioning an origin from meningeal fibroblasts metaplasia, others claiming that the tumor may arise from multipotential or perivascular mesenchymal cells or still, from aberrant nests of cartilage forming cells in the dura.

**Radiological evaluation**
On CT, the tumor appears as a well-circumscribed extra-axial mass with calcifications in at least 60% of cases.\textsuperscript{8}

On MRI, they show heterogeneous signal, due to the association of a cartilaginous matrix (characteristically hyperintense on T2 and hypointense on T1) with multiple calcifications (usually hypointense on T2/T2 and hyperintense on T1). After gadolinium these lesions present slight, heterogeneous enhancement, mainly at the periphery.

**Differential diagnosis**
Meningioma, chondrosarcoma

**Treatment options**
When indicated, surgical removal of the tumor with its dural attachment is the treatment of choice. Partial removal can be considered for neural decompression when there is high risk of eloquent structures damage with a more aggressive approach.

**Prognosis**
The intracranial chondromas usually follow a benign course with a slow growth rate and favorable prognosis.

**Discussion:**
Intracranial chondromas are rare benign tumors, which, usually, grow as solitary lesions, although an association with Ollier disease [and Maffucci’s syndrome has been reported. The majority of patients are between 20 and 60 years of age with a high frequency around the third decade. Although a slight female preference has been reported, there is no gender predominance. Our patient is a 48 years old female.

The first case of intracranial chondroma was reported in 1851, but only in 1982 the first surgical resection was reported. Depending on various authors, between 14-15 cases were reported in the literature.\textsuperscript{9} In a very recent study, Zivkovic et al. summarized 16 cases, including their, of falx cerebri chondroma.\textsuperscript{10}

The patients usually present with long time history of symptoms and signs because of the slow growing nature of these tumors.\textsuperscript{11} At time of surgery, most of the tumors are usually large. The clinical presentation of the tumor is mild and non-specific and mostly depends on anatomic location. Manifestations of the tumor are related to dysfunctions that are secondary to either, local parenchymal compression, epileptic seizures or increased intracranial pressure [23,24]. Our patient presented intermittent headaches, lack of concentration, and three months history of difficulty in naming objects and mild right hemiparesis. It has been reported that the mean diameter of convexity chondromas is 6 cm.\textsuperscript{12} In our case, the size of the tumor was $6.5 \times 5 \times 5$ cm.

The most frequently encountered imaging features of falx chondromas on CT scan and MRI include a well-circumscribed and demarcated mass, with mild to moderate patchy gadolinium enhancement, and a minimal peritumoral edema.\textsuperscript{13} In our case, the signal intensities where mixed and non-specific on both T1 and T2 weighted images.

Treatment of these tumors is entire tumor removal and resection of the attached falx, since they are well demarcated and there is little adherence to surrounding brain structures.\textsuperscript{14} We performed a complete resection of a large falxchordoma attached to the dura mater.

The long-term prognostic is good when complete resection of the falxchordoma is achieved, and no recurrence should be expected.\textsuperscript{15} Hardy et al.\textsuperscript{15} reported a patient with a survival period of 44 years after complete removal of a convexity chondroma. In our case we have done gross total resection of the tumor.
Conclusion

Tumors of falx cerebri origin with unusual radiological features may be considered as intracranial chondroma in the differential diagnosis.

References