Mesenchymal Chondrosarcoma of the Cerebellum*

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ABSTRACT

Mesenchymal chondrosarcoma is a rare malignant neoplasm of bone and soft tissues. An unique case is described of an 8-year-old child with a midline cerebellar lesion. Pertinent clinical and radiologic findings along with histopathologic features are described. To our knowledge, this is the first case of mesenchymal chondrosarcoma arising in the cerebellar parenchyma of a child.

Introduction

Sarcomas of the central nervous system are uncommon lesions. Mesenchymal chondrosarcoma represents a very small subset of intracranial sarcomas, which by themselves, constitute less than 2 percent of primary intracranial tumors.1,2 Mesenchymal chondrosarcoma was first described as a skeletal neoplasm in 19593 but it is now also known to occur in soft tissues, viscera and the meninges.4 Mesenchymal chondrosarcomas are composed of the two distinct histological components consisting of undifferentiated mesenchymal cells intermixed with islands of bland-looking neoplastic hyaline cartilage. The exact histogenesis of mesenchymal chondrosarcomas in the brain parenchyma is unknown but multiple theories have been proposed.5 A case described here is of an 8-year-old child who presented with a midline cerebellar mass mimicking radiologically medulloblastoma.

Case Report

An 8-year-old child was admitted to the hospital with a two-day history of occipital and bitemporal headaches, diplopia, blurry vision, ataxia and a nine-day history of abdominal pain and vomiting. His past medical history was significant for viral meningitis at 5 years of age. Physical examination revealed a right-sixth nerve palsy and positive cerebellar signs, including horizontal and upgaze nystagmus, dyscoordination, unsteady gait, intentional tremor, and positive Romberg's sign.

Radiologic Findings: A non-contrast head computer tomography (CT) demonstrated anterior displacement of the fourth ventricle by an isodense mass with associated obstructive hydrocephalus. Magnetic resonance imaging (MRI) revealed a 3 cm well-defined midline cerebellar mass, extend-

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ing into the fourth ventricle. The lesion was hypointense to gray matter on T1 and hyperintense on T2 weighted images with predominantly homogeneous contrast enhancement. Scattered foci of low signal were also noted on the T2 and post-contrast images.

A complete resection of the tumor was performed. Metastatic workup demonstrated no evidence of systemic disease. Surgery was followed by local posterior fossa radiation and a high dose of chemotherapy. The patient’s symptoms initially improved, and he remained symptom free for 8 months. He then presented with symptoms similar to those on initial presentation. The MRI scan revealed leptomeningeal dissemination of tumor extending throughout the entire length of the spinal cord. He is presently receiving intraventricular chemotherapy.

**Pathologic Findings:** On gross examination, the specimen measured 3.0 cm in greatest diameter, was red to grey and inhomogeneously firm in consistency with a glistening outer surface. Histologically, the lesion was composed of two distinctly different elements. The first included clusters of undifferentiated round to spindly mesenchymal cells arranged in monotonous sheets and parallel bundles. The individual cells were small, round to oval, with scanty cytoplasm, high nuclear cytoplasmic ratio, dark homogeneously dispersed chromatin and a very high mitotic rate. A few scattered cells were larger with more open vesicular chromatin and occasional inconspicuous nucleoli mixed with undifferentiated mesenchymal cells (figure 1). The undifferentiated cells were strongly positive for vimentin and negative for glial fibrillary acidic protein (GFAP), S-100 protein, synaptophysin, myoglobin, and smooth muscle actin.

The other distinct histologic component consisted of multiple tumor nests with cartilaginous differentiation. Islands of hyaline cartilage were seen in varying stages of maturity (figure 2). The cartilage was bland-looking and often comprised of single large vesicular nuclei within lacunae surrounded by abundant hyaline stroma. Occasional binucleated and multinucleated forms were seen (figure 3). These areas were GFAP-positive and merged inconspicuously with the undifferentiated mesenchymal elements. Multiple dilated and compressed capillary vessels were also seen throughout the tumor, particularly in areas at the junction of undifferentiated mesenchymal and chondroid elements. On the basis of these features, the diagnosis of a high-grade sarcoma with chondrosarcomatous and undifferentiated mesenchymal elements was made.

