

Intraspinal Space-Occupying Lesions in Children: Clinical Features, Neuroimaging and Surgical Outcomes of 27 Cases

Haigang Chang

The first Affiliated Hospital of Xinxiang Medical University

Jingkuo Yan

Ningxia Medical University

Yaxiao Wang

The First Affiliated Hospital of Xinxiang Medical University

Lei Hui

The First Affiliated Hospital of Xinxiang Medical University

Yan Li

The First Affiliated Hospital of Xinxiang Medical University

Pengju Ma

Xinxiang Medical University

Zhe Jin

Uppsala University

Feng Wang (✉ chg2010@126.com)

General Hospital of Ningxia Medical University

Research

Keywords: magnetic resonance imaging (MRI), primitive neuroectodermal tumors (PNET)

Posted Date: May 24th, 2021

DOI: <https://doi.org/10.21203/rs.3.rs-531588/v1>

License:  This work is licensed under a Creative Commons Attribution 4.0 International License. [Read Full License](#)

Abstract

Background: Pediatric intraspinal space-occupying lesions are relatively uncommon. However, these lesions can result in neurological disabilities due to misdiagnosis and delayed treatment. The main goal of the present study is to evaluate the clinical and radiological features and treatment options of pediatric intraspinal space-occupying lesions in order to improve the clinical recognition and management.

Methods: Clinical data of 27 children with intraspinal space-occupying lesions who underwent surgery treatment in a tertiary-care hospital between 2010 and 2018 were retrospectively reviewed and analyzed.

Results: Of these 27 patients, 14 (51.85%) were girls and 13 (48.15%) were boys. The most common age group affected was 10~14 years (62.96%, 9 girls and 8 boys in this age group). The mean age was 10.11 years old. Pain and weakness were the most common clinical symptoms. Preoperative magnetic resonance imaging (MRI) identified intramedullary (10 cases, 37.04%), intradural extramedullary (10 cases, 37.04%) and extradural (7 cases, 25.92%) lesions, respectively. The majority of the lesions were intraspinal tumors (23 cases, 85.19%). The histological diagnosis of tumors included embryonic residual tumors (6 cases, 22.22%), ependymoma (5 cases, 18.52%), primitive neuroectodermal tumors (PNET) (3 cases, 11.11%), schwannomas (2 cases, 7.4%), ganglioneuroma (1 case, 3.7%), Ewing's sarc (1 case, 3.7%), B-cell non Hodgkin lymphoma (1 case, 3.7%), Hodgkin lymphoma (1 case, 3.7%), chondrosarcoma (1 case, 3.7%), ganglioglioma (1 case, 3.7%), and glioma (1 case, 3.7%).

Conclusions: The incidence of pediatric intraspinal space-occupying lesions is low, and the clinical manifestation is lack of specificity. The prognosis for children with malignant tumors is poor and surgical resection is still the primary treatment option.

Background

The intraspinal occupying-lesions comprising tumoral and non-tumoral lesions are relatively rare in children [1–4], but can lead to serious neurological impairments if left untreated. The majority of the lesions are neoplastic lesions with the incidence less than 3 per 1000,000 persons per year, accounting for only 5% of pediatric central nervous system (CNS) tumors [5]. Notably, the most frequent pediatric intraspinal tumors are residual embryonic tissue tumors and neuroepithelial tumors. In contrast, meningiomas, shwannomas and ependymomas are more common in adults [6]. When symptoms of spinal cord compression occur in patients, a series of sophisticated imaging tools such as magnetic resonance (MR) and computed tomography (CT) could assist the early detection of intraspinal tumors [7]. However, the clinical presentation of intraspinal space-occupying lesions in children is insidious and lacks specific symptoms [1, 2]. Therefore, the actual diagnosis is usually delayed by months or more, which markedly affects the prognosis for the children [6]. Due to the rareness of pediatric intraspinal space-occupying lesions, only case series with limited number of patients are reported and therapeutic guidelines are not provided for pediatric intraspinal space-occupying lesions. In this study, we collected and analyzed the clinical data of pediatric intraspinal occupying-lesions at the General Hospital of Ningxia Medical University, China between 2010 and 2018, with the purpose to improve the diagnosis and treatment of these lesions in the future.

