Pilocytic Astrocytoma Arising from the Conus Medullaris in an Adult: A Case Report

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Abstract

Low-grade, sporadic, pilocytic astrocytomas (PAs) are rare spinal cord tumors diagnosed in adult patients. Their localization to the conus medullaris is exceedingly rare, having only been described in a limited number of case reports. Here, we describe a case of a 22-year-old female presenting with back pain, lower extremity weakness, hypoesthesia, and urinary incontinence. Imaging studies demonstrated a cystic lesion of the conus medullaris that was treated with subtotal resection and cyst-subarachnoid shunt placement. Final pathology report confirmed PA from the histology of surgical specimens. We discuss the current literature of conus medullaris lesions and their differential diagnosis.

Keywords
► pilocytic astrocytoma
► conus medullaris

Introduction

Pilocytic astrocytomas (PAs) are one of the most common primary central nervous system tumors in the pediatric population, accounting for 11.4% of primary central nervous system tumors, with an average incidence of 1.03 per 100,000.1 In contrast, PAs have a much lower incidence in adults, accounting for only 0.6% of primary brain tumors. In adults, PAs carry a worse prognosis; the 10-year survival rate for patients over 40 is 76.1% compared with 96% in children.1 Additionally, adult PAs are frequently located in the supratentorial region,2 with proclivity for the temporal and parietal lobes.3,4 In children, however, they are preferentially localized to the infratentorial compartment.5,6 There are rare case reports of primary spinal PAs presenting in adult patients,7-11 and a larger case series identified a potential predilection for the cephalad/superior regions of the spinal cord.12 PAs localized to the conus medullaris are diagnosed even less frequently, with only three documented and published reports, to the best of the authors’ knowledge.13-15 Therefore, clear documentation of the diagnosis and treatment strategies will help provide informed management of future patients presenting with conus medullaris PAs.

As noted previously, adult patients with PAs have a lower survival compared with pediatric patients and factors negatively impacting survival include subtotal resection or biopsy (compared with gross total resection), radiation treatment,
Case Report

Clinical Presentation
A 22-year-old female, with pertinent history of chronic back pain radiating to the right thigh and groin, presented to the emergency department with worsening pain and episodes of bowel incontinence. Her back pain started 2 years ago, and she was initially prescribed physical therapy alone that kept her symptoms stable until 4 months prior to presentation. On physical exam, she was noted to have hypoesthesia and weakness in her both lower extremities, specifically with more proximal weakness of 4/5 hip flexion, 4/5 knee extension, and 4/5 dorsiflexion.

Neuroimaging
The patient underwent magnetic resonance imaging (MRI) of the neuroaxis that demonstrated a 70 × 14 × 18 mm cystic intramedullary mass extending from the T11 to the conus tip. The mass demonstrated septations with fluid filled compartments that were hyperintense to cerebrospinal fluid (CSF) on T1 and isointense on T2-weighted MRI images. Contrast enhancement was noted centrally in the lesion (Fig. 1). No additional neuroaxis lesions were noted.

Surgical Treatment and Postoperative Course
The patient underwent a decompressive laminectomy from T11 to L1 for exposure. A durotomy was performed and the spinal cord exposed, noting a bluish hue to the suspected tumor site (Fig. 2). Intraoperative ultrasound was used to localize the thinnest part of the spinal cord in an effort to minimize injury to neural tissue from the myelotomy required to enter the tumor/cyst cavity. Once within the cavity, there was release of xanthochromic fluid, under moderate pressure. Multiple biopsies were taken from the cyst wall and pathologic examination of frozen sections indicated low-grade glioma. As there was no clear delineation between tumor and normal neural tissue, a subtotal resection was necessary to avoid neurologic injury. A cyst–subarachnoid shunt was placed to prevent future reaccumulation of cyst fluid (Fig. 2; Supplementary Video 1 [available in the online version only]). Histopathology of the tumor confirmed the diagnosis of PA demonstrating Rosenthal fibers, glial fibrillary acidic protein positivity, IDH1 wild-type, attenuated p53 expression, and a MIB-1 proliferation index less than 1% (Fig. 3). Her postoperative MRI confirmed subtotal resection with collapse of the cyst cavity and a residual area of central contrast-enhancement (Fig. 1). At her 1-month postoperative visit, the patient reported some residual saddle anesthesia; however, there was resolution of paresthesia and weakness in both legs and no episodes of urinary incontinence. Her recommended treatment plan was surveillance imaging in 6 months and then yearly to monitor for either tumor recurrence and or cyst formation.