**Discussion**

Mesenchymal chondrosarcoma is a high grade malignant neoplasm with a very high tendency for local recurrence and distant metastasis. It was first described by Lichtenstein and Bern-
stein. They reported two cases. Both neoplasms metastasized diffusely throughout the skeleton causing death of the patients in very short intervals. They interpreted these tumors as being derived from cartilage-forming mesenchymal areas and fields with cartilaginous differentiation are interrupted by prominent dilated and compressed capillary vessels. Nests of cells with abortive cartilaginous differentiation are also seen. (H&E ×450)

Figure 2. Transition between undifferentiated mesenchymal areas and fields with cartilaginous differentiation are interrupted by prominent dilated and compressed capillary vessels. Nests of cells with abortive cartilaginous differentiation are also seen. (H&E ×150)

Figure 3. Areas with chondroid differentiation showing chondrocytes and chondroblasts with round to oval nuclei, vesicular chromatin and inconspicuous nucleoli, surrounded by abundant hyaline matrix. (H&E ×450)
chyme and assumed to be multicentric in origin.  

Initially, mesenchymal chondrosarcomas were thought to be exclusively intraskeletal neoplasms but it is now apparent that both skeletal and extraskeletal forms exist. Approximately 50 percent are extraskeletal in origin and CNS meninges are known to be one of the most common extraskeletal sites of this rare neoplasm. Because of their rarity, meningeal location and high vascularity, clinically and radiologically, they are often misdiagnosed as vascular malformations, atypical meningiomas, calcific subdural hematomas and other vascular lesions.

This particular case, owing to its location, age of the patient, clinical and radiologic findings was thought to be a cerebellar medulloblastoma. Histologically, the tumor demonstrates two basic components. The first is composed of highly cellular, undifferentiated round to oval somewhat spindly mesenchymal cells with scanty cytoplasm randomly arranged in sheets containing scattered compressed capillary blood vessels. These undifferentiated mesenchymal cells, which may display alveolar and perivascular arrangement, are intermixed with islands of well-differentiated bland-looking neoplastic hyaline cartilage in varying stages of maturity. The transition between the two areas is usually abrupt, and the non-chondroid elements are dominant.

To our knowledge, 8 cases of mesenchymal chondrosarcoma of the cerebellum were reported, as summarized in Table I. The cases are presented in chronological order of publication with full references provided for each entry. The table includes the author of the report, age of the patient, sex, location of the tumor, and follow-up status.

### Table I

<table>
<thead>
<tr>
<th>Author</th>
<th>Age</th>
<th>Sex</th>
<th>Location</th>
<th>Follow-up</th>
</tr>
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<tbody>
<tr>
<td>Flyger et al</td>
<td>11</td>
<td>M</td>
<td>R frontal</td>
<td>Alive and well</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>(5 mo. postop)</td>
</tr>
<tr>
<td>Raskind et al</td>
<td>48</td>
<td>F</td>
<td>R frontal</td>
<td>Alive and well</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>(3 days postop)</td>
</tr>
<tr>
<td>Heros et al</td>
<td>26</td>
<td>F</td>
<td>L cerebellum</td>
<td>Alive and well</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>(2.5 years, postop)</td>
</tr>
<tr>
<td>Richman et al</td>
<td>59</td>
<td>M</td>
<td>R hemisphere</td>
<td>Alive and well</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>(7 mo. postop)</td>
</tr>
<tr>
<td>Harwood et al</td>
<td>22</td>
<td>M</td>
<td>Cerebellum</td>
<td>Died with local tumor</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>(4 mo. postop)</td>
</tr>
<tr>
<td>Banerjee et al</td>
<td>40</td>
<td>M</td>
<td>L hemisphere</td>
<td>Died</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>(8 days postop)</td>
</tr>
<tr>
<td>Parker et al</td>
<td>14</td>
<td>F</td>
<td>L thalamus</td>
<td>Died</td>
</tr>
<tr>
<td>Chhem et al</td>
<td>11</td>
<td>F</td>
<td>L parietal</td>
<td>Died with recurrence</td>
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<td>(1.5 years postop)</td>
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<tr>
<td>Present case</td>
<td>8</td>
<td>M</td>
<td>Midline cerebellum</td>
<td>Alive with recurrence</td>
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<tr>
<td></td>
<td></td>
<td></td>
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<td>(1.5 years postop)</td>
</tr>
</tbody>
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*Diagnosed at autopsy.


chymal chondrosarcoma have been reported to occur within the brain parenchyma without any dural attachment.\textsuperscript{5,8,9,11,12,13} (see table I). The ages of these patients ranged from 11 years to 59 years with the mean of 29 years. Two of the reported cases occurred in the cerebellum, one in a 26 year old female\textsuperscript{8} in whom the tumor was present in the left cerebellar hemisphere and the patient was alive without recurrent or metastatic disease 2.5 years after surgery. Another case was in a 22 year old male.\textsuperscript{6} No details were reported except that the patient died owing to a local tumor within four months of diagnosis.

In summary, intracranial mesenchymal chondrosarcoma is a rare and highly malignant form of extraskeletal chondrosarcomas. It seems to occur intracranially at almost any site, including the cerebellum, and may occur in adults as well as in children.

**Acknowledgment**

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**References**