

Intracranial Chondroma. Report of Two Cases and Review of the Literature

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ABSTRACT: *Introduction:* Chondromas are rare intracranial tumors. The authors present two cases of intracranial intradural chondroma, one originating from the falx cerebri and the other from the dura mater of the convexity. *Method and Results:* Diagnostic procedures, including magnetic resonance imaging, and surgical findings are described. In both cases, pre-operative diagnosis could have been at least suspected, and the tumor was completely removed, without recurrence after a follow-up of many years. The pathogenesis and pathological findings are discussed, and cases from the literature are reviewed. *Conclusion:* Benign intradural chondroma has a good prognosis, with no recurrence after surgical excision in most cases.

RÉSUMÉ: *Présentation de deux cas de chondrome intracrânien et revue de la littérature.* *Introduction:* Les auteurs présentent 2 cas exceptionnels de chondrome intracrânien intradural, une tumeur intracrânienne particulièrement rare. Un cas originait de la faux du cerveau et l'autre de la dure-mère de la convexité. *Méthode et Résultats:* Les données d'imagerie, incluant la résonance magnétique nucléaire, de même que les données chirurgicales sont présentées. Dans les deux cas, le diagnostic pré-opératoire pouvait être fortement suspecté, et une excision complète de la lésion a été pratiquée, sans signe de récurrence après une période de suivi de nombreuses années. La pathogénèse et la pathologie sont discutées, et les auteurs font une revue brève des cas de la littérature. *Conclusion:* Le chondrome intradural bénin est une tumeur à bon pronostic avec un risque très faible de récurrence après excision chirurgicale complète.

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Chondromas are rare intracranial tumors, representing 0.2-0.3% of all primary intracranial tumors.^{1,2} They usually arise from the synchondrosis at the base of the skull.^{1,3-6} Less frequently they arise from the dura (convexity or falx),⁷⁻²⁰ from inside the ventricles²¹⁻²⁵ or the cerebral parenchyma without apparent dural attachment.²⁶⁻²⁹ Their suspected development from embryonic remnants of the primordial cartilage in the synchondrosis at the base of the skull may explain their predilection for this location. However, the histogenesis of supratentorial tumors originating from the falx, the dura mater or brain parenchyma is not well understood. In the present report two dural chondromas are described. Their diagnostic criteria are discussed. An attempt is made to correlate the pathological and radiological features. Finally, the authors compare their findings with those reported in the literature.

CASE REPORT

Patient 1

This patient was a 22-year-old man with a 1 month history of focal motor seizures, with convulsions involving the left arm and leg. He had no headache or other symptoms of increased intracranial pressure. The clinical examination on admission revealed normal physical and neurological findings. Plain skull films demonstrated a ring-like calcification in the right frontoparietal area, without any sign of increased intracranial pressure (Figure 1). A computed tomographic scan showed a large hyperdense intracranial space-occupying lesion, located in the right

fronto-parietal area near the midline, with a calcified implantation base on the falx (Figure 1). There was no oedema, and neither significant mass effect nor enhancement. A right external and internal carotid angiogram revealed an avascular lesion with displacement of normal blood vessels.

A craniotomy was performed, and the opening of the dura-mater disclosed an intradural extracerebral tumor. The tumor was completely removed including its attachment to the falx. The tumor was easily cleaved from the brain surface. The post-operative course was eventless. The last CT scan was done in 1990, 5 years after the operation, and showed no evidence of recurrence. Pathologic examination revealed a 5.5 x 4.4 x 3.8 cm firm mass with a slightly lobulated surface. Section showed a lobulated greyish-white semilucid surface. Calcifications were rather extensive mostly close to the basal membranous surface. Histologically the lesion was made of nodules of perfectly mature cartilaginous tissue with thin sparsely cellular conjunctive and vascular strands (Figure 1). Calcifications developed within the cartilaginous matrix, as normally happens in chondromas. Bone formation was also focally extensive, with areas of hematopoietic activity.

