Intracranial osteolipomas and chondromas are rare types of benign tumors with reported prevalences of 0.01% and 0.5%, respectively, among all primary cerebral tumors. Meckel was the first to describe a hazelnut-sized, suprasellar lipoma in his seminal pathology textbook as early as 1816, while Hirschfeld first described a case of intracranial chondroma in 1851. Both entities are most commonly located in the sellar and interhemispheric regions. Chondromas are described as originating from synchondroses of the skull base, although rare cases of chondromas are believed to arise from cartilage remnants in the falx or the convexity. Associations with Maffucci syndrome (congenital mesodermal dysplasia, hemangiomatosis, and dyschondroplasia) and with Ollier disease (a polysystemic enchondromatosis) have been described in case reports. Chondromas are usually diagnosed in young adults. Pediatric cases are exceedingly rare, with just 2 reports in recent years documenting cases of supratentorial frontal chondromas in teenage boys. Osteolipomas are mainly located in relation to the corpus callosum, the tuber cinereum, and the paramesencephalic cisterns. They are believed to represent an abnormal differentiation of the meninx primitiva and are usually well vascularized and adherent to adjacent tissue components. Associations of intracranial lipomas and possibly an osteolipoma with Goldenhar-Gorlin syndrome (vertebro-auriculo-facial dysplasia) have been described in the literature. In this paper we present the case of a 9-year-old boy with an intratentorial osteochondrolipoma displaying components of both intracranial chondroma and osteolipoma.

**Case Report**

*History and Presentation.* This patient was born in 1992 in northern Ukraine to healthy unrelated parents after an uncomplicated full-term pregnancy. His weight at birth was 3450 grams, and he was 54 cm in length. He has 2 older siblings, both of whom are healthy. At birth, a tumorous occipital outgrowth in his skull was noticed, but was not further evaluated. The patient developed normally but presented to a hospital in February 2002 with headaches. A suboccipital tumor was diagnosed and a biopsy performed 1 week later, but the extracted tissue was nondiagnostic. The authors present a case of an osteochondrolipoma arising from the tentorium in a pediatric patient at the age of 9 years. The case and treatment are discussed, and a review of the literature is provided. (DOI: 10.3171/2009.1.PEDS08237)

**Key Words**
- brain tumor
- osteolipoma
- chondroma
- children
- cerebellum
- ataxia
- tentorium
- falx

**Abbreviations used in this paper:** EDTA = ethylenediaminetetraacetic acid; VP = ventriculoperitoneal.
Intratentorial osteochondrolipoma in a 9-year-old boy

Intratentorial tumor with a mass effect on infra- and supratentorial structures. The patient was then transferred to our hospital for further treatment in February 2006.

On admission the patient complained of intermittent and progressively worsening headaches and nausea. The neurological examination revealed moderate to severe ataxia and left-sided palsy of the third cranial nerve. Laboratory test results were unremarkable. The initial cranial MR imaging scan at our hospital showed an intratentorial tumor 7 × 4 × 4 cm in size, with expansion into both the supra- and infratentorial spaces, extending into a lipoma under the occipital bone. On MR imaging the tumor presented as a heterogeneous mass: it was hyperintense on T1- and T2-weighted imaging, showing slight contrast enhancement with hypointense margins. In addition, imaging revealed a large cerebellar hemispheric defect and enlargement of the ventricles. The tumor partially enclosed a segment of the straight sinus, which was also elevated and laterally displaced (Fig. 1).

**Operation and Postoperative Course.** The supratentorial part of the tumor was excised in April 2006. A vascular lipoma protruding through a 3-mm opening of the occipital skull was carefully removed. After opening the skull and the dura, the tumor was located inside the tentorium. Structurally, the tumor was composed of bony lamellae interspersed with highly vascularized lipomatous tissue. We discontinued this operation upon reaching the infratentorial segment of the tumor, when continuous bleeding from numerous areas of the tentorium became hemodynamically relevant. After successfully achieving hemostasis, the wound was closed in layers and the patient was scheduled for a second surgery in July 2006.

For the second procedure, a combined occipital and suboccipital approach was chosen via 1 incision and 2 craniotomies. Beginning from the suboccipital trephination, we were able to almost completely remove bone and lipomatous tumor tissue infratentorially, leaving only 1 bony lamella because it covered the vasculature draining into the straight sinus. Using occipital trephination we removed any remaining tissue supratentorially. Hemostasis was successfully attained and the patient was transferred to our neurosurgical intensive care unit.

