



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

Nathan K. Leclair¹ Avery Buehler² Qian Wu³ Kevin Becker⁴ Isaac L. Moss⁵ Ketan R. Bulsara⁴
Hilary Onyiuke⁴

¹School of Medicine, University of Connecticut, Farmington, Connecticut, United States

²Research Assistant, University of Connecticut, Storrs, Connecticut, United States

³Department of Pathology and Laboratory Medicine, UConn Health, Farmington, Connecticut, United States

⁴Department of Surgery, Division of Neurosurgery, UConn Health, Farmington, Connecticut, United States

⁵Department of Orthopedic Surgery, UConn Health, Farmington, Connecticut, United States

Address for correspondence Hilary Onyiuke, MD, FRCSC, FACS, FAANS, Department of Neurological Surgery, University of Connecticut Health Center, 263 Farmington Avenue, Farmington CT 06030, United States (e-mail: onyiuke@uchc.edu).

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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,

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and presenting location other than the cerebellum.¹⁶ Given their low-grade nature, many PAs can be managed with surgery and postoperative observation alone,¹⁷ and while subtotal resection can be a strong predictor of tumor recurrence,^{18,19} gross total resections must be balanced by the risk of damaging eloquent neural tissue. In patients that decline surgery or where surgery is not feasible, radiation can be used for tumor control,^{17,20} although some studies indicate that radiation and chemotherapy are associated with lower overall survival in PAs compared with surgery or observation alone.²¹ Newer agents, such as bevacizumab, have been used to manage recurrent PAs in adults.^{22,23} This treatment approach is in contrast to high-grade astrocytomas of the spine, which in adults carry a poor prognosis and require aggressive surgical treatment and postoperative adjuvant therapy.²⁴

Here, we describe a case of an adult presenting with a PA of the conus medullaris. We use this case to describe our experience in the diagnosis and management strategies for this unusual presentation. The patient has consented to publication of this report.

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.¹ 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 described¹³⁻¹⁵ (►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.¹⁵ 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,²⁵ and an astrocytoma of the conus in a patient with neurofibromatosis type 1²⁶ (►Table 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.^{8,27}

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.²⁸ They are typically low-grade benign tumors that can be treated with surgical resection alone. Ependymomas are most

