A RARE CASE REPORT INTRACRANIAL CHONDROMA

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Abstract:
Intracranial chondromas are benign tumors that constitute around 0.2 - 0.3 of all intracranial neoplasms. It commonly arises from skull base. Intracranial chondromas arising from duramater is rare. Hence this case has been presented. Radiologically it closely resembles meningioma. Cerebral angiography and histopathology are useful to differentiate them. It has good prognosis when resected completely.

Keyword: Chondroma, benign chondrocytes, S100, Vimentin

INTRODUCTION:
Chondromas are benign tumors that can be seen in any part of the body. Intracranial chondromas constitute around 0.2% - 0.3% of all intracranial neoplasms¹. Among intracranial neoplasms, tumors of cartilaginous origin are rare. Intracranial chondromas mostly involve base of skull with a predilection for sphenoidal region. It less commonly arises from the duramater, choroid plexus, leptomeninges, or brain parenchyma². Intracranial chondroma that arises from duramater constitutes around 15%³.

CASE REPORT
25 year old male admitted with complaint of recurrent episodes of seizures and vomiting of 3 months duration. He gave history of trauma ten years back. Family history and general examination were unremarkable. Neurological examination was normal. Routine laboratory investigations were within normal limits. Electroencephalogram showed focal slow waves in temperoparietal areas.

RADIODIAGNOSIS
X RAY SKULL Skull roentgenogram showed hyperostosis in the left temperoparietal region.

CT AND MRI BRAIN
CT and MRI imaging showed a large well defined lobulated dural based extra axial mass lesion with mixed signal intensity. Heterogenous enhancement and dense calcification was seen along left cerebral convexity causing mass effect over underlying brain and midline shift, probably meningioma (Fig1, 2).
The patient underwent craniotomy. When the bone flap was elevated from left parasagittal area a hard tumor was found attached to the duramater in the left temperoparietal region. The dural incision revealed a well defined, 8x6 cm, firm to hard, whitish-gray hypovascular tumor. Calcification was noted at the surface of the tumor. There was no adhesion between the tumor and the brain (Fig3). The tumor was completely resected along with the attached duramater. Postoperative MRI studies showed complete resection.

**Fig 3: Intra operative picture showing easy seperation of the tumor from the underlying brain parenchyma.**

**GROSS EXAMINATION:**

**Fig 1:** Axial non contrast CT image showing densely calcified mass lesion in left frontal region

**Fig 2:** MRI Axial T2 weighted image showing heterogenous extraaxial mass lesion with hypotense areas

**SURGERY – INTRAOPERATIVE FINDINGS**
Gross examination of the specimen revealed multiple cartilaginous bits in aggregates measuring 8x6x3cm. The tumor is pearl white in colour, firm to hard in consistency and covered with a thin transparent capsule (Fig4). Fig 4: Gross picture showing multiple pearly white cartilaginous bits.

MICROSCOPIC FINDING

Histopathological examination revealed a well-encapsulated cellular mass arranged in lobules composed of well-differentiated chondrocytes in the lacunae. Nuclear pleomorphism, increased mitotic activity or multinucleation were not seen. No meningothelial elements were noted. No hemorrhage or necrosis noted. There was no invasion into the brain parenchyma. The diagnosis of intracranial chondroma was made (Fig 5, 6, 7).

Fig 5: Benign chondrocytes in a chondroid matrix (4x)
Fig 6: Benign chondrocytes in a chondroid matrix (10x)
Fig 7: Benign Chondrocytes (40x)

IMMUNOHISTOCHEMISTRY

Immunohistochemistry done in this case revealed strong S100 and Vimentin positivity. EMA and Cytokeratin were negative (Fig 8, 9, 10, 11).
DISCUSSION

Hirschfield (1851) was probably the first to report intracranial cartilaginous tumor specifically chondroma\(^3\). The first operative resection was published by Nixon in 1982. Intracranial chondromas are rare and the majority of patients are between 20 and 60 years of age, with a peak incidence in the third decade. There is no gender predominance, although a slight female preference has been proposed\(^3\). It can be solitary or multiple. Solitary intracranial chondromas are usually located at the base of skull or in the paranasal sinuses with extension into the cranial cavity. Usually this tumor presents as a solitary mass, and rarely, it is associated with Ollier's multiple enchondromatosis, Maffucci's syndrome or choroid plexus papilloma\(^3\). WHO has grouped it under mesenchymal tumors as ICD 9220\(^5\). Intracranial chondromas can be classified into four groups\(^3\).

I GROUP - Tumors which arise from the base of the skull, particularly in the sphenoid region.
II GROUP - Tumors originating from the paranasal sinuses and extending into the cranial cavity.

III GROUP - Tumors arising from the choroid plexus

IV GROUP - Tumors with a dural attachment.

HISTOGENESIS:
Intracranial chondromas mainly develop from cartilaginous rests along the basilar synchondroses. However, the origin of chondromas arising from other locations is unclear and they are generally considered to be due to heterotopic embryonal cartilaginous rests, metaplasia of meningeal fibroblasts, metaplasia of perivascular mesenchymal cells, or displacement and migration caused by trauma or an inflammatory process.

