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Desmoplastic Medulloblastoma Arising From an Ovarian Teratoma: A Case Report and Review of the Literature

Shiuan-Li Wey, MD¹, Chi-Kuan Chen, MD¹, Tze-Chien Chen, MD¹, and Tung-Ying Chen, MD¹

Abstract
The primary neuroectodermal tumor of the ovary is extremely rare, and it manifests in 3 forms: differentiated, primitive, and anaplastic. The medulloblastoma belongs to the subgroup of primitive neuroectodermal tumor of the ovary. Only 3 cases of ovarian medulloblastoma have been reported in the literature, and all of them are presented without information about the specific pathological subtype. We present the fourth case of a 26-year-old woman who presented with abdominal fullness for months. Ultrasound exam showed a right adnexal mass with cystic feature and foci solid components. A partial oophorectomy was performed, and the mass was a desmoplastic medulloblastoma arising from an ovarian teratoma.

Keywords
ovarian medulloblastoma, primitive neuroectodermal tumor, poorly differentiated neuroectodermal tumor, teratoma, desmoplastic medulloblastoma

Introduction
Primary ovarian medulloblastomas belong to the subgroup of primitive neuroectodermal tumors of the ovary, which have the histological features of tumors that occur in the central nervous system of children.¹ According to the World Health Organization classification of ovarian tumors, primitive neuroectodermal tumors are categorized under the group of monodermal teratoma.¹,² Only 3 cases of primary ovarian medulloblastoma have been reported in the literature in English, and all of them belong to the group of classical medulloblastomas.¹,³ Other variants of ovarian medulloblastomas have never been reported in the literature. We present the first case of an ovarian desmoplastic medulloblastoma arising from a teratoma.

Case Report
A 26-year-old nulliparous female had a history of laparotomy for left ovarian mucinous cystadenoma torsion 3 years ago. The patient had suffered from abdominal fullness, a palpable progressively enlarging abdominal mass, and an irregular menstrual cycle for months prior to visiting the gynecologist’s clinic. Ultrasound exam showed a right adnexal mass measuring about 14.7 × 9.79 cm², with cystic features and a focal solid component. Abdominal computed tomography exam showed a well-defined mass lesion with mixed density in the lower abdomen. The mass lesion consisted of well-formed calcifications, fat components, and focal solid parts, suggesting a teratoma. Serum CA-125, carcinoembryonic antigen, and α-fetoprotein levels were within normal limits. The patient received a partial oophorectomy.

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On gross examination, the ovarian cystic mass was 12 cm in diameter with a smooth external surface and filled with sebum. A yellow-gray, solid protruding nodule was observed in the cystic wall. Microscopically, most of the cystic mass showed features of a mature cystic teratoma composed of mature squamous epithelium, sebaceous gland, choroid plexus, and glial tissue (Figure 1A). A mural tumor, measuring 2.1 cm in diameter, protruded into the cystic space (Figure 1B). This tumor mass was composed of many hypercellular and nodular structures (Figure 1C). The pale nodular area was surrounded by densely packed hyperchromatic cells (Figure 2A). The cells in the pale nodular area were more uniform and mature than those in the extranodular areas, which exhibited a reduced nuclear cytoplasmic ratio and increased fibrillary matrix (Figure 2B). Cells with perinuclear halo and oligodendrocyte-like appearance were found in some pale nodular areas (Figure 2C). The tumor cells in the extranodular regions showed increased cellularity, pleomorphism, and high-grade atypia (Figure 2D). The immunohistochemical stains revealed synaptophysin to be strongly positive, particularly in the pale nodular region (Figure 3A). The glial fibrillary acidic protein stain was positive with a long, branched pattern in pale nodules and diffusely positive in the cells in extranodular areas (Figure 3B). Vimentin and CD56 were also strongly positive. Reticulin stain appeared negative in the pale nodular regions but positive in the extranodular areas (Figure 3C). The tumor cells were negative for cytokeratin, CD99, desmin, inhibin-α, estrogen receptor, progesterone receptor, and HMB-45. Based on the morphology and immunohistochemical results, the final diagnosis was a desmoplastic medulloblastoma arising from an ovarian teratoma.
Discussion