Macroscopically, the tumor displayed both softer yellow, partly nodular fat tissue and firmer white-yellow tissue components with inclusion of many bone fragments. The softer parts of the tissue were fixed in formaldehyde, embedded in paraffin, cut into 7-μm sections, and stained with H & E. The firm components were initially decalcified in EDTA. Microscopically, the softer tumor parts appeared as mature univacuolated fat tissue and hyaline
cartilage partially arranged in lobules with strands of connective tissue (Fig. 2A). The hypocellular cartilage contained isomorphic S100-positive chondrocytes with uniform, small, round, and hyperchromatic nuclei (Fig. 2A). The decalcified firmer parts of the tissue revealed hyaline cartilage surrounded by a rim of osteoid and bone tissue (Fig. 2B). Throughout the different tumor components the proliferation rate (MIB-1, Ki 67) was 1–2%. Mitotic figures could not be observed (Fig. 2C). The characteristic findings of a chondroma as well as those of a lipoma with osseous and cartilaginous differentiation led to the diagnosis of an osteochondrolipoma, an entity that had previously been observed only extracranially.

After an uneventful postoperative course the patient was discharged to a rehabilitation facility. There he developed a CSF fistula, which presented with symptoms such as headaches and nausea. Cerebrospinal fluid microbiology tested positive for *Staphylococcus epidermidis*, and antibiotic treatment using vancomycin and cefotaxime was started immediately after implanting an external ventricular drain. After 7 days of treatment, no bacteria were present in the CSF. Because there was consistently elevated production of CSF (> 250 ml/24 hours at 15 cm H2O), we implanted a VP shunt. After several days of nausea and headaches in the morning hours, the patient improved markedly on the 4th day after implantation of the VP shunt. We discharged the patient in stable condition (Glasgow Outcome Scale score of 4) to the rehabilitation facility.

After living abroad in reportedly good health, the patient was invited to a follow-up examination in early 2008, ~ 2 years after the first resection. The patient’s neurological evaluation revealed improved yet still apparent gait insecurity, palsy of the sixth cranial nerve, and a slight mental retardation. No seizures were reported since discharge. He communicated freely and was independent in the majority of his daily activities. Magnetic resonance imaging revealed only minor changes from the imaging study at discharge, with parenchymal defects unchanged and normal ventricles.

**Discussion**

Initially, the tumor in the patient was diagnosed as a chondroma, which was supported by its close relationship to the falx and, moreover, to the tentorial structures. Intracranial chondromas are a rare type of primary brain tumor. The review by Matz et al. from 1981 is often cited, which notes a total of 139 cases of intracranial chondromas and chondrosarcomas. Since then, and to the best of our knowledge, another 170 cases have been reported in the literature, 45 of which constituted cases of intracranial chondromas whereas 125 were chondrosarcomas. Chondromas are commonly believed to arise from the cartilaginous remnants at the synchondroses of the skull base, a theory that is bolstered by the high number of parasellar cases published. Chondromatous metaplasia might explain the occurrence of intracerebral or meningeal tumors. Traumatic displacement is considered an alternative explanation of these clinical entities; in 2005, Hong et al. reported a case of intracranial chondroma at the site of a skull fracture that had occurred years earlier. According to Ramamurthi et al., as well as Russell and Rubinstein, aberrant nesting of cartilaginous tissue could also be an explanation for the development of these tumors. Chondromas are usually opalescent, firm, relatively large, and

![Fig. 2. Photomicrographs of excised tumor tissue. A: The hyaline cartilage mostly appears hypocellular. The chondrocytes have uniform and isomorphic nuclei. Binucleate cells were not identified. H & E. Bar = 200 μm. B: The decalcified tumor fraction revealed osteoid tissue besides cartilaginous tissue. H & E (after EDTA decalcification). Bar = 200 μm. C: Low proliferation activity of tumor cells (~ 1–2%). MIB-1/ Ki 67. Bar = 100 μm.](image-url)
Intratentorial osteochondrolipoma in a 9-year-old boy