Patient 2

This 32-year-old woman was brought in for evaluation of headache and slight weakness and fine tremor of the left arm. The neurological

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examination revealed slight weakness on the left side. The skull films showed a 2 x 1 cm area of hyperostosis of the inner table on the right frontal area (Figure 2). The CT scan showed a very large and well circumscribed lesion in the right frontal area, with an hypodense central part and a peripheral hyperdense enhancing part (Figure 2). A pre-operative angiogram revealed vascular displacements without tumoral blush, in agreement with an extra-axial avascular lesion. An MRI was done, showing a large lesion with well-defined margins and a right posterior frontal dural implantation. The T1-weighted sequence showed a markedly hypointense central part and a slightly hypointense peripheral part (Figure 2). The proton time and T2-weighted sequences showed a hyperintense signal in the central part and a nearly isointense heterogeneous signal in the peripheral part, with a thin hypointense margin and no oedema (Figure 2).

A right frontal craniotomy was performed, and the tumor was totally removed. It originated from the dura-mater of the convexity, without evidence of brain invasion. The dural attachment was easily cleaved from the skull, without any evidence of dural transgression or bone invasion. The last CT scan was done 4 years later, in 1991, with no sign of recurrence. Grossly, the lesion measured close to 8 x 6 x 6 cm and weighed 124 grams. The dural attachment measured 6 x 5.3 cm. The surface was discretely lobulated. The cut surface was the same as that of Patient 1, except for a slightly eccentric deep, soft, depressed, sharply limited but irregularly shaped zone of yellowish-white tissue (Figure 3). The main histologic picture was that of mature hyaline cartilaginous tissue. In rather extensive areas, lacunae were not so well developed and cellularity was slightly denser, but these, in absence of nuclear atypia, larger nuclei, binucleated cells and mitosis, were not considered as indicative of even borderline malignancy. The soft sub-central area described grossly was not made of necrotic tissue but of a very loose-textured connective vascular tissue, identical with that normally present in chondromas,³⁰ separating the cartilaginous matrix lobules (Figure 3). This tissue was made of widely separated fibroblasts and small bloodvessels. The wide intercellular spaces were most often unstained; in some areas they were occupied by a slightly acidophilic homogeneous material with no polysaccharidic staining. This connective tissue thus appeared as mainly edematous. Liquid loss from the cut surface explained its tendency to shrink and collapse, then appearing as nearly cystic.

DISCUSSION

Intracranial chondromas are rare lesions. Mapstone et al.¹³ in 1983 reviewed the literature and found 120 cases of intracranial chondromas, of which 29 originated outside the cranial base (dura, falx, cerebral parenchyma or choroid plexus). We found 18 additional cases in our analysis of the world literature, increasing the total number of published cases to 47, including our 2 cases. About 70% of the cases originated from the dura mater, the remaining cases (30%) were located inside the brain parenchyma or the choroid plexus (Table 1). The tumors at the convexity were located more often in the fronto-parietal area (Table 1).

The histogenesis of intracranial chondromas and chondrosarcomas has been debated. Those originating from the basal synchondrosis are believed to arise from embryonic remnants of chondrogenic cells in such areas.^{1,4,6,31} The origin of dural chondromas and chondrosarcomas is, at first glance, more speculative. Hypotheses include development from heterotopic chondrocytes,^{5,32} metaplasia of meningeal fibroblasts^{1,7,31} or perivascular mesenchymal tissue²⁶ and origin from meningeal connective tissue as a result of cartilaginous activation of fibroblasts by trauma of inflammation.²⁷

Russell & Rubinstein⁵ and Sheithauer & Rubinstein,³³ favour an origin from pluripotential mesenchymal cells or from their more differentiated descendants abnormally present in meningeal tissue. Nevertheless, the recent observation by Katayama et al.³⁴ of a meningeal chondrosarcomatous tumor

