

Giant dural supratentorial chondroma generating the question of how large can a tumor become without revealing itself

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Abstract

Chondromas usually affect the small bones of hand and feet and account for only 0.5% of all intracranial tumors. We present a case of a giant, supratentorial meningeal chondroma in

a 19-year old male patient and discuss the pre-operative diagnostic findings as well as the appropriate treatment options. A 19-year old male presented with headache, new onset of focal seizures and paresis of left upper extremity. Magnetic resonance imaging revealed a large right parietal tumor in the precentral region with local mass effect. The patient underwent right parietal craniotomy and gross total resection of the tumor. The histopathological report revealed a chondroma. Intradural supratentorial chondromas are extremely rare. As with other slow growing intracranial masses, they often reach a relatively large size before generating symptoms. Maximal surgical resection is the treatment of choice and if this is achieved no adjuvant therapy is necessary.

Introduction

Intracranial chondromas account for only 0,5% of all primary intracranial tumors. They usually arise in the skull base and especially in the sphenopetrosal, sphenoclivar or petroclivular junctions.¹⁻³ They tend to engulf cranial nerves or major arteries and become sympto-

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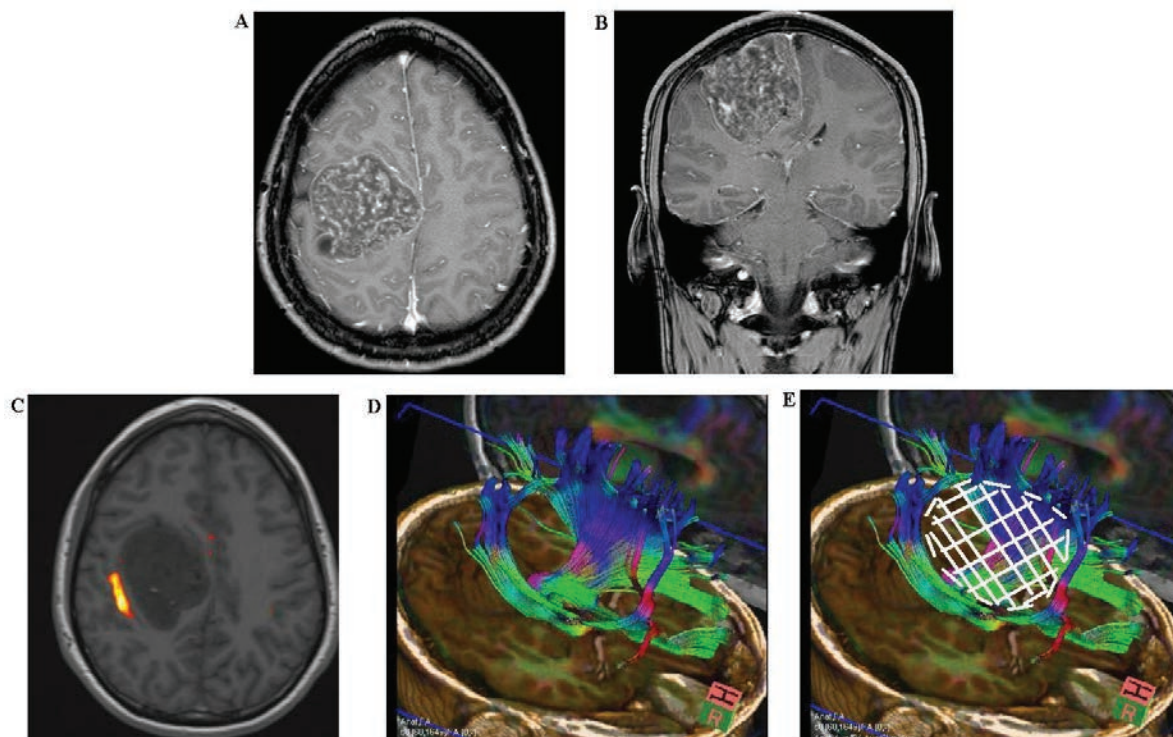


Figure 1. A) Axial T2WI of the brain showing a tumor with a honeycomb appearance; B) Coronal section of the tumor (T2WI); C) Functional magnetic resonance imaging illustrating the motor area of the left hand. The tumor seems to grow in the precentral gyrus pushing fibers of the motor cortex aside; D) Fibertracking image shows the corticospinal fibers pushed anteriorly and posteriorly from the tumor; E) The white area illustrates the tumor.

matic with cranial nerve palsies or headaches. They grow slowly and are considered benign as opposed to their malignant relatives the chondrosarcomas. The histological differentiation between the two can be challenging and has enormous significance regarding their therapy.