Primary ovarian neuroectodermal tumors are extremely rare, and only about 60 cases have been reported in the literature in English. These tumors resemble neoplasms of the central nervous system with different degrees of differentiation. They are divided into 3 categories based on the histological features: differentiated, primitive, and anaplastic tumors. The most common group is the differentiated tumors, including ependymoma and astrocytoma. The anaplastic tumors have the histological features of glioblastoma multiforme. The primitive tumors include neuroblastoma, medulloblastoma, medullopithelioma, and ependymoblastoma. The primitive neuroectodermal tumors are composed of small cells with hyperchromatic, round to oval nuclei and scanty cytoplasm. About 21 cases of primitive neuroectodermal tumors of the ovary have been reported in the literature, of which only 3 cases were medulloblastomas. The average age of these patients was 35.5 (23-69) years, and all the tumors were unilateral. The tumors ranged from 11 to 17 cm in diameter (Table 1). Two of the 3 cases presented with the histological feature of a uniformly undifferentiated pattern. In the third case, the tumor was arranged in

Figure 2. Desmoplastic/Nodular medulloblastoma: A. The pale nodular area is surrounded by hypercellular and hyperchromatic cells (10× objective). B. Tumor cells in pale nodule show increased cytoplasm and neuropils (10× objective). C. Cells with perinuclear halo with an oligodendrocyte-like appearance in the pale nodular area; the cells in the extranodular area show high-grade atypia and high nuclear-cytoplasmic ratio (20× objective). D. The border between cellular and pale islands (40× objective)
A poorly delineated trabecular pattern with neuropil differentiation and pseudo-rosette formation. All these 3 cases were classical medulloblastomas. However, the tumor in our case had a feature of desmoplastic medulloblastoma, showing focal differentiation with characteristic reticulin-free nodular zones surrounded by densely packed, hyperchromatic cells. The reticulin-free zones have increased cytoplasm and neuropil. The intervening background of desmoplastic medulloblastoma is composed of highly cellular, actively proliferative, reticulin-rich areas. The tumor cells display neuronal maturation within the nodular area, which is strongly stained with synaptophysin and to a lesser extent with glial marker. The distinctive histological features and the immunohistochemical stain patterns help in distinguishing the desmoplastic medulloblastoma from other variants of medulloblastomas.

The previously reported cases of ovarian medulloblastoma contained teratomatous components, and our case was not exceptional. The distinction between ovarian primitive neuroectodermal tumor and immature teratoma is sometimes controversial. By definition, the immature teratoma contains a variable amount of immature neuroectodermal tissues intermingled with other teratomatous elements. Conversely, the neuroectodermal tumors are composed exclusively of primitive neuroectodermal tissue; therefore, they have been separated from teratomas and are treated as a distinct group of neoplasms. The tumor in our case was a single mural mass protruding into the cystic space, and most of it was composed of primitive neuroectodermal tissues that did not mix with other teratomatous elements. This tumor was composed of reticulin-free nodular zones surrounded by densely packed, hypercellular, hyperchromatic cells, which are characteristics of desmoplastic medulloblastomas.

The prognosis for ovarian primitive neuroectodermal tumors depends on the stage of the tumor. Among the 3 previously reported patients with ovarian medulloblastomas, 1 patient was lost to follow-up, 1 passed away 6 months later, and the other was successfully treated with operation and chemotherapy. Although the tumor stage in our patient is IA, extraovarian extension and distant metastasis may still occur. Thus, our patient needs further close follow-up.

In conclusion, we present an exceedingly rare case of an ovarian desmoplastic medulloblastoma arising from a teratoma. Its distinctive histological pattern is the first case reported in the literature.
Table 1. Ovarian Medulloblastomas

<table>
<thead>
<tr>
<th>Age</th>
<th>Tumor Diameter (cm)</th>
<th>Symptoms</th>
<th>Clinical Stage</th>
<th>Nonneural Tissue</th>
<th>Survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kleinman et al</td>
<td>24</td>
<td>17</td>
<td>Mass</td>
<td>IA</td>
<td>Skin</td>
</tr>
<tr>
<td>Kleinman et al</td>
<td>69</td>
<td>11</td>
<td>Mass ascites</td>
<td>III</td>
<td>Respiratory epithelium</td>
</tr>
<tr>
<td>Clinkard et al</td>
<td>23</td>
<td>13</td>
<td>Abdominal pain</td>
<td>IIIC</td>
<td>Epidermis and appendages, fat thyroid tissue</td>
</tr>
<tr>
<td>Wey et al (present study)</td>
<td>26</td>
<td>12</td>
<td>Abdominal fullness</td>
<td>IA</td>
<td>Squamous epithelium, sebaceous gland, choroids plexus</td>
</tr>
</tbody>
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