Intracranial chondromas grow predominantly by expansion and do not invade the brain parenchyma. Therefore the clinical manifestations of these tumors are usually mild and nonspecific, although some patients may present with focal signs due to local compression of adjacent brain tissues, increased intracranial pressure, or seizures. In this case, patient presented with symptoms of headache and seizures.

Bone destruction occurs in over 50% of cases, and the tumor may also produce hyperostosis of inner table of skull. Chondromas are avascular lesions that compress the brain but do not invade it. In addition to the clinical manifestations, imaging studies such as CT or MRI are important for the detection of intracranial chondromas. Both CT and MRI of chondromas usually show a well-circumscribed, extra axial lesion, with variable heterogeneous and nonspecific patterns and the tumors usually have delayed slight-to-moderate enhancement. The heterogeneous features probably reflect the varying extent of calcification.

As a whole, chondroma should be considered if imaging studies reveal a ring-shaped circumscribed extra axial tumor with a thick rim and delayed slight-to-moderate enhancement. In this case, radiological imaging revealed a well-defined lobulated extra axial mass lesion with mixed signal intensity. Heterogenous enhancement and calcification were also noted in this patient.

In cerebral angiography chondromas present as avascular extra cerebral, space-occupying lesions which cause compression of the arteries in the vicinity of the tumors. Cerebral angiography was not done in this case.

Microscopically, the tumor consists of a fine fibrous capsule surrounding lobules of well-differentiated chondrocytes. The chondrocytes are seen in an abundant chondroid matrix, show no evidence of atypical cells, multinucleation, or mitotic activity. This case revealed similar histological findings.

IMMUNOHISTOCHEMISTRY:
In chondroma S100 and vimentin will be strongly positive. Cytokeratin and EMA will be negative. But in Meningioma vimentin and EMA will be strongly positive. Cytokeratin and S100 will be focally positive.

Immunohistochemistry done in this case revealed strong S100 and Vimentin positivity. EMA and Cytokeratin were negative.

DIFFERENTIAL DIAGNOSIS:
1. Metaplastic meningioma with extensive cartilaginous differentiation
2. Chordoid meningioma
3. Chondrosarcoma
The current WHO classification considers meningeal tumour with pure cartilaginous mesenchymal differentiation as a metaplastic subtype. Various types of metaplasia including xanthomatous, osseous, lipomatous or cartilaginous differentiation might occur in meningiomas. However, the extensive cartilaginous metaplastic changes are extremely rare. Chondromas of the duramater and falx are difficult to distinguish from Metaplastic meningiomas with extensive cartilaginous differentiation. Meningiomas usually display intense, homogeneous contrast enhancement whereas chondromas are heterogeneous and shows calcification. Early contrast enhancement is suggestive of meningioma, but marked delayed contrast enhancement is suggestive of chondroma. In cerebral angiography meningioma is vascular while chondroma is avascular. Metaplastic meningioma can be differentiated histologically by the presence of foci of cells with meningothelial features. In the present case, even with extensive sampling meningothelial cells could not be made out histologically and it was avascular thus favouring chondroma ruling out metaplastic meningioma with extensive chondroid differentiation.

Chordoid meningiomas are usually supratentorial masses. The tumor is often vaguely lobulated with abundant basophilic mucin and epithelioid to spindled cells arranged in ribbons or cords. Focally foamy or vacuolated "physaliferous-like" cells can be seen. A prominent lymphoplasmacytic infiltrate is occasionally seen. Foci of conventional meningothelial or transitional meningioma may be seen. Psammoma bodies are also uncommon. Chordoid meningiomas are especially likely to recur, particularly following subtotal resection hence graded as WHO grade II meningioma. Chordoid meningiomas with extensive cartilaginous metaplasia are rare. In this case chordoid like areas and meningothelial cells were not seen thereby ruling out Chordoid meningioma.

Microscopically, chondrosarcoma can be differentiated by the presence of nuclear pleomorphism and mitotic activity, but sometimes very well differentiated cartilaginous areas can be found within chondrosarcoma. This emphasizes the need for careful pathological evaluation of the specimen before labeling it as benign chondroma. Low grade chondrosarcoma differs from chondroma by invasion. Invasion and pleomorphism were not seen in this case ruling out well differentiated chondrosarcoma.

**PROGNOSIS:**
Since dural chondromas are well demarcated and have minimal adherence to surrounding tissues, the treatment of choice for these tumors is total tumor excision with removal of attached durameter. The long-term prognosis is good after total excision of the tumor and tumor recurrence is exceptional. Malignant transformation into chondrosarcoma has been reported after partial resection of chondroma. Therefore, in cases with subtotal resection of chondroma, long-term imaging follow up may be necessary, and local invasion or recurrence may suggest malignant transformation into chondrosarcoma. However chondromas associated with Maffucci syndrome has showed increased incidence of chondrosarcomatous change around 16%.

**CONCLUSION**
Intracranial chondromas are circumscribed masses of well differentiated, cytologically benign hyaline cartilage. They tend not to invade or destroy the surrounding parenchyma and there is little tendency for sarcomatous change. Total excision is frequently
possible and is curative, with no recurrence reported on long-term follow-up.

BIBLIOGRAPHY