Case Report

A 19-year old male was admitted with complains of headache, hyperventilation and new onset of focal seizures. There was no significant family history, especially no history of tumor. The patient received no medication and had no history of radiation therapy. On clinical examination higher mental functions were intact. The patient was in postictal recovery and presented a paresis of the left upper extremity.

Magnetic resonance imaging (MRI) scan revealed a giant mass in the right precentral gyrus measuring 5×6×6 cm (Figure 1A and B). The corticospinal fibers as well as the motoric areas in the precentral gyrus were displaced anteriorly and posteriorly around the tumor (Figure 1C-E) On MRI imaging the tumor mass showed intermediate signal intensity on

T1 weighted and high signal intensity on T2 weighted images as well as heterogeneous enhancement with typical honeycomb appearance. The patient underwent surgical treatment via a right parietal craniotomy. The dura overlying the tumor mass was irregularly distended. The partially involved dura and the tumor were dissected from the cortical surface without difficulty. The intracranial mass was completely removed in a piecemeal fashion. Since the bone did not seem to be involved in the tumor process we drilled the inner table of the calvaria but did not remove the bone completely. The removed tumor mass was macroscopically multinodular, ivory colored with a very cartilaginous consistency (Figure 2). The patient had postoperatively no motor or sensory deficits. The histological diagnosis revealed a chondroma (Figure 3). The postoperative computed tomography (CT) brain scan confirmed the total excision of the tumor (Figure 4A). The patient was placed on appropriate antiepileptic drugs and made a good recovery. He was discharged on the 12th postoperative day. Following gross total resection and after establishment of the histological diagnosis no adjuvant therapy was needed. The whole body scintigraphy did not show further tumors (Figure 4B and C).

Discussion

Chondromas are recognized as a dyschondroplasia caused by developmental errors in enchondral ossification and are slow-growing, benign tumors of cartilaginous origin.^{1,3} They originate in the skull base and especially arise from the sphenopetrosal, sphenoclivar or petroclivular junctions. Rarely, they can be seen in the parasellar region, at the falx or arising from the dura convexity. Multiple theories have been suggested to explain the genesis of these tumors. In their paper Colpan and colleagues listed four of them:⁴ i) metaplasia of meningeal fibroblasts; ii) multipotential or perivascular mesenchymal cells in the dura mater; iii) aberrant nests of cartilage forming cells in the dura mater; iv) traumatic displacement of cartilage.

They can be found as solitary lesions or as part of Ollier's disease (multiple polysystemic enchondromatosis) or Maffucci's syndrome^{5,6} (multiple enchondromatosis associated with soft tissue angiomas). The publications regarding convexity chondromas are shown in Table 1.^{2,7-28} A given predominance of the left hemisphere can be seen. The frontoparietal region is the most common localization of the tumor. The oldest patient that has been

Table 1. Literature review with patients with convexity chondromas.

Publication	Age/Gender	Localization	Histologic type	Tumor dimensions
Berkmen and Blatt ⁸ (1968)	33 m	Right frontoparietal	N/A	550 g
Wu and Lapi ⁹ (1970)	32 f	Left parietal	N/A	8.5×6.5×4 cm
Hardy <i>et al.</i> ¹⁰ (1978)	22 f	Left frontoparietal	N/A	250 g
Matz <i>et al.</i> ¹¹ (1981)	20 m	Left frontoparietal	I	7×8 cm
Sebbag <i>et al.</i> ¹² (1990)	25 m	Left frontal	N/A	4×7 cm
Nakazawa <i>et al.</i> ¹³ (1993)	16 f	Left parietal	II	5×4 cm
Salazar-Calderon Perrigo <i>et al.</i> ¹⁴ (1993)	27 f	Right frontoparietal	II	3×4 cm
Lacerte <i>et al.</i> ¹⁵ (1996)	32 f	Right frontal	II	8×6×6 cm
Khosrovi <i>et al.</i> ¹⁶ (2000)	14 m	Left frontoparietal	II	N/A
Nakayama <i>et al.</i> ¹⁷ (2001)	47 f	Right frontal	II	N/A
Colpan <i>et al.</i> ¹⁸ (2003)	40 f	Right frontoparietal	I	5×3.5×2.5 cm
Bergmann <i>et al.</i> ¹⁹ (2004)	30 f	Bifrontal	N/A	8×8×5 cm
Cosar <i>et al.</i> ²⁰ (2005)	21 f	Left frontal	I	2×2×2 cm
Hong <i>et al.</i> ²¹ (2005)	18 m	Left frontal	I	3×3×4 cm
Erdogan <i>et al.</i> ²² (2006)	14 m	Left frontal	II	6×7×4 cm
Delgado-Lopez <i>et al.</i> ⁷ (2007)	18 m	Left frontoparietal	N/A	N/A
Laghmari ²³ (2007)	50 m	Frontal parasagittal	N/A	N/A
Kawabata <i>et al.</i> ²⁴ (2010)	48 f	Left frontal	N/A	1.5×1.5×6.3cm
Maheshwari <i>et al.</i> ²⁵ (2011)	40 m	Left frontal	N/A	8 cm
Uddin <i>et al.</i> ²⁶ (2012)	23 m	Left frontal	N/A	10.0×7.5×1.4cm
Yeung <i>et al.</i> ²⁷ (2012)	22 f pregn	Right frontal	N/A	4.2×3.2×3.1 cm
Park <i>et al.</i> ²⁸ (2013)	55 f	Left frontal	N/A	5.9×3.5 cm
Atalay <i>et al.</i> ² (2014)	52 f	Left frontal	N/A	2×1 cm