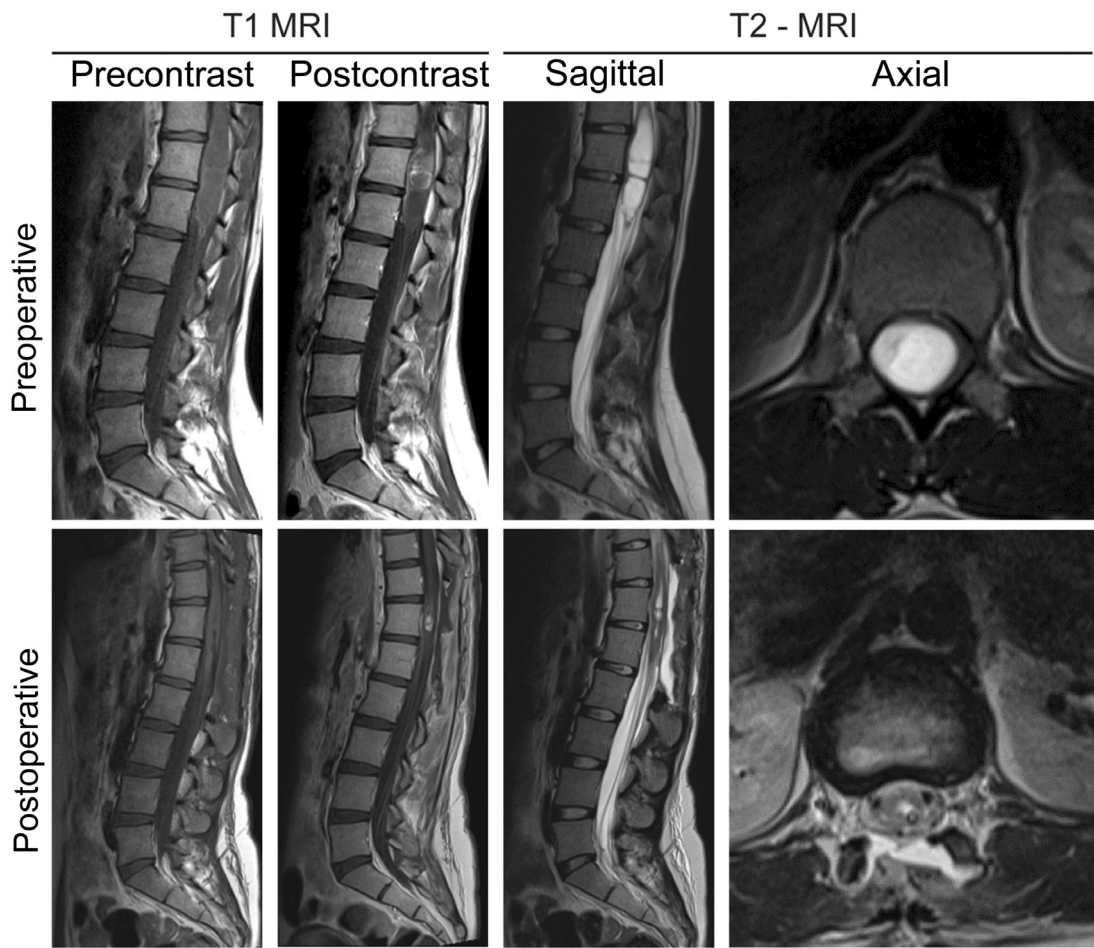


Fig. 1 Neuroradiology of a conus pilocytic astrocytoma. T1 (pre- and postcontrast administration) and T2-magnetic resonance imaging (MRI) of the lumbar spine demonstrating a large cystic lesion in the conus region of the spine extending to T11. The lesion demonstrates heterogenous enhancement centrally. 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.

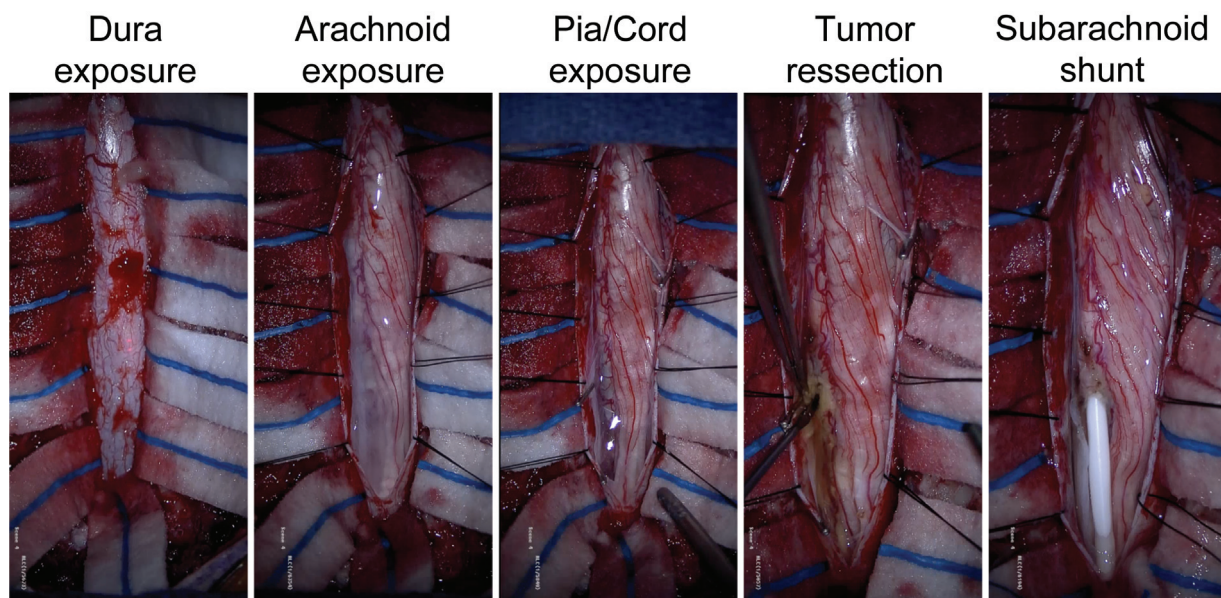


Fig. 2 Intraoperative view of pilocytic astrocytoma resection. T11 to L1 laminectomy was performed to expose the dura and underlying spinal cord. Myelotomy was performed with a release of xanthochromic fluid, and a cyst-subarachnoid shunt was placed for continual drainage of the cyst into the cerebrospinal space.

