Case report

Dorsal intramedullary spinal pilocytic astrocytoma: a case report and review of published cases

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ABSTRACT

Background: Spinal pilocytic astrocytoma (PA) is an uncommon intramedullary neoplasm, with the literature dominated by isolated case reports and a few small series.

Case presentation: This report describes a 10-year-old boy with a D9–D12 intramedullary solid-cystic PA treated with gross total resection, followed by a focused review of published spinal PA cases and series, emphasizing epidemiology, radiological characteristics, surgical strategy, and outcomes.

Conclusion: Excellent early neurological recovery can be achieved when a safe plane of dissection is present and careful microsurgical technique is used.

Key words: pilocytic astrocytoma, spine, spinal tumor

INTRODUCTION

Pilocytic astrocytoma is a World Health Organization (WHO) grade 1 glioma that typically arises in the cerebellum, optic pathways, hypothalamus, and brainstem in children and young adults. Spinal localization is distinctly rare, accounting for a small minority of pediatric low-grade gliomas and intramedullary tumors overall. Because of this rarity, most knowledge of spinal PA is extrapolated from intracranial disease, while available spinal data derive from single-center series, multicenter registries, and case reports [1–6]. The present case describes a dorsolumbar intramedullary PA in a 10-year-old boy treated at a tertiary neurosurgical center in North India, with classic imaging and histopathology but an uncommon holosegmental dorsal–lumbar extent and good functional recovery following gross total excision. A narrative review of the literature is provided, consolidating data from recent series and representative case reports to contextualize this presentation [1–4, 7].

CASE PRESENTATION

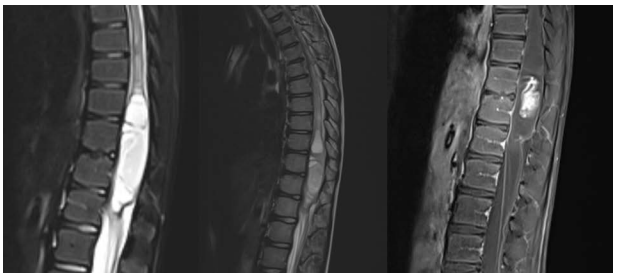
Clinical presentation

A previously healthy 10-year-old boy presented with several weeks of low backache and progressive difficulty in walking, while bowel and bladder function remained intact. There was no history of trauma, infection, prior surgery, or significant family illness. On neurological examination he was alert with normal cranial nerve and upper-limb function, but had left lower-limb weakness maximal distally, reduced left knee and ankle jerks, and preserved right-sided strength and tone.

Preoperative imaging

Contrast-enhanced MRI of the dorsolumbar spine (fig. 1) demonstrated a relatively well-defined expansile intramedullary solid-cystic lesion extending from D9 to D12, involving the lower dorsal cord and conus.

Figure 1. Contrast-enhanced MRI of the dorsolumbar spine demonstrated a relatively well-defined expansile intramedullary solid–cystic lesion extending from D9 to D12, involving the lower dorsal cord and conus.



The lesion showed an avidly enhancing nodular solid component with internal cystic locules, visible fluid–fluid levels, and a thin peripheral rim suggestive of hemorrhagic by-products, along with a small syrinx at D8 and pressurised intramedullary hyperintensity extending cranially to D6–D7. Vertebral alignment and osseous structures were normal, with no abnormal enhancement of the vertebral bodies or epidural space, and the remaining spinal cord segments appeared unremarkable.

Operative details

After multidisciplinary discussion and informed consent, the child underwent D9–D12 laminoplasty with excision of the intramedullary lesion under general anesthesia. In the prone position, a midline dorsal incision and subperiosteal muscle dissection exposed D9–D12 laminae, and a "scorpion-tail" laminoplasty was fashioned to allow decompression while preserving posterior elements. The dura was bulging and tense; following midline durotomy, the cord was noted to be rotated slightly to the right with bilateral nerve roots in view.

A midline myelotomy over the point of maximal expansion exposed a yellowish-grey, moderately vascular, CUSA-suckable intramedullary tumor with relatively ill-defined margins blending with adjacent cord parenchyma. Internal debulking with the ultrasonic aspirator, complemented by microsurgical dissection along any identifiable plane, permitted circumferential mobilization and apparent gross total excision while respecting pial boundaries and rootlets. After meticulous hemostasis, the dura was closed in a watertight fashion reinforced with fibrin sealant, the laminae were replaced to complete the laminoplasty, and layered wound closure was performed over a drain.