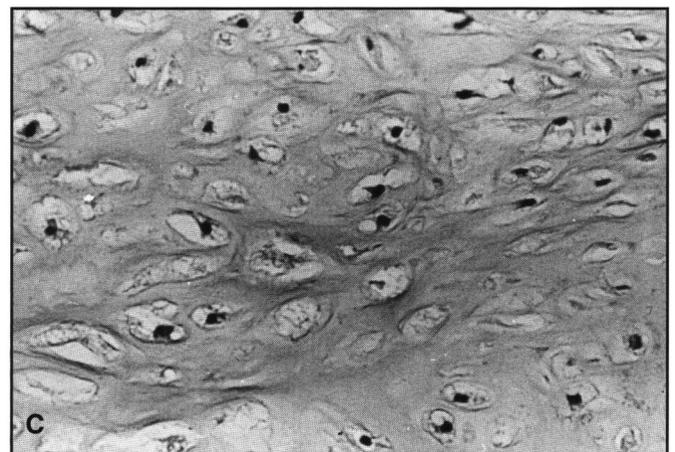
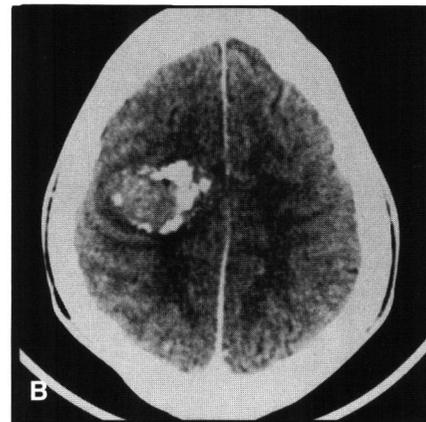
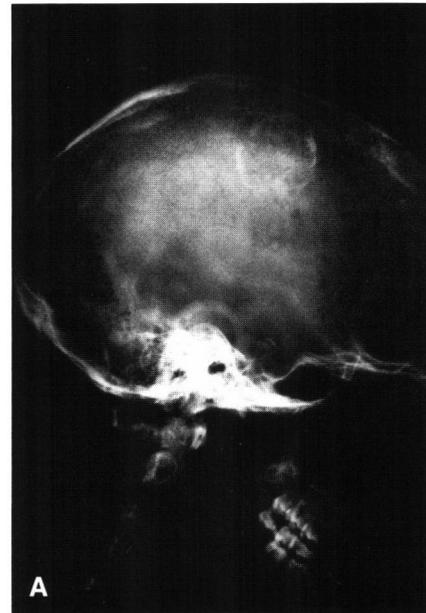


Figure 1: Patient 1. **A)** Lateral skull radiograph showing a ring-like calcification in the right frontoparietal area. **B)** CT scan with contrast enhancement showing the lesion with a calcified implantation base on the falx, and little enhancement. **C)** Representative photomicrograph, microscopic appearance of tumor from Patient 1, with well differentiated chondrocytes. H&E x 160.