Discussion
PA of the spine is a rare diagnosis to make, especially in the adult population where they account for less than 1% of central nervous system tumors. The patient described here has an even more unusual presentation with the tumor arising from the conus medullaris. To the authors knowledge, only three other reports of sporadic low-grade conus PAs have been described (Table 1). In all cases, these tumors presented with back pain, lower extremity weakness, and hypoesthesia. Bladder or bowel dysfunction was noted in two of the four cases. MRI findings demonstrated a cystic mass that is hypointense on T1 and hyperintense on T2 compared with CSF and enhances with contrast. Treatment is surgical resection with the goal to preserve function. Communicating the cystic component to the subarachnoid space was done in our case, using a shunt, and in one other case through marsupialization. Due to the benign course of PAs, postoperative adjuvant treatment is not necessary, and surveillance imaging should be done to monitor cyst fluid accumulation or tumor progression.

Additional cases of astrocytomas of the conus medullaris have been described: a case of anaplastic glioma of the conus medullaris with morphological features of PA, and an astrocytoma of the conus in a patient with neurofibromatosis type 1. In both cases, these tumors presented with similar symptoms of low back pain and saddle anesthesia; however, both required postoperative adjuvant treatment due to aggressive nature of high-grade lesions. In addition, there are rare reports of adult holocord astrocytomas extending into the conus medullaris as well, often accompanied by motor and sensory dysfunction in all extremities given the extent of the tumor involvement.

The differential diagnosis of intramedullary tumors of the conus medullaris includes PA as well as ependymomas. Ependymomas are much more common, accounting for 25% of intramedullary spinal cord tumors in adults. They are typically low-grade benign tumors that can be treated with surgical resection alone. Ependymomas are most...
Fig. 1 Neuroradiology of a conus pilocytic astrocytoma. T1 (pre- and postcontrast administration) and T2-magnetic resonance imaging (MRI) of the lumbar spine demonstrating a large cystic lesion in the conus region of the spine extending to T11. The lesion demonstrates heterogeneous enhancement centrally. On T2-axial cuts, the cystic component obliterates the spinal cord in this region. Postoperatively, residual tumor is noted with a central contrast-enhancing lesion on T1, but with collapse of the cystic components.

Fig. 2 Intraoperative view of pilocytic astrocytoma resection. T11 to L1 laminectomy was performed to expose the dura and underlying spinal cord. Myelotomy was performed with a release of xanthochromic fluid, and a cyst-subarachnoid shunt was placed for continual drainage of the cyst into the cerebrospinal space.
Histological characterization of resected conus medullaris pilocytic astrocytoma. Hematoxylin and eosin staining demonstrates a tumor composed of piloid astrocytes with Rosenthal fibers (arrow). Eosinophilic granular cell bodies are not present in appreciable numbers. There is no evidence of necrosis, significant mitotic activity, or endothelial proliferation. Immunohistochemical stains for glial fibrillary acidic protein (GFAP) confirm the astrocytic origin of the tumor. Stains for mutant IDH1R132H were negative and total p53 expression was very low. Stains for MIB-1, a marker of cellular proliferation, indicate a proliferative index that is approximately 1%.