Histopathology and immunohistochemistry

Microscopic examination of the tumor showed a biphasic architecture with compact piloid areas composed of bipolar spindle cells with elongated hair-like processes and more microcystic regions containing loose, pale, stellate cells. Numerous Rosenthal fibers and eosinophilic granular bodies were seen, without significant mitotic activity, necrosis, or microvascular proliferation, consistent with WHO grade 1 pilocytic astrocytoma. Representative hematoxylin–eosin stained sections from this tumor are illustrated below [6].

Immunohistochemistry demonstrated diffuse GFAP positivity and a low proliferative index, with Ki-67 labeling restricted to a small fraction of tumor cell nuclei, while ATRX expression was retained and p53 showed only scattered weak nuclear staining. This morphologic and immunophenotypic profile supported the diagnosis of classic pilocytic astrocytoma without anaplastic features. A composite panel of the immunostains is shown [6].

Postoperative course and follow-up

The child was extubated on table and transferred to the neurosurgical ward for close monitoring. He received perioperative antibiotics, analgesia, and a short course of high-dose methylprednisolone, and the postoperative period remained uneventful without cerebrospinal fluid leak, wound complication, or new neurological deficit. Over the first postoperative week he demonstrated progressive improvement in left lower-limb power, with knee and ankle strength improving to near-normal, and he was mobilised with assistance.

He was discharged in a satisfactory condition, ambulatory, afebrile, and with a healthy wound, on a tapering steroid regimen, oral antibiotics, and analgesics, together with advice regarding wound care, physiotherapy, and scheduled neurosurgical follow-up. Interval MRI is planned to document the extent of resection and to monitor for recurrence, given the known potential for late progression and leptomeningeal dissemination in a minority of spinal PAs [4–8].

REVIEW OF THE LITERATURE

Search strategy and scope

Published data on spinal pilocytic astrocytoma remain limited; most series combine both adult and pediatric patients and include only small numbers of cases. For this review, emphasis was placed on recent comprehensive series and illustrative case reports that together capture the clinical spectrum of intramedullary spinal PAs, including a 12-patient single-center series with literature review by Hu et al., a multicenter study by the Neurospinal Society of Japan, a radiologic series detailing MRI features, a molecular characterization study of pediatric spinal PAs, and selected case reports of multicentric, holocord, and disseminated disease [1–4, 7–10].

Because of heterogeneity in reporting and the absence of a dedicated meta-analysis that enumerates every individual case, the compiled data presented here draw primarily from these higher-yield publications rather than claiming absolute exhaustiveness.

Epidemiology and demographics

Spinal PA accounts for a minority of intramedullary spinal cord gliomas, with ependymoma and diffuse astrocytoma more frequent in adults. In the multicenter Japanese series of primary spinal PA, the cohort included both children and adults, with a median age in young adulthood but a substantial pediatric subset. The MRI-based series from Huashan hospital identified 13 consecutive patients with pathologically confirmed spinal PA from a neuro-oncology database, again spanning a wide age range but showing a predilection for younger patients [3, 4, 7, 11].

Pediatric spinal low-grade gliomas and glioneuronal tumors, including PA, constitute approximately 2.8–5.2% of pediatric low-grade gliomas overall, underscoring the rarity of spinal localization even in this age group. Several reports highlight unusual extremes, such as holocord PAs in children, multicentric spinal PAs, and long-delayed spinal leptomeningeal dissemination from an initial intracranial PA treated in childhood [2, 7, 9, 10, 12].

Anatomical distribution and radiological characteristics

Across series, spinal PAs most often arise in the cervical and thoracic cord, with fewer lesions in the conus–cauda region, and some tumors spanning multiple contiguous segments. Holocord involvement from the cervicomedullary junction to the conus has been described, mainly in children, and presents formidable challenges for surgical management [1, 3, 4, 9, 13, 14].

Typical MRI features include an eccentric intramedullary mass with mixed solid-cystic architecture, often associated with syringomyelia; in the Huashan cohort, all 13 tumors showed solid-cystic components, and a majority had associated syrinx. The solid nodule usually enhances vividly after contrast, while the cystic component may demonstrate fluid–fluid levels or internal septations, and diffusion-weighted imaging often reveals relatively high apparent diffusion coefficient values compared with normal cord, reflecting the low-grade nature of the lesion. These features overlap with spinal ependymoma and diffuse astrocytoma, and misdiagnosis as ependymoma or inflammatory myelitis remains common preoperatively [1, 3, 7].

Histopathology and molecular profile

Histologically, spinal PAs recapitulate the classic biphasic piloid and microcystic pattern seen in their intracranial counterparts, with variable numbers of Rosenthal fibers and eosinophilic granular bodies and generally low mitotic activity. Anaplastic transformation is rare but has been documented, including an adult primary intramedullary spinal cord PA with anaplastic features and higher Ki-67 index [1, 3, 6, 15].