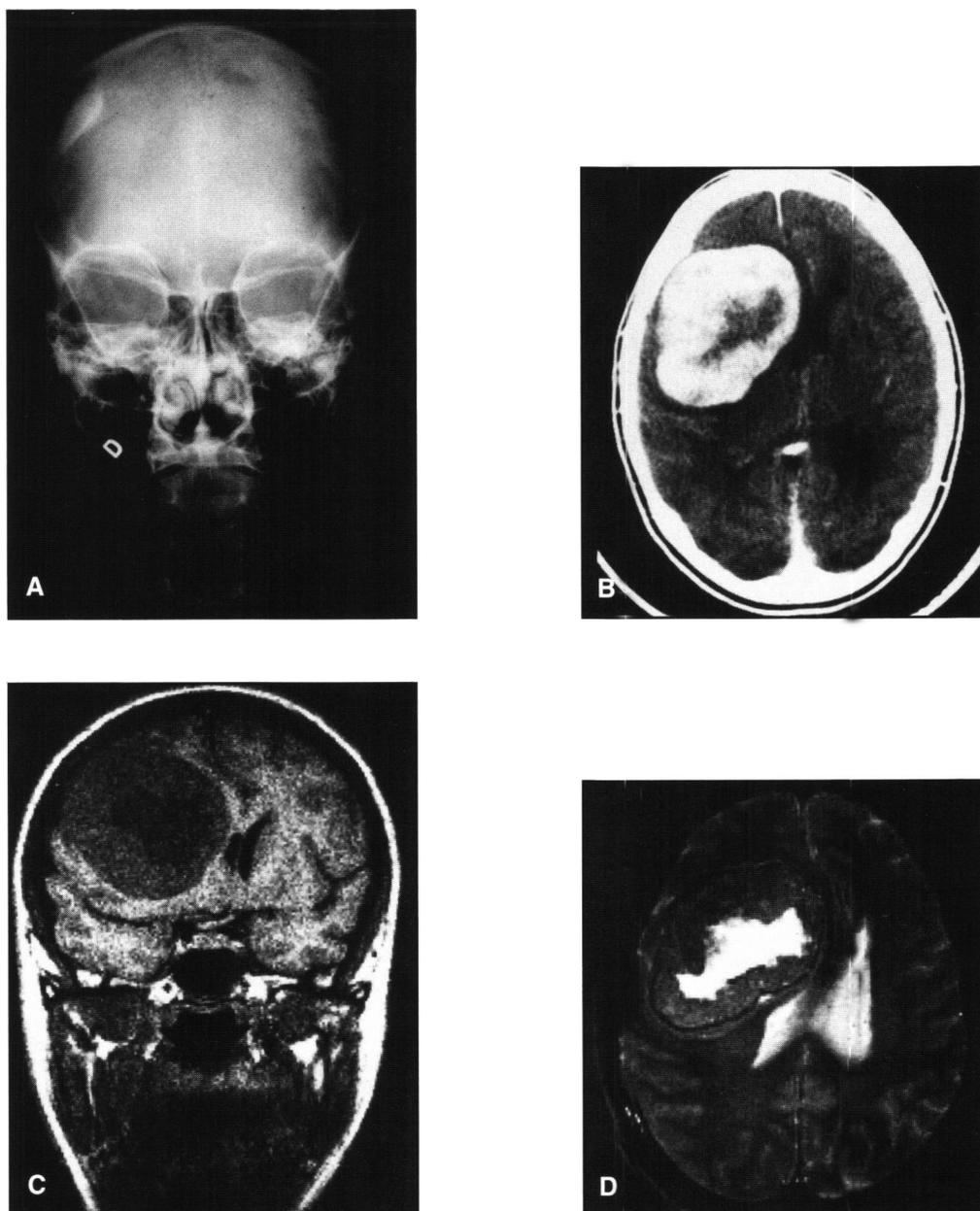


Figure 2: Patient 2. A) AP skull radiograph showing hyperostosis of the inner table of the skull. B) CT scan with contrast enhancement showing the large lesion with hypodense central part and peripheral enhancing part. Coronal T1-weighted magnetic resonance image C) showing the very hypointense central part and slightly hypointense peripheral part. On axial T2-weighted sequence D) the central part became highly hyperintense and the peripheral part isointense. Gadolinium was not used (not available in our center in 1987).

associated with meningocytic differentiation has revived the hypothesis proposed by Cushing & Eisenhardt (cited by the authors),³⁴ according to which at least an occasional meningeal cartilaginous tumor might originate from meningocytes through a form of metaplastic differentiation. Katayama et al. have found a few similar reported cases. The same process probably explains Russell & Rubinstein's⁵ observations of transitional aspects between meningioma and haemangiopericytoma. By analogy, such a metaplastic phenomenon can explain the occurrence of cartilaginous differentiation in glial tumors.³⁵