Methods

This study was approved by the hospital's Ethics Committee and the informed consent was obtained from each child's parent or guardian prior to inclusion in this study. We reviewed the clinical data of 27 pediatric patients with intraspinal space-occupying lesions who underwent surgery or biopsy from January 2010 to February 2018 at the department of neurosurgery, the General Hospital of Ningxia Medical University, China. The inclusion criteria include: 1) The age of patients ranged from 0 to 14 years; 2) The intraspinal space-occupying lesions confirmed by operation or biopsies and diagnosed by imaging examination. Pre- and postoperative magnetic resonance imaging (MRI) and electromyography were performed for all patients. Preoperative MRI was performed after admission and postoperative MRI was performed within one week after surgery. The extent of resection was evaluated by operative records and postoperative MRI re-examination. Subsequently, clinical and MRI examinations were performed at 6 months, 1 year, and once a year during follow-up. The modified McCormick Scale was used to rate the patient's global functional impairment during clinical examination. The score categories were: 1 = neurologically intact; 2 = mild motor or sensory deficit, but functional independence; 3 = moderate deficit and limitation of function; 4 = severe motor or sensory deficit, dependent; and 5 = paraplegia or quadriplegia. The clinical manifestations, imaging features, tumor location, resection degree, pathological diagnosis and prognosis of the patients were analyzed. Gross total resection was attempted for all lesions whenever possible. The intraoperative physiologic monitoring and motor evoked potential measurement were used during the surgery. The surgery was performed under the standard microsurgical conditions using the conventional laminectomy in the early years and osteoplastic laminotomy in recent years.

Results

Fourteen out of the 27 patients were female (51.85%) and 13 were male (48.15%). The patients that underwent surgery were divided into three age groups, including 0-4 years group (4 patients), 5-9 years group (7 patients) and 10-14 years group (16 patients). The median age was 10.11 years old. The mean age was 10.11 years old (range 4 – 14 years). The clinical data of all patients are summarized in Table 1.

Clinical symptoms

The intraspinal space-occupying lesions manifested with duration from onset to admission about 4-day to 5-year (mean time 7.9 months). The most common symptom at presentation was weakness in the limbs (18 cases, 66.67%), followed by pain (17 cases, 62.96%), dysesthesia (7 cases, 25.92%), gait abnormality (7 cases, 25.92%), amyotrophy (3 cases, 11.1%), incontinence (3 cases, 11.11%), oliguresis and anuria (2 cases, 7.41%), fever (2 cases, 11.11%). There were other symptoms including involuntary twitch, abnormal hair distribution, weight loss, hyperhidrosis and itching.

Neurological examination.

The most frequent neurological signs included muscle weakness (18 cases, 66.67%), hypesthesia (13 cases, 48.15%), Babinski sign (+) (7 cases, 22.22%), percussion pain (5 cases, 18.52%), increased tendon reflexes (5 cases, 18.52%), weakened tendon reflex (5 cases, 18.52%), abdominal wall, perianal and testicular reflex disappeared (4 cases, 14.81%), hyperpathia (3 cases, 11.11%). In addition, straight-leg raising test, muscular tension dysfunction, red skin rash, developmental deformity and lymphadenectasis were found in a few cases.

Magnetic resonance imaging (MRI) features

Magnetic resonance imaging scans were performed to reveal the location of lesions. MRI images showed intramedullary (10 cases, 37.04%, Fig. 1 and 3), intradural extramedullary (10 cases, 37.04%, Fig. 4), and extradural lesions (7 cases, 25.93%, Fig. 5). Three lesions were located in the cervicothoracic spine (11.11%), 11 in the thoracic spine (40.74%, including 2 malignant tumors from posterior mediastinum and thoracic vertebrae), 4 in the thoracolumbar spine (14.81%), 6 in the lumbar spine (22.22%), and 3 in the lumbosacral spine (11.11%).

