Case Report
A rare presentation of giant pediatric pilomyxoid astrocytoma involved in the sellar region: case report and literature review

Zhihua Li¹, Mengzhao Feng², Fuyou Guo², Jing Cui¹

¹Department of Parasitology, Medical College, Zhengzhou University, Zhengzhou 450052, Henan, China; ²Department of Neurosurgery, The First Affiliated Hospital, Zhengzhou University, Zhengzhou 450052, Henan, China

Received September 12, 2017; Accepted January 4, 2018; Epub February 15, 2018; Published February 28, 2018

Abstract: Pilomyxoid astrocytoma (PMA) is a rare malignant tumor with a poor prognosis and a high incidence of dissemination in children. We present a case of an unusual giant pediatric PMA with unique clinical imaging. A 13-month-old female presented with three months of nystagmus and developmental retardation. Magnetic resonance imaging (MRI) showed the size of the tumor was approximately 4.5×4.2×4.0 cm³, and there was significant enhancement with an abundant blood supply. Three-dimensional printing (3D-P) of the tumor demonstrated that the bilateral anterior cerebral arteries and the anterior communicating artery were completely wrapped by this giant tumor. Subsequently, the patient underwent total resection with good recovery with the assistance of 3D-P. The presumed diagnosis of the lesion was suprasellar germinoma or craniopharyngioma, however, postoperative histopathological examination identified it as being a rare PMA. Uncommon PMA with unique clinical features should be emphasized as differential diagnosis in the sellar region. 3D-P is a safe and effective tool for successful treatment of a rare hypervascular PMA.

Keywords: Pediatric brain tumor, pilomyxoid astrocytoma, 3D printer, surgery

Introduction
The predominant pediatric glioma located in the sellar region is pilocytic astrocytoma. However, pilomyxoid astrocytoma (PMA) is rare based on previous literature. PMA is a rare WHO grade II tumor, accepted as a variant of pilocytic astrocytoma in the World Health Organization (WHO) classification of central nervous system tumors in 2007 [1]. PMA has previously been described as a solitary rare case in the previous recent literature [2-10] (see Table 1). Here, we report a rare case of PMA with an abundant intratumoral blood supply in the hypothalamic/chiasmatic region. The total resection of the giant PMA was achieved with the assistance of three-dimensional printing (3D-P). Uncommon PMA with unique clinical features should be emphasized as the differential diagnosis in the hypothalamic and chiasmatic region. 3D-P is a safe and effective tool for successful treatment of a rare hypervascular PMA.

Case report
A 13-month-old female was admitted to our hospital with a three-month history of nystagmus. Physical examination was positive for developmental retardation. There was no neurological deficit. A laboratory evaluation revealed the levels of AFP and HCG were 2.57 ng/ml (normal range: 0~10) and less than 0.1 mIU/ml (normal range: 0~5), respectively. A hormonal assay of pituitary function was normal except for a slight decrease in growth hormone in the serum. Computer tomography (CT) revealed the levels of AFP and HCG were 2.57 ng/ml (normal range: 0~10) and less than 0.1 mIU/ml (normal range: 0~5), respectively. A hormonal assay of pituitary function was normal except for a slight decrease in growth hormone in the serum. Computer tomography (CT) revealed a slightly hypodense lesion in the suprasellar region (Figure 1A). Magnetic resonance imaging (MRI) showed a tumor located in the hypothalamic and chiasmatic region and the size of the tumor was approximately 4.5×4.2×4.0 cm³. The tumor appeared hypointense and hyperintense to the brain on T1-weighted (Figure 1B, 1C) and T2-weighted MRI, respectively, with remarkable homogeneous enhancement after administration of gadolinium (Figure 1D-F),
which suggested that the tumor was a hypervascular lesion with an abundant intratumoral blood supply. 3D-P of the tumor was applied in the present patient (Figure 2A-C). It demonstrated the bilateral anterior cerebral arteries and anterior communicating artery were completely wrapped by this giant tumor (Figure 3A-D).