The clinical context of chondroma is different from that of a much more frequent meningeal tumor, the meningioma, which

most commonly occurs in the fourth through sixth decades of life with a peak incidence at 45 years,³⁶ and which is twofold more frequent in women than in men.³⁶ For intracranial chondroma, we found in our review a mean age of 30 years at presentation, and a slight male predominance (60% versus 40%) (Table 2). This seems to imply a different biological context and pathogenesis. Clinically, patients often present with a long standing history of headache progressing toward a more obvious picture of increased intracranial pressure, seizures (more often focal motor seizures), or a progressive lateralized deficit which varies according to tumor location. This is not surprising if we consider the slow-growing nature of these tumors, their very

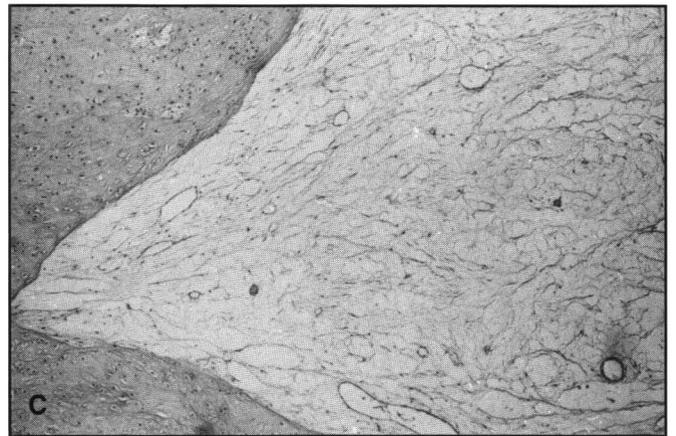
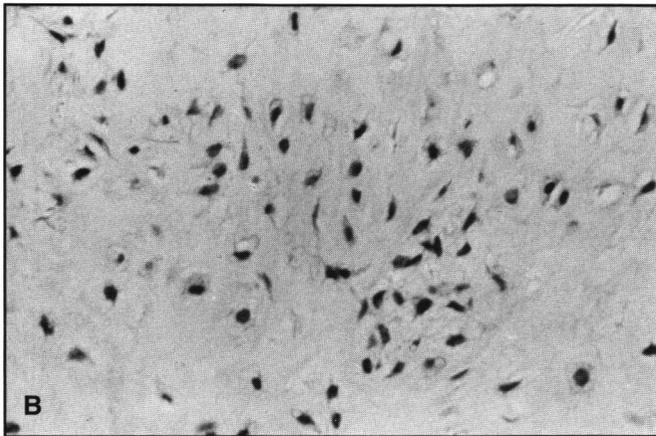
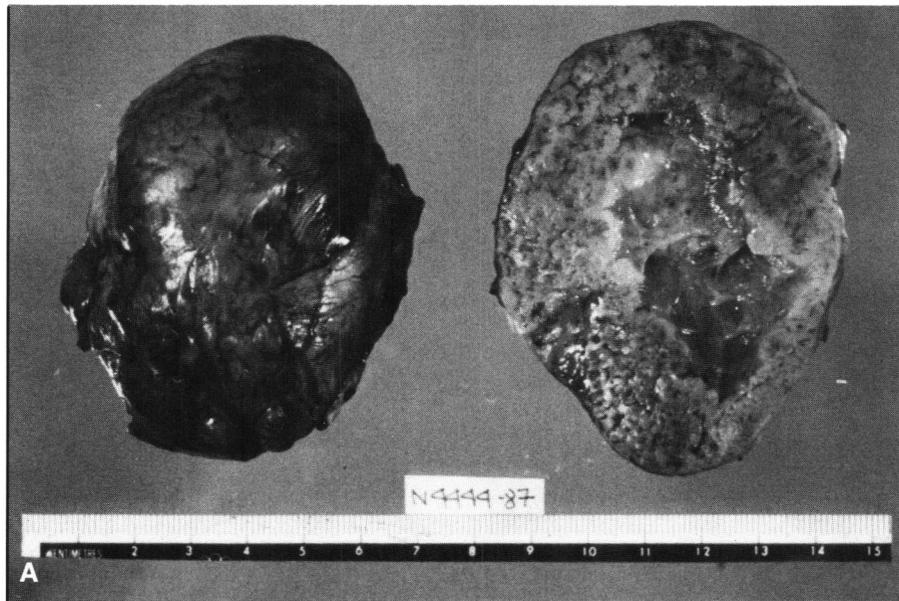