Surgical treatment

The tilt angle of operating bed was adjusted according to lesions. For extramedullary tumors, the prone position was usually used; for intramedullary tumors, the lateral position was usually selected, which was more beneficial to keep the surgical field clear. The surgical approach for all lesions was usually through a midline incision centered on the spinal lesion, based on bone surface markers. The lamina and spinous processes dorsal to the intraspinal occupying lesions were then removed. The exposure should be appropriate for adequate decompression and exposure of occupying lesions as well as maintaining the spinal stability. After the lamina was opened, the operation was performed under a microscope. For intradural tumors, a clear plane of masses between the tumor and the surrounding normal tissue was carefully dissected and resected. In the case of infiltrating tumors, it was attempted to reduce the volume as much as possible from within the lesions. Surgical resection was performed with an ultrasonic knife. If the lesions were tightly attached to the spinal cord/cauda equina nerve/filum terminale, and the boundary was unclear, the capsule was first incised to remove the tissue in the lesion, followed by careful separation of the nerve rootlets and spinal cord along the capsule. If necessary, the filum terminale was cut off, and the lesion was subtotally resected in blocks to preserve the nerve integrity. Before suturing the dura mater, all nerves should be combed as far as possible to avoid nerve adhesion or stretch. The spinal dura was closed with a watertight suture and the laminar and spinal process were restored. Normally no drainage tube was placed except for the inflammatory lesion.

Histopathological examination

Histopathological examination was performed in 27 cases. Intraspinal tumors were found in 23 cases (84.62%): embryonal remnants tumors in 6 cases (epidermoid cyst in 3 cases, lipoma in 2 cases, cystic teratoma in one case), ependymomas in 5 cases (myxopapillary ependymoma in 3 cases, Fig. 6A and B; ependymoma (WHO II) in 2 cases, Fig. 6C and D), primitive neuroectodermal tumors in 3 cases, schwannomas in 2 cases (malignant peripheral epithelioid schwannoma in one case), ganglioneuroma in one case, Ewing's sarcoma in one case, B-cell non-Hodgkin lymphoma in one case (Burkitt lymphoma subtype), Hodgkin lymphoma in one case, chondrosarcoma in one case, ganglioglioma in one case, glioma in one case. Intraspinal non-neoplastic lesions were found in 4 cases (15.38%): chronic granulomatous inflammation in 2 cases, tuberculous granuloma in one case, and nerve root sleeve cyst in one case.

Postoperative outcome and follow-up

Preoperative and postoperative Modified McCormick scale (MMCS) [8, 9] (Table. 2) was performed in 27 patients. One to three weeks after surgery, the clinical symptoms and neurological function of 20 patients were improved in varying degrees. The clinical symptoms were remained unchanged in 4 patients and even deteriorated in one patient. There was no operative (in-hospital) death. The postoperative infection occurred in one case. A total of 27 cases were followed up via outpatient service and telephone. The clinical follow-up period ranged from two months to seven years. Two patients with intraspinal primitive neuroectodermal tumors (PNET) died. Two cases of myxopapillary ependymoma relapsed from L2-3 to L5-S2 and T12-L3 to S1-3 at 11 and 12 months after surgery, respectively. One patient with ependymoma experienced recurrence three years postoperatively. Malignant peripheral epithelioid schwannoma invaded into the spinal canal from posterior mediastinum in one case (Fig. 2). In one case Hodgkin lymphoma invaded from the thoracic vertebrae to the spinal canal. The lesions in 2 cases were large, involving many organs and having poor surgical outcomes, further received chemotherapy. The volume of the lesions reduced after chemotherapy.

Discussion

Intraspinal space-occupying lesions are rare in pediatric patients and lack of uniform evidence-based management strategies [5, 7]. In contrast to adult intraspinal space-occupying lesions, pediatric cases have their own characteristics in clinical manifestations, histopathology, and surgical outcomes. The physiological features of child growth and development require a relatively independent system for diagnosis and treatment of diseases. The potential factors that result in misdiagnosis and missed diagnosis of pediatric intraspinal space-occupying lesions could be due to insufficient medical history and lack of comprehensive neurological examination. Therefore, the pathological, physiological, anatomical and immunological uniqueness of children should be fully taken into account when dealing with pediatric intraspinal space-occupying lesions, and a multi-disciplinary team can ensure timely diagnosis and optimal prognosis [1].