Molecularly, pediatric spinal PAs have been shown to form a distinct epigenetic subclass, separate from PAs in other CNS locations and from diffuse leptomeningeal glioneuronal tumor (DLGNT). In that series, FGFR1 alterations were enriched in spinal PAs, whereas KIAA1549:BRAF fusions were more characteristic of cerebellar and optic pathway PAs, supporting the concept of location-specific oncogenic drivers. Recognition of this molecular distinction is important to avoid misclassification of spinal low-grade gliomas and to guide targeted therapy in selected progressive cases [2, 6] (fig. 2, 3).

Figure 2. Histopathology of dorsal spinal pilocytic astrocytoma (D9–D12, intramedullary lesion). (A) Low power hematoxylin–eosin (H&E) section showing a moderately cellular intramedullary glial tumour composed of bipolar piloid cells in a fibrillary background without necrosis or significant atypia. (B) Higher magnification highlighting loose microcystic and myxoid areas with scattered eosinophilic granular bodies and Rosenthal type fibres, typical of pilocytic astrocytoma. (C) Tumour with entrapped thick walled hyalinised vessels and surrounding microcystic change, without endothelial proliferation or microvascular tufting. (D) Compact nodule of piloid spindle cells arranged in whorled fascicles, further supporting the diagnosis of pilocytic astrocytoma.

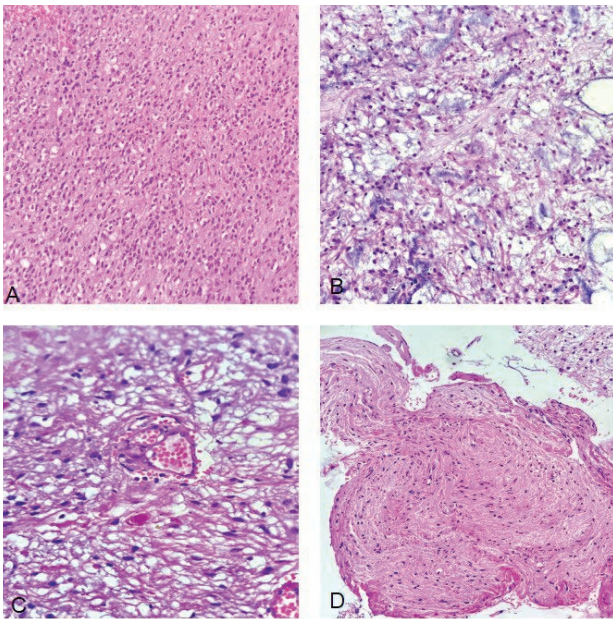
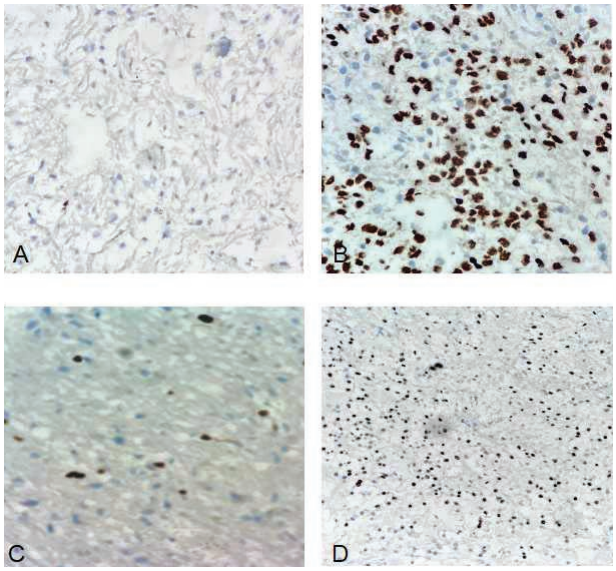


Figure 3. Immunohistochemical profile of spinal pilocytic astrocytoma. (A) Epithelial membrane antigen (EMA) immunostain showing absence of membranous or perinuclear dot like staining in tumour cells, arguing against ependymoma. (B) Olig2 immunostain with strong, diffuse nuclear positivity in tumour cells, confirming glial/astrocytic lineage. (C) Ki 67 (MIB 1) immunostain demonstrating only scattered positive nuclei, with a low proliferative index (<3%), in keeping with a WHO grade 1 lesion. (D) Glial fibrillary acidic protein (GFAP) showing diffuse strong positivity of tumour cells and their processes, supporting the diagnosis of pilocytic astrocytoma.