**TABLE 1: Summary of falcine and tentorial chondromas in adults reported in the literature from 1932 to 2008**

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Patient Age (yrs), Sex</th>
<th>Location</th>
<th>Clinical Symptoms</th>
<th>Imaging Findings</th>
<th>Pathology</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Verbrugghen, 1932</td>
<td>39, M</td>
<td>rt frontoparietal</td>
<td>seizures, Lt hemiparesis</td>
<td>radiograph: calcified tumor</td>
<td>chondroma w/ some calcification, no bony structures</td>
<td>favorable</td>
</tr>
<tr>
<td>Hardy et al., 1978</td>
<td>22, F</td>
<td>rt frontoparietal</td>
<td>headaches, hemiparesis, dizziness</td>
<td>CT: hypodense center, contrast-enhancing; MRI: T1 hypointense, T2 hyperintense, center hypointense, rim enhancement</td>
<td>lobulated hyaline cartilage, central cavitary degeneration, immunohistochemistry positive</td>
<td>NA</td>
</tr>
<tr>
<td>Ozgen et al., 1984</td>
<td>39, M</td>
<td>It frontal, parasagittal</td>
<td>headaches, seizures</td>
<td>CT: calcifications; MRI: T1 rim hyperintense, heterogeneously enhancing mass</td>
<td>low-cellular cartilage, grouped chondrocytes</td>
<td>&quot;well&quot; at 6 mos</td>
</tr>
<tr>
<td>Kretzschmar et al., 1989</td>
<td>43, F</td>
<td>It frontoparietal</td>
<td>headaches, amnesia, difficulty concentrating, aphasia</td>
<td>NA</td>
<td></td>
<td>NA</td>
</tr>
<tr>
<td>Hadadian et al., 1991</td>
<td>25, F</td>
<td>frontal, parasagittal</td>
<td>headaches, seizure</td>
<td></td>
<td></td>
<td>NA</td>
</tr>
<tr>
<td>Kurt et al., 1996</td>
<td>27, M</td>
<td>frontal</td>
<td>headache, amnesia, lack of concentration</td>
<td></td>
<td></td>
<td>favorable</td>
</tr>
<tr>
<td>De Coene et al., 1997</td>
<td>35, M</td>
<td>rt posterior</td>
<td>&quot;mental deterioration&quot;</td>
<td></td>
<td></td>
<td>NA</td>
</tr>
<tr>
<td>Pallini et al., 1997</td>
<td>22, M</td>
<td>lt parietal, parasagittal</td>
<td>flaccid paralysis, distal rt hemiparesis, seizure</td>
<td></td>
<td></td>
<td>favorable</td>
</tr>
<tr>
<td>Ustun et al., 1997</td>
<td>28, F</td>
<td>rt temporoparietal</td>
<td>headache, double vision, lt paresthesia, nausea, vomiting</td>
<td></td>
<td></td>
<td>NA</td>
</tr>
<tr>
<td>Ishibashi et al., 1999</td>
<td>60, M</td>
<td>infratentorial, cerebellar</td>
<td>unsteady gait, truncal ataxia</td>
<td></td>
<td></td>
<td>favorable</td>
</tr>
<tr>
<td>Schreckenberger et al., 1999</td>
<td>21, F</td>
<td>rt frontal</td>
<td>headache, sensory jacksonian seizures</td>
<td></td>
<td></td>
<td>favorable</td>
</tr>
<tr>
<td>Bergmann et al., 2004</td>
<td>19, F</td>
<td>frontal, parasagittal</td>
<td>headache, apraxia, transient rt hemiparesis</td>
<td></td>
<td></td>
<td>favorable</td>
</tr>
<tr>
<td>Cosar et al., 2005</td>
<td>44, M</td>
<td>frontal, parasagittal</td>
<td>headache</td>
<td></td>
<td></td>
<td>favorable</td>
</tr>
<tr>
<td>Li et al., 2005</td>
<td>26, F</td>
<td>frontal, parasagittal</td>
<td>headache</td>
<td></td>
<td></td>
<td>favorable</td>
</tr>
<tr>
<td>Erdogan et al., 2006</td>
<td>50, F</td>
<td>frontal, inside falx</td>
<td>headache</td>
<td></td>
<td></td>
<td>NA</td>
</tr>
<tr>
<td>Fountas et al., 2008</td>
<td>30, M</td>
<td>lt parietal, parasagittal</td>
<td>seizure</td>
<td></td>
<td></td>
<td>favorable</td>
</tr>
</tbody>
</table>

* NA = not available.

lobulated. Fifteen percent of intracranial cases are of dural origin, arising from either the falx cerebri or the convexities.5,6 We reviewed 16 cases of chondromas of falcine origin in adults that have been published between 1932 and 2008 (Table 1).

