Intracranial chondroma

see Skull Base Enchondroma.

Intracranial chondroma are cysts of chondroid tissue, first reported by Hirschfeld in 1851.

**Epidemiology**

They are extremely rare and account for only 0.2% to 0.3% of all intracranial tumors.

They can be present at different regions within the cranial cavity especially the skull base, intrasellar, parasellar, intradural and especially falx.

Very occasionally observed in combination with intratumoral hemorrhage.

Despite a purported lack of any sex predilection there are reports of a slight female predominance.

Intracerebral location is extremely rare and has only been described in a few cases.

**Etiology**

Various theories have been proposed to determine the etiology of intracranial chondromas but none has succeeded to ascertain a definite cause of origin. The most commonly accepted explanation for skull base chondromas is embryonic remnants of chondrogenic cells along the base.

They grow slowly by expansion and mostly originate from rests of cartilaginous cells at sphenoethmoidal suture and sphenoorbital suture.

The chondromas arising from the dura matter, choroid plexus, and cerebral cortex have been proposed to develop from metaplasia of meningeal fibroblasts and perivascular meninges.

Similarly, proliferation of ectopic embryologic rests of cartilage cells, traumatic displacement of cartilage elements or inflammatory cartilaginous activation of fibroblasts have been suggested to be the cause of development of intracranial chondromas.

**Clinical features**

The presenting symptoms range from headaches to lower cranial nerve palsy. In some cases, proptosis, diplopia and varying degrees of visual activity impairment along with orbital extension have been reported. Patients often complain of forgetfulness and lack of concentration.

Generalized tonic–clonic seizures are also usually the presenting complaints in intracranial chondromas, which develop because of the gradual destruction of a large number of neurons that begin to fire at regular intervals. Focal neurological deficits may also result from mass effects of tumor.

Intracranial chondroma has also been reported as a component of Ollier’s multiple chondromatosis.

Pontine hemorrhage has also been associated with parasellar intracranial chondromas. Association of skull base chondromas has also been reported with Maffucci syndrome.

Intracranial chondromas may develop in a person at any age but they have been most frequently
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observed in the third decade.

**Diagnosis**

Bone destruction occurs in over 50% of the cases, whereas irregular calcifications are seen in about 60%. Intracranial chondromas may also produce hyperostosis of the inner table of the skull [17] [18] [19].

On X-ray, intracranial chondromas represent hyperostosis of the internal table of the skull [20].

Enhanced intracranial pressure and calcified portions [21]. Intradural convexity chondromas possess carved, tufted, ring-shaped calcified areas [22].

MRI has become an important diagnostic tool for intracranial chondromas. Brownlee et al. reported variable signal intensity at different levels of MRI in a case of intracranial chondroma. At T1 they reported less intensity whereas at T2 the signal appeared to be of middle to high intensity [23].

They are typically DWI hypointense with high apparent diffusion coefficient (ADC) values while meningiomas are typically DWI hyperintense with low ADC values [24].

A study reported that intradural chondromas possess two different CT appearances. The usually found type 1 shows mixed density with minimal or moderate enhancements. The rare type 2 shows an innermost less dense area containing a cyst [25].

Angiography shows displacement of vessels but no tumor stain [26] [27] [28].

Chondromas showed low uptake in PET images, which might be useful for differentiation between chondromas and chordomas [29].

In the past pneumoencephalography revealed displacement of basal cisterns and the ventricular system. A radionuclide brain scan may show abnormal uptake in the tumor [30].

**Differential diagnosis**

Preoperatively, chondromas can be difficult to distinguish from meningiomas. They may also be confused with chordomas, craniopharyngiomas or even arterial aneurysms [31] [32].

Tanohata et al. reported two instances of skull base chondromas that exhibited delayed contrast enhancement on CT after a high-dose of the contrast medium was administered. They suggested this CT feature to be employed in differential diagnosis of intracranial chondromas from meningiomas and neurinomas [33].
Treatment

In symptomatic patients, operative resection is sensible. In most cases total removal of the tumor is possible and leads to full recovery. When the finding is merely incidental in older patients, a watchful waiting approach is acceptable, given the benign and slow-growing nature of the lesion \(^{34}\).

The current popular surgical approach for parasellar lesions is transcranial such as the orbitozygomatic approach, subtemporal approach. In surgical removal of skull base chondromas, it is advisable to try to confirm the diagnosis preoperatively with characteristic image findings and to consider the best approach in each case to decompress the involved nerves without any complications \(^{35}\).

In cases of convexity chondroma, it is additionally recommended to remove the dural attachment \(^{36}\) \(^{37}\) \(^{38}\).

Outcome

Usually postoperative observation reveals no recurrence of the lesion after complete resection. An adjuvant therapy is not necessary and the long-term prognosis is good \(^{39}\) \(^{40}\) \(^{41}\).

The malignant form, chondrosarcoma, generally occurs later in life, presenting mostly in the fifth and sixth decades \(^{42}\).