Figure 3: Patient 2. **A)** Macroscopic appearance of the tumor. Note the central gelatinous part of the tumor. **B)** Mature hyaline cartilaginous tissue. Lacunae not well developed but no evidence of nuclear atypia or mitosis. H&E x 160. **C)** Clear demarcation line between the typical cartilaginous tissue and the central loose-textured connective tissue. H&E x 64.

large size at presentation (mean diameter 6 cm and mean weight 170 grams) (see Table 2), and their tendency to be localized in the frontoparietal area (Table 1). Three patients died suddenly; the tumor was discovered at autopsy, and was responsible for brain herniation.^{21,22,28}

The radiological features can warrant, in some cases, a presumptive pre-operative diagnosis. The findings are summarized in Table 3. Calcifications and hyperostosis are frequent on plain skull films. In our analysis of the reported cases, we found two distinct CT-scan presentations (see Table 3). The first pattern or type 1 can be considered "classical".^{9,13,14,17,23,25} The second pattern or type 2, in which there is a hypodense central area, has been reported less frequently.^{11,15,20,37} A pathological correlation with this second pattern was made in one case and showed that the hypodense central part contained "central cystic degeneration".¹¹ Pathological data in our Patient 2 demonstrated that this central area was composed of a very loose-textured edematous connective tissue (without evidence of necrosis). Furthermore,

this well hydrated tissue explained the well demarcated hyperintense central area on the long TR MRI sequences.^{15,37} Four cases of benign intradural chondromas imaged with MRI have been reported (including our case), and the findings are summarized in Table 3. The signal either on CT scan or MR scan is often heterogeneous without the typical homogeneous and densely enhancing aspect of a meningioma. On an angiogram, these tumors present as avascular extracerebral space-occupying lesions.^{11,13,14,24-26,29,37} The latter is a good distinguishing feature from a meningioma, which is usually associated with a blush on the angiogram. The presence of hypervascularity in an otherwise typical lesion should raise the possibility of a chondrosarcoma.³⁸

Surgical resection is the treatment of choice for benign intradural chondromas. These tumors are well demarcated and have no tendency to invade or destroy surrounding parenchyma, and recurrence is rare after total removal. Our two patients remain asymptomatic and without radiological evidence of recurrence, with a follow-up of 6 and 7 years, respectively. The

Table 1: Reported locations of intracranial chondromas*.

1. Meninges: 34 (72%)
Convexity: 19
Frontal/Parietal: 17
others: 2
Falx: 15
Frontal/Parietal: 14
others: 1
2. Intracerebral: 7 (15%)
Frontal/Parietal: 3
Temporal: 2
Parieto-occipital: 2
3. Choroid Plexus/Intraventricular: 6 (13%)
Lat Ventricular: 4
IIIrd Ventricular: 1
IVth Ventricular: 1

* Total number of cases: 47.

long term prognosis is very good with prolonged survival following excision.^{12,16} On the other hand the malignant counterpart, the chondrosarcoma, has a variable clinical course with frequent recurrences and occasional distant osseous and visceral metastatic spread, particularly with the mesenchymal sub-type of chondrosarcoma.^{33,38-40}

Usually, benign cartilaginous tumors can be differentiated from the malignant counterpart on the basis of histopathological criteria.⁴¹⁻⁴⁴ A benign chondroma has an expansile and noninfiltrative pattern of growth and typically contains well differentiated cartilaginous cells within lacunae, with a low cellularity, and no mitoses.⁴² However, the degree of differentiation is situated along a continuum and the distinction between a benign chondroma and a low-grade chondrosarcoma is sometimes not obvious. The criteria to diagnose a low-grade (grade 1) chondrosarcoma are based mostly on the morphology of the

Table 2: General features intradural chondromas*.