TREATMENT STRATEGIES

Surgery is the mainstay of treatment for primary spinal PA, with the goal of maximal safe resection while preserving neurologic function. In the Japanese multicenter study, gross total resection was achieved in a subset of patients and was associated with lower local recurrence, although functional outcome was primarily related to preoperative neurological status and intraoperative monitoring findings [1, 3, 4].

Because PAs are often relatively well circumscribed compared with diffuse astrocytomas, an identifiable plane between tumor and normal cord may permit near-total or gross total excision, particularly in focal cervical or thoracic lesions; however, infiltrative behavior and adherence to critical tracts can limit resectability in long-segment or holocord tumors. Internal debulking with ultrasonic aspirator, meticulous pial preservation, and use of intraoperative neurophysiological monitoring are recommended to optimize the balance between extent of resection and functional preservation [1, 4, 9, 14].

Adjuvant radiotherapy or chemotherapy is generally reserved for subtotal resection, recurrence, leptomeningeal dissemination, or histologic anaplasia. Pediatric protocols for low-grade glioma, often including carboplatin and vincristine-based regimens, have been used successfully for disseminated or unresectable spinal PAs, achieving radiologic response and prolonged disease control [2, 4, 6, 8, 12].

Outcomes and prognostic factors

Overall survival for spinal PA is favorable compared with higher-grade spinal astrocytomas, but functional outcome and progression-free survival vary with age, preoperative deficits, tumor length, and extent of resection. In the Neurospinal Society of Japan study, most patients were alive with stable or improved neurological status at long-term follow-up, although some experienced late recurrence necessitating further treatment [4, 9].

Case reports of holocord and multicentric spinal PAs suggest that even extensive disease can be compatible with long survival, but the risk of neurological morbidity is substantial, and symptom control rather than radiologic cure may be the realistic surgical goal. Leptomeningeal dissemination, either synchronous at diagnosis or metachronous many years after initial treatment, is rare

but underscores the need for long-term surveillance with craniospinal imaging in selected patients, especially those with progressive symptoms or atypical imaging [7–10, 12, 14].

Summary of published series and illustrative cases

The table 1 below synthesizes key features from major recent series and representative case reports to provide context for the present case.

by pediatric spinal tumor surgery literature. Midline myelotomy through the dorsal median sulcus, judicious use of CUSA for internal debulking, and progressive circumferential dissection permitted gross total excision despite the tumor's moderate vascularity and somewhat indistinct plane from the surrounding cord [4, 9, 14].

Histologically, the tumor exhibited quintessential pilocytic features without anaplasia, and immunohistochemistry showed

Table 1. Summary of reported cases of primary spinal pilocytic astrocytoma in the literature and the present case, showing patient demographics, tumour location and extent, radiological characteristics, surgical strategy, adjuvant treatment, and outcome (follow-up duration and recurrence status).

Stud/report	Design and cohort	Key findings relevant to spinal PA
Hu et al. [1]	Single-center retrospective series of 12 spinal PAs (2010–2021) with literature review	Highlighted rarity, frequent preoperative misdiagnosis as ependymoma, predominance of cervical–thoracic lesions, solid-cystic MRI appearance, and good prognosis after surgery with low-grade histology.
Métais et al. [2]	Molecular and epigenetic study of 28 pediatric spinal low-grade gliomas and glioneuronal tumors	Demonstrated that pediatric spinal PAs form a distinct methylation and genetic subclass enriched for FGFR1 alterations, separable from PAs of other locations and DLGNT.
She et al. [3]	Imaging series of 13 consecutive spinal PAs	Described consistent solid-cystic intramedullary masses, frequent syringomyelia, eccentric location, vivid nodular enhancement, and high ADC values aiding differentiation from other intramedullary tumors.
Muto et al. [4]	Multicenter retrospective study of primary spinal PAs in Japan	Analyzed clinical characteristics and long-term surgical outcomes; gross total resection associated with lower recurrence; preoperative neurologic status predicted postoperative function.
Alanazi et al. [7]	Adult case of multicentric spinal PA with syringomyelia	Reported multiple mural nodules within a cervical syrinx; emphasized rarity of multicentric disease and role of meticulous microsurgical excision.
Holocord PA reports [9, 10, 13, 14]	Case reports and small pediatric series of holocord PAs	Highlighted challenges of managing long-segment tumors; strategy often combines limited debulking, cyst fenestration, and adjuvant therapy with focus on preserving function.
Leptomeningeal dissemination reports [8, 12]	Pediatric and adult cases of spinal or craniospinal dissemination of PA	Demonstrated that PA, although low-grade, can disseminate along neuraxis after long latency; management typically involves chemotherapy and/or radiotherapy with prolonged follow-up.