### Table 1. Pediatric pilomyxoid astrocytoma reported in the recent literature

<table>
<thead>
<tr>
<th>No</th>
<th>Author</th>
<th>Year</th>
<th>Age/Sex</th>
<th>Clinical features</th>
<th>Location</th>
<th>Treatment</th>
<th>Prognosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Ceppa E [2]</td>
<td>2007</td>
<td>6.5 Y/Female</td>
<td>Weight gain, irritability, abnormal eye movements</td>
<td>Hypothalamic</td>
<td>Biopsy + chemotherapy</td>
<td>Tumor recurrence after one year</td>
</tr>
<tr>
<td>2</td>
<td>Alimohamadi M [3]</td>
<td>2009</td>
<td>12 Y/Male</td>
<td>Visual impairment, cognitive disturbance</td>
<td>Sellar and suprasellar</td>
<td>Surgery + radiotherapy + chemotheray</td>
<td>No recurrence in follow-up of one year</td>
</tr>
<tr>
<td>3</td>
<td>Paraskevopoulos D [4]</td>
<td>2011</td>
<td>12 Y/Female</td>
<td>Gait disturbance, motor and sensory deficits of the left side</td>
<td>C2-C7 intramedullary</td>
<td>Surgery</td>
<td>Recurrence after 3 M and development of GBM; dead at 12 M</td>
</tr>
<tr>
<td>5</td>
<td>Chonan M [6]</td>
<td>2013</td>
<td>3 Y/Male</td>
<td>Truncal ataxia, drowsy</td>
<td>Right cerebellum</td>
<td>Surgery</td>
<td>Dead</td>
</tr>
<tr>
<td>6</td>
<td>Pereira FO [7]</td>
<td>2013</td>
<td>11 Y/Female</td>
<td>Diplopia, gait disturbance</td>
<td>Brainstem</td>
<td>Surgery</td>
<td>Recurrence after 9 M</td>
</tr>
<tr>
<td>7</td>
<td>Tjahjadi M [8]</td>
<td>2015</td>
<td>7 Y/Female</td>
<td>Headache, nausea, vomiting, bitemporal hemianopia</td>
<td>Suprasellar region</td>
<td>Surgery + radiotherapy</td>
<td>Tumor reduction after one year</td>
</tr>
<tr>
<td>9</td>
<td>Homma T [10]</td>
<td>2017</td>
<td>4 M/Male</td>
<td>Vomiting, slow weight gain</td>
<td>Right temporal lobe + suprasellar region</td>
<td>Surgery + chemo theray</td>
<td>Tumor recurrence after 4 Y</td>
</tr>
<tr>
<td>10</td>
<td>Present case</td>
<td>2017</td>
<td>13 M/Female</td>
<td>Nystagmus, developmental retardation</td>
<td>Hypothalamic + chiasmatic region</td>
<td>Surgery</td>
<td>No recurrence in follow-up of 3 M</td>
</tr>
</tbody>
</table>

**Figure 1.** A 13-month-old female with a PMA in the hypothalamic and chiasmatic region shows the tumor compressing the brain stem on CT (A). A preoperative axial and sagittal T1-weighted MRI scan shows a suprasellar lesion with hypointense signals, respectively (B, C). A coronal axial, sagittal gadolinium-enhanced T1-weighted MRI scan shows a significant homogeneously enhanced tumor (D-F).
The patient underwent resection of the tumor via a frontal basal interhemispheric approach with assistance of the 3D-P model. The lamina terminalis was cut for increased exposure during the operation, and a gross total tumor resection was achieved after one week as seen in CT and MRI examinations (Figure 4A-D). No postoperative radiotherapy or chemotherapy was administered in this patient. During a three-month follow-up, no further tumor progress or recurrence was observed. Histological examination revealed monomorphous bipolar cells with a loose myxoid background (Figure 5A, 5B). Immunohistochemical examination showed the tumor cells were positive for glial fibrillary acidic protein (GFAP), S-100, and Olig2 (Figure 5C-E). Immunohistochemical staining for Ki-67 was performed, and the Ki-67 labeling index was approximately 5% (Figure 5F).