Age at presentation	Mean: 30 years
	Range: 5-64 years
Sex	Female: 15 (40%)
	Male: 22 (60%)
Duration of symptoms to presentation	Range: 0.5 to 132 months
	Mean: 39 months
Tumor size (maximal diameter)	Mean: 6.0 cm
	Range: 1.5-10 cm
Tumor weight (grams)	Mean: 170
	Range: 67-250

* Sufficient clinical information was available in 37 published cases.

nuclei in the tumor chondrocytes, and include the presence of numerous multiple (2 or more) nuclei within one lacuna and the exclusive presence or marked preponderance of small and densely-staining nuclei.⁴¹ In some cases, a small number of larger and somewhat pleomorphic nuclei are present in isolated areas.⁴¹ Sanerkin emphasizes invasiveness as a reliable guide.⁴⁴ Dense cellularity, presence of significant numbers of larger nuclei and mitotic figures are not features of grade 1 chondrosarcoma (if present, indicate higher grade).⁴¹ Some diagnostic difficulties can arise because some cartilage neoplasms, both benign and malignant, contain myxoid areas in which characteristic chondrocytes are not apparent.⁴³

The higher grades (2 and 3) contain more striking features (mitotic figures, necrosis, marked anisokaryosis, multinucleate cells) and are easy to identify.^{41,44} However, any chondrosarcoma can contain areas of very well differentiated chondroid tissues,

Table 3: Radiological features

Modality	Findings	Comments
Skull X-Ray	Calcifications: Usually dense or mottled Rarely shell-like or curvilinear	Inner table hyperostosis reported in 8 Cases (13,17,19,42)
CT Scan	Huge calcified mass Type 1 (*) mixed density signal minimal to moderate enhancement Type 2 (II) hypodense well defined central area with hyperdense enhancing peripheral part	Calcifications; no reported case without calcification on CT scan No or minimal surrounding oedema
MRI	Mixed intensity signal Predominantly hypointense on T1 spin-echo and hyper-intense on T2 spin-echo Enhancement minimal to moderate except for more intense enhancement within peripheral part in type 2 (ring enhancement). (&)	Distinctive pattern: well defined central part with high intensity signal in T2 (Patient #2): corresponding to CT scan type 2
Angiography	Avascular extracerebral lesion	The presence of hypervascularity in an otherwise typical lesion should raise the possibility of a chondrosarcoma

(*); Based on references 9,13,14,17,23,25.

(II); Based on references 11,15,20,37

(&); Based on references 9,15,25

and this emphasizes the need for the extensive and meticulous examination of the pathological specimen, which is mandatory before a definite diagnosis of benign chondroma is made.⁴³ Benign intracranial intradural chondromas seem to have very little tendency for malignant transformation.⁸ Accordingly, a tumor first diagnosed as a "benign" chondroma but showing rapid recurrence, invasion or metastasis should be suspected of being a chondrosarcoma. In such a situation, it is very likely that a benign section of a chondrosarcoma was investigated initially.⁴²

The authors have presented two cases of intradural chondroma. These rare cartilaginous tumors have a particularly benign course, present usually at a younger age than meningioma, and without sex predominance. The pre-operative diagnosis can be made, on the basis of some radiologic features that are suggestive, despite the rarity of such a neoplasm. The treatment is surgical excision usually with good results and no recurrence. This benign tumor can be distinguished in most cases from its malignant counterpart, the chondrosarcoma. There is no reported case of malignant transformation of an intradural benign cartilaginous tumor.

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