Case series

2011

Xin et al. retrospectively analyzed the clinical data of 30 patients (12 males and 18 females; mean age 35.4 years; age range 16-60 years) who had pathologically confirmed intracranial chondroma treated at our hospital from September 1996 to June 2008. Surgery was performed on all 30 patients: five patients underwent postoperative radiotherapy; 26 patients were followed up postoperatively for a mean duration of 45.8 months. The surgical approach was selected according to tumor location. Total resection was achieved in 11 patients, subtotal resection in 13, and partial resection in nine (three patients had recurrent chondroma). Follow-up showed that 21 patients recovered without recurrences, three had recurrence, and two patients died. The clinical manifestations included headache and multiple cranial nerve lesions. Imaging usually showed a well-demarcated extramedullary tumor, centrally located, without surrounding brain edema, partially calcified (73.3%) and with minimal vascularity, often accompanied by erosion and destruction of surrounding bone (56.7%). It is difficult to totally remove an intracranial chondroma, and it is not possible to differentiate a chondroma from a myxoma or chordoma at the cranial base on the basis of clinical manifestations and neuroradiological findings. Selection of the appropriate surgical approach is important for resection of the tumor \(^{43}\).

1976

Four new cases are added to the previously recorded 122 cases \(^{44}\).

Case reports
2018

A 25-year-old male patient with a giant convexity chondroma with meningeal attachment in the right frontal lobe that was detected after a first generalized seizure. Based on the putative diagnosis of meningioma, the tumor was completely resected via an osteoplastic parasagittal craniotomy. The postoperative MRI confirmed the complete tumor resection. Histopathological analysis revealed the presence of a chondroma.

2017

Giant convexity chondroma with dural involvement: Case report and review of literature.

2013

A 55-year-old female presented to the emergency room with a complaint of aphasia. Her initial brain computed tomography scan showed an intracranial hemorrhage in the left frontal area. After surgery, histopathological examination confirmed the diagnosis of a chondroma. Intradural chondroma is a rare, slow growing, benign intracranial neoplasm, but is even rarer in combination with an intratumoral hemorrhage. Chondromas are generally avascular cartilaginous lesions. This case was thought to be caused by the rupture of abnormally weak vessels derived from the friable tumor. Intradural chondromas may be included in the differential diagnosis of intracranial tumors with acute hemorrhages.

2012

A 23-year-old Asian man presenting with intracerebral chondroma of the left frontal lobe, which was eroding the dura matter. The intracranial chondroma was completely removed by surgery.

2011

A 45-year old female is presented with a solitary intracerebral chondroma located in the right frontal lobe with no meningeal attachment.

An intracranial chondroma with intratumoral and subarachnoidal hemorrhage.

2007

Higashida et al. reported two cases of intracranial skull base chondroma and discussed the differential diagnosis and the treatment strategies. The first case was a 39-year-old male who presented with left exophthalmos, visual loss and oculomotor disturbance. MRI showed a huge tumor occupying the bilateral cavernous sinus. Partial removal of the tumor was performed through the left orbitozygomatic subtemporal approach. The second case was a 54-year-old male who presented with left hemiparesis. MRI showed a brain stem infarction with a huge tumor located at the right middle fossa. Partial removal was performed through the right orbitozygomatic subtemporal approach. In these two cases, the histopathological diagnosis of the tumors was benign chondroma and the size of residual tumors have not changed for one year without any additional therapy.
Intracranial chondroma

A Osteochondroma of the skull base.

2003
A rare case of a chondroma arising from the convexity dura mater.

2001
A case of intracranial giant chondroma originating from the dura mater of the convexity.

2000
Intradural convexity chondroma: a case report and review of diagnostic features.

1993
A rare case of chondroma originated from the dura mater of the cerebral convexity in a 16-year-old girl. Radiologic findings are reported with emphasis on computed tomography and magnetic resonance imaging scans, and histogenesis is briefly discussed.

1991
A rare case of Maffucci's syndrome associated with enchondroma at the skull base, left internal carotid artery aneurysm, and goiter is reported. Two other previously reported cases of Maffucci's syndrome with associated aneurysms and the present case suggest that Maffucci's syndrome may be associated with aneurysm.

1990
A rare case of parasellar chondroma accompanied by pontine hemorrhage is described. A review is made of the previously reported 6 cases of intracranial chondromas complicated with hemorrhage. A 21 year-old woman was admitted because of consciousness deterioration progressing to coma within a day, and right hemiparesis. CT scan showed a contrast-enhanced mass in the parasellar region and a hematoma in the brain-stem, which was clearly demonstrated by MRI to be abutted on the dorsal part of the tumor mass. The tumor was removed through frontotemporal craniotomy and confirmed histologically as chondroma. Postoperatively, the patient gradually regained consciousness and is hospitalized to rehabilitate hemiparesis.

1989
A case is presented in which a solitary chondroma arose from the clivus of a patient with Ollier's disease.

1983
Intradural chondroma: a case report and review of the literature.
1981

A case of a huge intracranial frontoparietal osteochondroma in a 20-year-old man is reported. The presenting symptoms were headache, vomiting, and blurred vision. Apart from papilledema, no other abnormal neurological signs were present. A specific preoperative diagnosis could not be reached from the information provided by plain skull films, angiography, and radionuclide scan. The findings on computed tomography were those of a high density mass interspersed with small foci of lower densities, producing a honeycomb appearance, and surrounded by deposits of nodular calcification. The postcontrast scan showed a moderate degree of enhancement with preservation of the precontrast honeycomb pattern. These particular features may enable a correct preoperative histological diagnosis to be offered with a high degree of probability.

1969

Osteochondroma of the base of the skull causing an isolated oculomotor nerve paralysis. Case report emphasizing microsurgical techniques.


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