Cx – chemotherapy; GTR – gross total resection; MMCS – modified McCormick scale; PA – pilocytic astrocytoma; PR – partial resection; RT – radiotherapy; STR – subtotal resection.

DISCUSSION

Uniqueness and learning points from the present case

This case illustrates a classic solid-cystic dorsolumbar intramedullary PA in a child, but with certain features that make it instructive for neurosurgeons and neuroradiologists. The lesion extended over multiple segments (D9–D12) with associated syrinx and cranial pressurised cord signal change, closely mimicking more common intramedullary tumors such as ependymoma or diffuse astrocytoma on clinical grounds. Preoperative MRI demonstrated the characteristic avidly enhancing mural nodule with internal cystic locules and fluid–fluid levels, imaging clues that, when correlated with the patient's age, could raise a strong suspicion of PA and potentially influence counselling and surgical planning [1, 3, 4].

From a technical perspective, laminoplasty rather than laminectomy was chosen to reduce the risk of postoperative spinal deformity in this skeletally immature patient, an approach supported

a low proliferative index, suggesting a favorable prognosis; this aligns with data indicating that most spinal PAs behave indolently when completely resected, although rare cases of anaplastic transformation or late dissemination mandate continued vigilance [4, 12, 15].

Comparison with published series

When compared with aggregated data from recent series, the current case fits within the expected spectrum of spinal PAs in terms of age, thoracic location, solid-cystic architecture, and association with syrinx (tab. 1). Presentation with progressive gait difficulty but preserved sphincter function and subsequent improvement in motor power following gross total resection mirrors the favorable functional trajectories observed in many reported pediatric patients [1, 3, 4, 9].

However, the presence of a relatively long-segment D9–D12 lesion places this tumor towards the more extensive end of the spec-

trum, approaching the holocord examples that often demand staged or limited debulking; in that sense, this case underscores that even multisegment intramedullary PAs may be amenable to complete excision when a plane exists and meticulous microsurgical technique is used. The use of laminoplasty and preservation of posterior elements also addresses a critical concern in growing children – postlaminectomy kyphosis – highlighting the importance of tailoring spinal approaches to skeletal maturity [4, 9, 14].

Practical implications for diagnosis and management

The literature suggests that in a child or young adult with a long-segment, eccentric, solid-cystic intramedullary spinal cord mass with a vividly enhancing mural nodule and associated syrinx, PA should be strongly considered in the differential diagnosis, even when ependymoma appears more likely on epidemiologic grounds. Recognizing this possibility can inform preoperative counselling regarding prognosis and the likelihood of achieving gross total resection [1, 3, 7].

For surgeons, intraoperative assessment of tumor consistency and the presence of a cleavage plane should guide the extent of resection; aggressive pursuit of gross total excision is reasonable when neuromonitoring signals remain stable and a plane is evident, but subtotal resection may be appropriate in infiltrative or holocord disease to avoid permanent deficits. In cases with subtotal resection or disseminated disease, multidisciplinary discussion with neuro-oncology is essential to select appropriate adjuvant chemotherapy or radiotherapy based on age, molecular profile, and prior treatments [2, 4, 8, 9, 12, 14].

Long-term follow-up with serial MRI is mandatory, given reports of late recurrence and very delayed leptomeningeal spread, sometimes decades after initial surgery. Counselling families about this low but real risk, even in the context of a histologically benign lesion, sets realistic expectations and encourages adherence to surveillance protocols [6, 8, 12, 16–18].

CONCLUSION

Spinal pilocytic astrocytoma is a rare but important consideration in the differential diagnosis of pediatric intramedullary spinal cord tumors, particularly when MRI shows an eccentric, solid-cystic lesion with a vividly enhancing mural nodule and associated syrinx. The present case of a D9–D12 intramedullary PA in a 10-year-old boy, managed with laminoplasty and gross total excision, illustrates that excellent early neurological recovery can be achieved when a safe plane of dissection is present and careful microsurgical technique is used.

Recent series and molecular studies underscore that spinal PAs, while sharing histologic features with intracranial counterparts, form a distinct clinicoradiologic and epigenetic entity with generally favorable prognosis but a non-negligible risk of late recurrence and leptomeningeal dissemination, necessitating long-term follow-up. Continued reporting of well-documented cases with integrated imaging, histopathology, molecular profiling, and long-term outcomes will refine understanding of this rare tumor and support evidence-based management strategies.

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All authors have contributed equally.

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