N/A, not available.

described was 55 years old, whereas most of the patients had symptoms in a young age. There was no difference among men and women. The tumor reached in most of the cases a size of 6 cm. The neurological symptoms that the patients developed depended on the localization of the tumor and were usually epileptic seizures or motor deficits. Delgado-Lopez *et al.* published a patient with Noonan's syndrome and chondroma,⁷ a combination which was never described before. All authors agree that operative excision of the tumor is the best treatment option. No adjuvant therapy is then needed. Since intracranial chondromas are rare tumor entities, there are no grade 1 recommendations regarding their treatment. In all patients that were surgically treated and where the tumor was completely resected, no recurrence occurred. In our patient and after macroscopically complete tumor resection we planned the next MRI scan 6 months postoperatively. The further MRI scans will then be scheduled in one, two and five years or earlier, if symptoms appear.

Imaging

The diagnostic work-up includes MRI and

CT sequences. In the contrast enhanced MRI images the tumor appears ringlike with intratumoral nodular enhancement and no edema.⁷ In the computed tomography the tumor appears as an irregularly lobulated mass with obvious calcification. Nonetheless, a definitive diagnosis can only be made after histological examination of the tumor.

Differential diagnosis and immunohistochemistry

Intracranial chondromas present rare benign tumors of the central nervous system. The differential diagnosis primarily includes chondrosarcomas. In our case the initial diagnosis was suspicious for chondrosarcoma and therefore the histological specimen was sent to a specialized reference center. Histopathological analysis revealed a monomorphous tumor of low-moderate cellularity surrounded by a fibrous pseudocapsule. The mononucleated cells were embedded within a chondroid matrix, without signs of atypia. The tumor cells were strongly positive for S100 and negative for cytokeratine, GFAP, and EMA. Proliferative activity as determined by immunohistochemistry for Ki67 was almost

absent. Therefore, the final diagnosis was chondroma. Other tumors that have to be considered in the differential diagnosis are meningiomas,² hemangiopericytomas, gliomas and metastases. Immunohistochemistry is an important tool in establishing the diagnosis. Since chondromas are benign tumors and grow slowly, they are only then symptomatic, when the mass effect affects vital functions of the brain. Sugiura *et al.* performed a molecular analysis in their case of convexity chondroma which revealed wild type IDH1/2 and expression of HMGA2.²⁹

Therapy

Our patient presented with symptoms of focal mass effect and high intracranial pressure as well as seizures. As a result, we preferred a gross total resection over a possible stereotactic biopsy in order to alleviate the symptoms and to obtain a permanent histological diagnosis. Because of its huge size we also did not consider biopsy and radiosurgery as an option. Since the tumor was located in the eloquent precentral region, we preoperatively performed a digital tractography (fibertracking) and a functional MRI, which showed displace-