Discussion

PMA is usually implicated in the suprasellar region on the floor of the third ventricle and in the thalamic region. It has been reported that a non-enhancing solid portion of both a primary and disseminated mass have more frequently been found in PMAs compared to PAs (100% versus 32%). The reason for the lack of enhancement may be the myxoid component, based on the previous literature. [11] However, the MRI manifestation of our case was not consistent with previous reports, and there were several striking features obtained from the present patient. First, the tumor was very large due to the length of the largest diameter surpassing 4 cm. Second, the tumor showed remarkably homogeneous enhancement after administration of gadolinium. PMA usually exhibit unremarkable enhancement due to low grade glioma, however, hypervascular PMA with significant enhancement was observed from this patient. The pre-
Rare giant pediatric pilomyxoid astrocytoma with unique clinical imaging

The clinical features of PMAs are that they require a more aggressive course, they have a

sumed diagnosis of the lesion was suprasellar germinoma or craniopharyngioma and postoperative histopathological examination identified it as being a rare PMA. This giant tumor severely compressed the adjacent hypothalamo/chiasmatic vital structures and brain stem. Consequently, it was considerably too dangerous to totally remove such tumors in infants. In our previous cases, there were occasional mortalities with giant suprasellar tumors due to limited exposure and excessive bleeding caused by hypervascular lesions as well as injuries of the hypothalamus, because patients are easily susceptible to shock or even death during surgical intervention because of the very limited blood volume in infants or children, especially for hypervascular tumors. [12] How should the operational risk of hypervascular tumors be reduced in infants? In addition to the basic skills described in our previous paper, we found that a preoperative 3D-P model could offer the best visualization in three-dimensional space, which not only contributed to the design of the preoperative approach but also played an important role in decreasing vascular injury during the operation because bilateral anterior cerebral arteries and the anterior communicating artery were completely wrapped by this giant tumor on 3D-P. The use of 3D-P can facilitate the identification of the 3D anatomical relationship between the tumor and adjacent artery and it is helpful to maximally reduce the damage to the surrounding artery in deep-seated tumors. Through our experience in this case, we found that a 3D-P model is an extremely safe and useful tool for giant PMAs with hypervascularity.

Figure 4. Postoperative CT demonstrated that the tumor was removed fully and there was excellent decompression of the brain stem (A). A postoperative sagittal, axial, and coronal gadolinium-enhanced T1-weighted MRI scan indicates that the tumor was totally removed at 7 days after the operation (B-D).

Figure 5. Hematoxylin-eosin staining demonstrate that the tumor consisted of monomorphous bipolar cells with a loose myxoid background (A: Original magnification ×100 & B: Original magnification ×200). Immunohistochemical examinations for GFAP, S-100, and Olig2, respectively (C-E: Original magnification ×400), and Ki-67 (F: Original magnification ×200).
higher recurrence rate, and they disseminate along the cerebrospinal fluid pathways. Consequently, PMAs are still challenging tumors for neurosurgeons. Moreover, there is no consensus for standard treatment for PMAs until now. Our results show that surgery has a clear role for diagnosis and tumor control as well as relief of the mass effect in optic pathway/hypothalamic gliomas in children. Simultaneously, the authors noted that primary surgical debulking of the tumor without adjuvant therapy was a safe and effective management. [13] Due to the toxicity of chemotherapy or radiotherapy for infants, adjuvant therapy was not recommended because this patient underwent total resection. However, dynamic observation will be very important for the present patient in the future.

Conclusion

PMA is a rare malignant tumor with unique manifestation on MRIs. PMA as low grade glioma exhibited uncommon remarkable enhancement in this patient and 3D-P provided better visualization for observation of the tumor and adjacent vital structures. Total resection of the PMA without complications was achieved with the assistance of 3D-P. Rare PMA with abundant blood supply should be emphasized as differential diagnosis in the hypothalamic and chiasmatic region.

Acknowledgements

This study was supported in part by the Natural Scientific Foundation of China (No. U1204807 and No. 81172612).

Disclosure of conflict of interest

None

Address correspondence to: Jing Cui, Department of Parasitology, Medical College, Zhengzhou University, 40 Daxue Road, Zhengzhou 450052, Henan, China. E-mail: cuij@zzu.edu.cn; Fuyou Guo, Department of Neurosurgery, The First Affiliated Hospital, Zhengzhou University, Zhengzhou 450052, Henan, China. E-mail: chyou666@hotmail.com

References