The present tumor specimen also showed osseous and lipomatous components. Friede11 conducted a detailed literature review yielding a total of 12 cases of osteolipomas by 1977. When combined with cases published afterward as well as those omitted by Friede, 21 intracranial osteolipomas have been reported to date.3,9,12,28,33,37,42–44

Unlike previous cases of osteolipomas described in the literature, the present tumor was not located at the tuber cinereum or the corpus callosum, but within meningeal structures. Intraoperatively, no structures adherent to brain parenchyma were observed, which is also unlike the characteristic osteolipomas described by Friede11 and by Sinson et al.37 These findings are consistent with an outside reference and with pathology reports of chondromas published in the literature.19,33 Yet main structures of this tumor specimen also had osteolipomatous components. According to Rau et al.,32 the nomenclature of osteochondrolipomatous lesions is controversial and, because the predominant component is a mature fat tissue, the tumor is still best considered a lipoma with mature cartilaginous and osseous differentiation, or more specifically an osteochondrolipoma. Among the cases reviewed, 3 pediatric cases of falcine chondroma and 4 pediatric osteolipomas could be identified, none of which arose intratentorially.3,4,12,13,22,41,44 These cases are listed in

**TABLE 2: Summary of falcine chondromas in pediatric patients reported in the literature from 1931 to 2008**

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Patient Age (yrs), Sex</th>
<th>Location/Size</th>
<th>Clinical Symptoms</th>
<th>Imaging Findings</th>
<th>Pathology</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Brüt, 1931</td>
<td>17, F</td>
<td>lt frontoparietal/10 × 10 × (unknown) cm</td>
<td>headaches, seizures</td>
<td>radiograph: calcified tumor</td>
<td>chondroma w/ some calcification, no bony structures</td>
<td>favorable at 15 mos, patient working</td>
</tr>
<tr>
<td>Luzardo-Small et al., 1999</td>
<td>14, M</td>
<td>lt frontoparietal, parasagittal/size unspecified</td>
<td>headache, proximal rt hemiparesis, seizure</td>
<td>CT: hypodense center, contrast-enhancing; MRI: T1 hypointense, T2 hyperintense, center hypointense, rim enhancement</td>
<td>lobulated hyaline cartilage, central cavitary degeneration, immunohistochemistry positive</td>
<td>NA</td>
</tr>
<tr>
<td>Geramizadeh et al., 2007</td>
<td>17, M</td>
<td>frontal, midline anterior fossa/5 × 5 × 4 cm</td>
<td>headache</td>
<td>CT: calcifications; MRI: T1 rim hyperintense, heterogeneously enhancing mass</td>
<td>low-cellular cartilage, grouped chondrocytes</td>
<td>&quot;well&quot; at 6 mos</td>
</tr>
</tbody>
</table>


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drolipomas in this location have not yet been described on CT/MR imaging. Osteolipomas and even osteochondrolipomas may show very similar features, both clinically as well as on CT/MR imaging. However, this could not be clearly distinguished from the mass effect of the tumor itself because the headaches were aggravating before admission. The patient, however, be due to a marginally compensated hydrocephalic state. This made the complete removal of the bony capsule impossible in our case. Nevertheless, at follow-up after more than 2 years, no tumor growth was detectable on MR imaging and the boy displayed significant neurological improvement. This unique intracranial tumor shows the characteristic findings of a chondroma as well as those of a lipoma with osseous and cartilaginous differentiation, thus leading to the diagnosis of an osteochondrolipoma.

This tumor can be clearly distinguished from a chondroma lipoma, which has a considerably more immature aspect with multivacuolated pleomorphic cells and myxoid changes. Chondromas of falx cerebri and tentorial tumor of this kind can be considered in patients presenting with a slightly enhancing mass on MR imaging that is sharply demarcated from the parenchyma, with a hypointense rim and a heterogeneous center.

Conclusions

Intracranial chondromas and osteolipomas are rarities in pediatric patients. In this paper we present the first case of an intracranial osteochondrolipoma arising from the tentorium. The location of the tumor presented here caused significant difficulties in resection because the straight sinus passed through its firm cartilaginous tissue. This made the complete removal of the bony capsule impossible in our case. Nevertheless, at follow-up after more than 2 years, no tumor growth was detectable on MR imaging and the boy displayed significant neurological improvement. This unique intracranial tumor shows the characteristic findings of a chondroma as well as those of a lipoma with osseous and cartilaginous differentiation, thus leading to the diagnosis of an osteochondrolipoma.

This tumor can be clearly distinguished from a chondroma lipoma, which has a considerably more immature aspect with multivacuolated pleomorphic cells and myxoid changes. Chondromas of falx cerebri and tentorial origin may show very similar features, both clinically as well as on CT/MR imaging. Osteolipomas and even osteochondrolipomas in this location have not yet been described in the literature. The differential diagnosis of a falcine or tentorial tumor of this kind can be considered in patients presenting with a slightly enhancing mass on MR imaging that is sharply demarcated from the parenchyma, with a hypointense rim and a heterogeneous center.

Disclaimer

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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* Adapted and expanded from Friede, 1977.
Intratentorial osteochondrolipoma in a 9-year-old boy

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