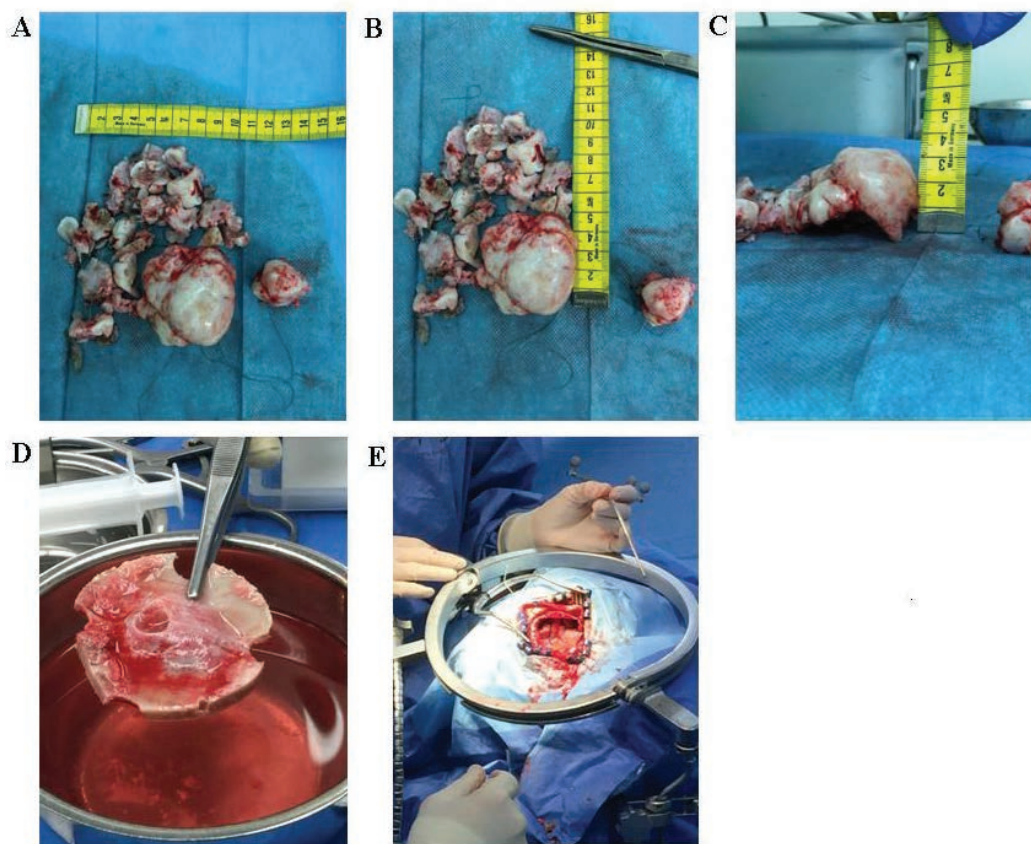


Figure 2. Macroscopical image of the resected tumor. The tumor was resected piecemeal as shown in A-C; D) Tumor growth into the calvaria. The tumor seems only to grow towards the calvaria and not to originate from it. The dura on the other side was infiltrated with tumor; E) Intraoperative image of the brain after tumor resection.

