Case Report

Intracranial Extraskeletal Myxoid Chondrosarcoma: Case Report and Literature Review

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Intracranial extraskeletal myxoid chondrosarcoma is extremely rare and is thought to arise from the choroid plexus, dura, or in rare instances the pineal region.¹⁻³,⁸⁻¹²,¹⁷) Since its first description in 1972, only seven intracranial extraskeletal myxoid chondrosarcoma tumors have been reported.²,⁴⁻⁵,⁸⁻¹³⁻¹⁵,¹⁷) We describe a patient with an intracranial myxoid chondrosarcoma originating from the choroid plexus of the left lateral ventricle.

INTRODUCTION

Intracranial myxoid chondrosarcoma is extremely rare and is thought to arise from the choroid plexus, dura, or in rare instances the pineal region.¹⁻³,⁸⁻¹²,¹⁷) Since its first description in 1972, only seven intracranial extraskeletal myxoid chondrosarcoma tumors have been reported.²,⁴⁻⁵,⁸⁻¹³⁻¹⁵,¹⁷) We describe a patient with an intracranial myxoid chondrosarcoma originating from the choroid plexus of the left lateral ventricle.

CASE REPORT

A 21-year-old woman was admitted with weakness in the right extremities that had persisted for over a month. She had a known brain tumor, which has been diagnosed in another country. At that time, she experienced headaches as well as visual and hearing disturbances. Previous brain magnetic resonance imaging (MRI) showed a well-enhanced mass, measuring 26×40×29 mm, in the posterior horn of the left lateral ventricle. Ventricular dilatation was absent, and peritumoral edema was observed. For financial reasons, however, she was not treated at that time.

Upon admission to our hospital, she complained of both visual and hearing loss. She also showed a grade II weakness of her right extremities and symptoms of increased intracranial pressure. Magnetic resonance imaging (MRI) revealed hydrocephalus and a well-enhanced large mass around her left thalamus. A left parietal craniotomy and a cortisectomy at the superior parietal lobule were performed. Total surgical resection was also performed, and pathology results confirmed an extraskeletal myxoid chondrosarcoma. Postoperative MRI showed no residual tumor, and the patient underwent radiotherapy. After six months of radiotherapy, the patient’s headache and weakness had improved to grade IV. This malignant tumor showed high rates of recurrence in previous reports. We here report another occurrence of this highly malignant and rare tumor in a patient treated using total surgical excision and adjuvant radiotherapy.

Key Words: Brain neoplasms · Chondrosarcoma · Choroid plexus.
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dispersed chromatin, and a moderate amount of eosinophilic cytoplasm that was often finely vacuolated (Fig. 2B). Mitotic figures were rarely observed.

The tumor cells were further examined by immunohistochemistry, and antibodies were used at the dilutions listed. Tumor cells were found to be focally and strongly positive for epithelial membrane antigen (1 : 25, Dako, Glostrup, Denmark) (Fig. 2C), weakly positive for class III β-tubulin (1 : 200, clone TU-20, Genetex, Irvine, CA, USA), diffusely positive for microtubule-associated protein 2 (1 : 200, clone AP18, NeoMarkers, Fremont, CA, USA) (Fig. 2D), and positive for vimentin (1 : 250, Zymed, San Francisco, CA, USA) (Fig. 2E). In contrast, the tumor cells were negative for S-100 protein (1 : 1000, Zymed, San Francisco, CA, USA), cytokeratin (1 : 250, Zymed San Francisco, CA, USA), and glial fibrillary acidic protein (GFAP, 1 : 200, Biogenex, San Ramon, CA, USA). Final pathologic analysis of the above results led to a diagnosis of extraskeletal myxoid chondrosarcoma. Postoperative MRI showed no residual tumor. The patient then underwent adjuvant radiotherapy, at a total dose of 6080 cGy, as well as rehabilitation. After six months of treatment, the headache and weakness symptoms had improved to grade IV, but other neurologic deficits, including blindness and deafness, were unchanged.

DISCUSSION

Histologically, three subtypes of cranial and intracranial chondrosarcomas have been described: classic, mesenchymal, and myxoid5,7,9. Intracranial extraskeletal myxoid chondrosarcomas are extremely rare, with only seven cases previously reported to date2,5,8,13-15,17). A summary of these previous patients, including imaging results, the surgical extent of the tumor, postoperative radiation treatment, and patient outcomes is shown in Table 1.

Intracranial extraskeletal myxoid chondrosarcomas are thought to originate from the dura, leptomeninges, parenchyma, and choroid plexus2,5,8,13-15). Our findings in the present case suggest that the tumor originated from the choroid plexus of the lateral ventricle.