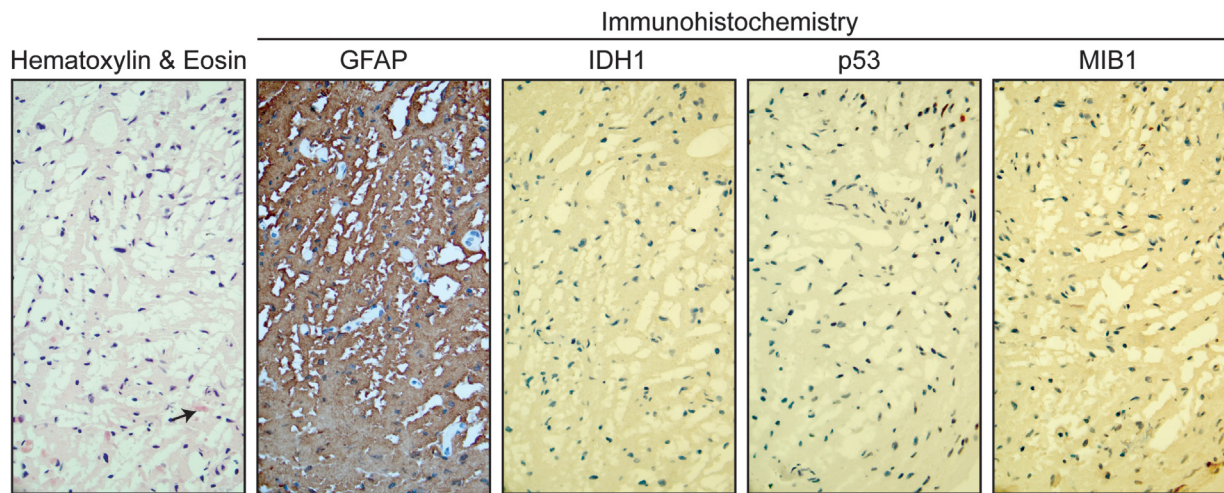


Fig. 3 Histological characterization of resected conus medullaris pilocytic astrocytoma. Hematoxylin and eosin staining demonstrates a tumor is composed of piloid astrocytes with Rosenthal fibers (*arrow*). Eosinophilic granular cell bodies are not present in appreciable numbers. There is no evidence 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 IDH1^{R132H} 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

Study	Year published	Presentation	Radiology findings	Histopathologic diagnosis	Treatment course
Baréa et al ¹⁴	1999	20-year-old female. Lower back pain with radiation to lower extremities. Saddle anesthesia.	Cystic mass, hypointense on T1 and hyperintense on T2 to CSF	Low-grade PA	GTR, no adjuvant treatment
Kumar et al ¹³	2012	44-year-old female. Lower back pain with radiation to bilateral lower extremities. Hypoesthesia and low voiding pressures on physical exam.	Cystic mass, hypointense on T1 and hyperintense on T2 to CSF, peripheral enhancement with contrast	Low-grade PA	GTR, no adjuvant treatment
Lavrador et al ¹⁵	2017	69-year-old female. Gait disturbances and bilateral lower extremity pain. Hypoesthesia on physical exam.	Cystic mass, hypointense on T1 and hyperintense on T2 to CSF	Low-grade PA	STR, cyst marsupialization into subarachnoid space
Uchi et al ²⁶	2020	69-year-old male with NF1, back pain and bilateral hypoesthesia of the lower extremities.	Cystic mass, hypointense on T1 and hyperintense on T2 to CSF, peripheral enhancement with contrast	Grade 2 diffuse astrocytoma	Laminectomy and biopsy followed by radiation. Patient died after 1 year and 10 months
Palpan Flores et al ²⁵	2021	20-year-old male. Back pain, paraparesis, and urinary incontinence.	Cystic mass, hypointense on T1 and hyperintense on T2 to CSF, enhancement with contrast	Anaplastic astrocytoma features, H3K27 mutant glioma	STR, Stupp protocol (radiotherapy and temozolomide)
Current case	NA	22-year-old female	Cystic mass, hypointense on T1 and hyperintense on T2 to CSF, peripheral and central enhancement with contrast	Low-grade PA	STR, cyst to subarachnoid space shunt

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, hypoesthesia, 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.

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None.

Conflicts of Interest

None declared.

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