### Table 1 Additional reports of adult astrocytomas localized to the conus medullaris

<table>
<thead>
<tr>
<th>Study</th>
<th>Year published</th>
<th>Presentation</th>
<th>Radiology findings</th>
<th>Histopathologic diagnosis</th>
<th>Treatment course</th>
</tr>
</thead>
<tbody>
<tr>
<td>Baréa et al14</td>
<td>1999</td>
<td>20-year-old female. Lower back pain with radiation to lower extremities.</td>
<td>Cystic mass, hypointense on T1 and hyperintense on T2 to CSF</td>
<td>Low-grade PA</td>
<td>GTR, no adjuvant treatment</td>
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<tr>
<td>Kumar et al13</td>
<td>2012</td>
<td>44-year-old female. Lower back pain with radiation to bilateral lower extremities. Hypoesthesia and low voiding pressures on physical exam.</td>
<td>Cystic mass, hypointense on T1 and hyperintense on T2 to CSF, peripheral enhancement with contrast</td>
<td>Low-grade PA</td>
<td>GTR, no adjuvant treatment</td>
</tr>
<tr>
<td>Lavrador et al15</td>
<td>2017</td>
<td>69-year-old female. Gait disturbances and bilateral lower extremity pain Hypoesthesia on physical exam.</td>
<td>Cystic mass, hypointense on T1 and hyperintense on T2 to CSF</td>
<td>Low-grade PA</td>
<td>STR, cyst marsupialization into subarachnoid space</td>
</tr>
<tr>
<td>Uchi et al26</td>
<td>2020</td>
<td>69-year-old male with NF1, back pain and bilateral hypoesthesia of the lower extremities.</td>
<td>Cystic mass, hypointense on T1 and hyperintense on T2 to CSF, peripheral enhancement with contrast</td>
<td>Grade 2 diffuse astrocytoma</td>
<td>Laminectomy and biopsy followed by radiation. Patient died after 1 year and 10 months</td>
</tr>
<tr>
<td>Palpan Flores et al25</td>
<td>2021</td>
<td>20-year-old male. Back pain, paraparesis, and urinary incontinence.</td>
<td>Cystic mass, hypointense on T1 and hyperintense on T2 to CSF, enhancement with contrast</td>
<td>Anaplastic astrocytoma features, H3K27 mutant glioma</td>
<td>STR, Stupp protocol (radiotherapy and temozolomide)</td>
</tr>
<tr>
<td>Current case</td>
<td>NA</td>
<td>22-year-old female</td>
<td>Cystic mass, hypointense on T1 and hyperintense on T2 to CSF, peripheral and central enhancement with contrast</td>
<td>Low-grade PA</td>
<td>STR, cyst to subarachnoid space shunt</td>
</tr>
</tbody>
</table>

Abbreviations: CSF, cerebrospinal fluid; GTR, gross total resection; NF1, neurofibromatosis 1; PA, pilocytic astrocytoma; STR, subtotal resection.
frequently found in the cervical and upper thoracic spine, but myxopapillary ependymomas can be found in the conus medullaris.29

The management in our patient included subtotal resection in an effort to reduce functional loss associated with possible resection of normal neural tissue within the spinal cord. Additionally, a cyst−subarachnoid shunt was placed to allow for continual drainage of the cyst and prevent future cord compression secondary to cyst reaccumulation and expansion. A potential complication to this approach is seeding the rest of the neuroaxis with tumor cells; however, widespread metastatic central nervous system disease from a conus tumor would only be expected in high-grade lesions.30 Seeding of distal areas from shunt placement has been described as a rare but serious complication in pediatric patients with CSF shunts and high-grade tumors.31,32

Conclusion

Adult PA localized to the conus medullaris are exceedingly rare and typically present with lower backpain, lower extremity weakness, hypoaesthesia, and incontinence. Given the low recurrence and slow growth rate of these tumors, treatment should be aimed at preserving function through surgical resection and postoperative surveillance imaging.

Informed Consent

The authors received informed consent for publication from the patient described in this case.

Funding
None.

Conflicts of Interest
None declared.

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