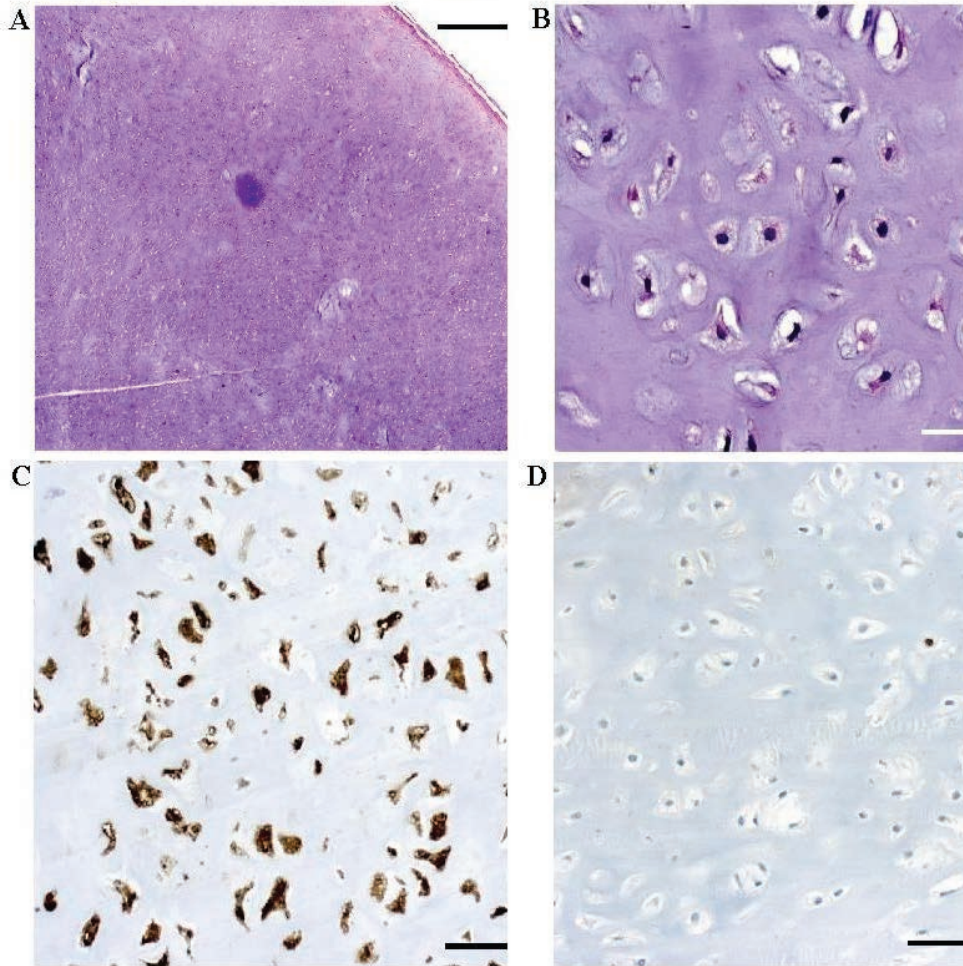


Figure 3. Histopathological analysis reveals a monomorphous tumor of low-moderate cellularity surrounded by a fibrous pseudocapsule [A: hematoxylin and eosin (H&E); scale bar = 1000 μm]. The mononucleated cells are embedded within a chondroid matrix, without signs of atypia (B: H&E; scale bar = 50 μm). The tumor cells are strongly positive for S100 (C: scale bar = 100 μm) and negative for cytokeratine, GFAP, and EMA (not shown). Proliferative activity as determined by immunohistochemistry for Ki67 is almost absent (D: scale bar = 100 μm).

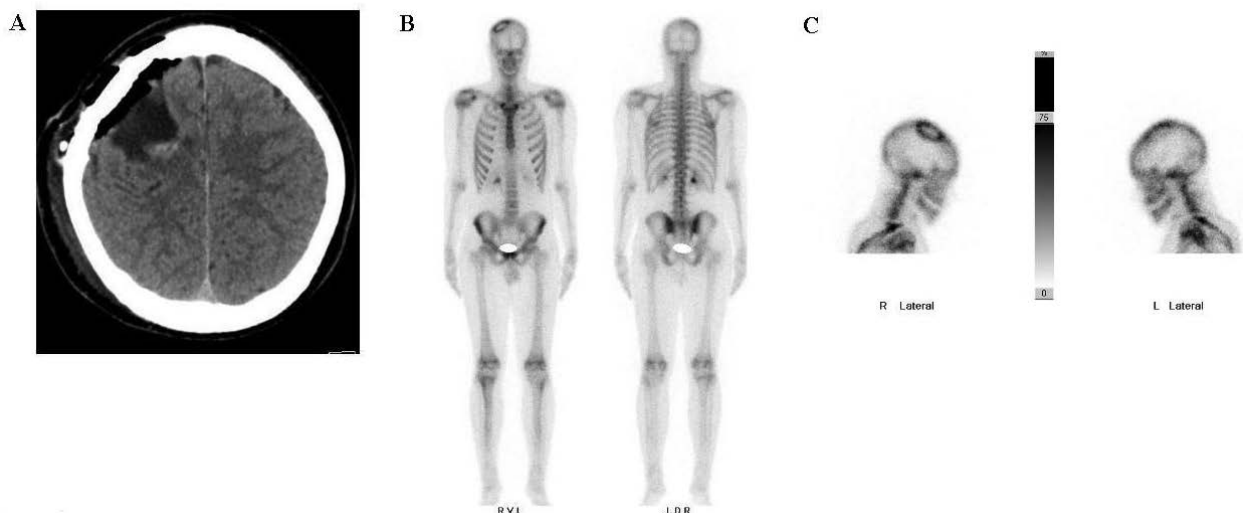


Figure 4. A) Postsurgical computed tomography showing tumor removal; B and C) Whole body scintigram showing no further tumor dissemination.

ment of the fibers posteriorly and anteriorly to the tumor. Since the tumor was well defined and did not diffusely infiltrate the neighboring fibers gross total resection was feasible.

Conclusions

Supratentorial dural chondromas are rare lesions that grow slowly and cause symptoms due to local mass effect or increased intracranial pressure. If they are located in eloquent brain areas, a thorough diagnostic imaging should be performed including functional imaging and fibertracking in order to pick the best possible approach with no or minimum manipulation of the critical structures. Aim of the operation should be gross total resection. The histological diagnosis can be challenging, but once established no adjuvant therapy has to be undertaken.

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