Preoperative imaging methods, including CT and MRI, revealed similar results in previous patients with intracranial extraskeletal myxoid chondrosarcomas. Precontrast CT scans have shown isodensity of tumors in five of the seven previously reported patients; however, it should be noted that the two remaining patients had preoperative tumor bleeding16,17). Most
previous tumors of this type were calcified, with one showing greater than 50% calcification. All of these previous tumors also showed contrast enhancement on CT scans. T1-weighted MRI scans usually showed contrast enhancement of the tumors. Previous immunohistochemical analyses showed that all seven of the previously reported cases expressed vimentin, with some expressing epithelial membrane antigen, neurofilament, and synaptophylin. In contrast, S-100 staining is very rare and usually focal.

The optimal treatment for intracranial myxoid chondrosarcoma is radical excision, and total removal of the tumor is critical. In six of the eight patients diagnosed with this tumor to date, including our current patient, the tumors were completely removed by surgical excision. Fortunately, these tumors have well-defined margins and are clearly distinct from normal brain tissue, making them amenable to complete resection. However, it is difficult to completely remove tumors located in the cerebellopontine angle, the pineal region, or the sellar portion of the brain. In addition, intracranial myxoid chondrosarcomas are not associated with a good prognosis. Of the seven patients previously described, two showed tumor recurrences and two died due to postoperative complications. Adjuvant therapies, including chemotherapy and radiation therapy, have been used to improve patient outcomes in cases of intracranial myxoid chondrosarcoma. These therapies may include surgery, radiation, and chemotherapy, as well as targeted therapies and immunotherapy.

### Table 1. The characteristics of previous cases of intracranial myxoid chondrosarcoma

<table>
<thead>
<tr>
<th>Authors</th>
<th>Reference year</th>
<th>Age (years)/sex</th>
<th>Location/origin</th>
<th>Size</th>
<th>Surgical extent</th>
<th>Postoperative radiotherapy, dose</th>
<th>CT</th>
<th>MRI</th>
<th>Postoperative course</th>
</tr>
</thead>
<tbody>
<tr>
<td>Scott et al.</td>
<td>1976</td>
<td>39/male</td>
<td>4th ventricle, choroid plexus</td>
<td>Not described</td>
<td>STL</td>
<td>Not done</td>
<td>Not done</td>
<td>T1: hypointense</td>
<td>13 days died d/t ventriculitis</td>
</tr>
<tr>
<td>Salzman et al.</td>
<td>1992</td>
<td>28/female</td>
<td>Left parafalcine &amp; dura of falx</td>
<td>Not described</td>
<td>70×50×40 mm</td>
<td>TR</td>
<td>Not done</td>
<td>Isodense</td>
<td>20 months alive and local recurrence</td>
</tr>
<tr>
<td>Sato et al.</td>
<td>1993</td>
<td>43/female</td>
<td>Pineal gland &amp; dura</td>
<td>Not described</td>
<td>PR</td>
<td>Yes, 6000 cGy</td>
<td>Enhancement (+)</td>
<td>T1: hypointense</td>
<td>3 years dead d/t tumor progression</td>
</tr>
<tr>
<td>Chaskis et al.</td>
<td>2002</td>
<td>69/male</td>
<td>Right F. cortex</td>
<td>Not described</td>
<td>TR</td>
<td>Not done</td>
<td>Not described</td>
<td>Enhancement (+)</td>
<td>1 months died with septic shock d/t diverticulitis</td>
</tr>
<tr>
<td>González et al.</td>
<td>2002</td>
<td>17/female</td>
<td>Right F-P cortex</td>
<td>23×20 mm</td>
<td>TR</td>
<td>No, 6000 cGy</td>
<td>Enhancement (+)</td>
<td>T1: hypointense</td>
<td>20 months alive, twice had tumor recurrence</td>
</tr>
<tr>
<td>Im et al.</td>
<td>2003</td>
<td>43/male</td>
<td>Left P. cortex</td>
<td>20 mm</td>
<td>TR</td>
<td>Yes, 5940 cGy</td>
<td>Enhancement (+)</td>
<td>T1: hypointense</td>
<td>3 years alive, no recurrence</td>
</tr>
<tr>
<td>Sorimachi et al.</td>
<td>2008</td>
<td>37/female</td>
<td>Pineal region</td>
<td>Not described</td>
<td>1st : PR</td>
<td>2nd : TR</td>
<td>Not done</td>
<td>Mixed dense</td>
<td>1st : 13 months recurrence 2nd : 7 months alive, no tumor recurrence</td>
</tr>
<tr>
<td>Present case</td>
<td>2011</td>
<td>21/female</td>
<td>Left lateral ventricle/choroid plexus</td>
<td>32×63×48 mm</td>
<td>TR</td>
<td>6080 cGy</td>
<td>Isodense</td>
<td>T1: hypointense</td>
<td></td>
</tr>
</tbody>
</table>

recurrence in previous reports. Here, we present a case report of this tumor treated with total tumor resection and adjuvant radiotherapy.

References