Pediatric intraspinal space-occupying lesions include a variety of tumors and non-neoplastic lesions. These tumors are originated from e.g. embryonic remnants, spinal cord and nerves, spinal dura mater and adipose tissue. In contrast to schwannomas, ependymomas and meningiomas that are most common in adults, intraspinal tumors in children are predominantly embryonic residual tissue tumors (teratomas, epidermoid cysts, dermoid cysts, enterogenous cysts, lipomas), and neuroepithelial tissue tumors [6]. A previous study has shown that the extradural and intramedullary lesions are most frequent in children as compared to adults, suggesting the location of tumors within the spinal cord, the meninges, and the spine differs between children and

adults[10]. In the present study, intramedullary, intradural extramedullary and extradural lesions account for 37.04 %, 37.04% and 25.93% of all children cases, respectively.

The onset of intraspinal lesions in children is insidious, and the clinical symptoms are nonspecific. Clinical symptoms occur only after the spinal cord and nerves are compressed by the lesion. The most frequently encountered complaint at presentation is dyskinesia, including progressive reduction of myodynamia, unstable walking, gait abnormality and hemiparesis[8]. Similarly, the most common clinical symptoms in this study are progressive reduction of myodynamia (66.67%) and unstable walking (25.92%). Another relatively common symptom is pain (nerve root pain, dull pain or radiation pain), which is usually caused by nerve root compression, but the children often have difficulties to explain it verbally. Back pain caused by intradural lesions is usually exacerbated in the supine position at night and could be associated with dural swelling resulted from the venous congestion. Other symptoms include sciatica, limb numbness, muscle atrophy, and autonomic nerve dysfunction caused by spinal cord compression, the latter consists of urinary weakness, urinary retention, incontinence, and defecation difficulties. These symptoms did not normally precede motor or sensory defects, and the condition was quite serious at the time of diagnosis.

Early neurological dysfunction is easily overlooked due to the relatively difficult or incomplete description of symptoms from children, particularly for infants. Some clinical manifestations such as instability of walking are important for early diagnosis. Young children could not clearly describe the nature and location of pain and in infants it could be manifested as crying for unknown reasons, sweating or scratching local skin. More attention should be paid to children with newly occurred back pain without trauma. In this study, one child with intraspinal primitive neuroectodermal tumor has suffered from aggravated abdominal pain for six months and sought medical advice at night, but it was difficult to make a definite diagnosis before a thorough medical examination. Moreover, autonomic dysfunction such as diurnal voiding was also easily overlooked in children with intraspinal lesions. In addition, as one-third of children with intraspinal lesions can develop scoliosis, new-onset scoliosis should also be given more attention. In particular, for scoliosis with lumbar back pain the imaging examination should be performed as early as possible [4].

A comprehensive and structured neurological examination is very important for infants who cannot accurately describe symptoms. Asymmetric nerve reflex may be an early sign of intraspinal space-occupying lesions. Tendon hyperreflexia of lower extremities may indicate that lesions are located in the lower cervical spine, thoracic spine and lumbar spine, whereas hyperreflexia of the higher extremities may indicate lesions in the upper cervical spine. The subtle symptoms of intraspinal space-occupying lesions can be well revealed by assessing the gait of the child, including walking postures of the toes and heels. Because of the incompatibility of physical examination, the children are instructed to change from sitting position to standing position, and subtle changes in proximal lower limb muscle strength are observed. The tenderness on percussion over vertebral spinous process may provide a clue for the location of lesion. Digestive system and bladder dysfunctions usually occur at late stages, but sphincter tension and urodynamic examinations are still helpful. In addition, the presence of brown spots, subcutaneous nodules or Lisch's ganglia in the skin may indicate the presence of neurofibromatosis. Clustered, convoluted hair may appear on the skin near dermoid cysts, epidermoid cysts, and teratomas.

When the intraspinal space-occupying lesion is suspected, MR imaging examination should be performed as early as possible. High-resolution contrast-enhanced MRI can not only display the solid components of intraspinal space-occupying lesions, secondary cysts, and edema, but also detect scoliosis, which provide adequate information for further diagnosis[4]. In addition, the heterogeneity of embryonic residual tumors such as intraspinal endothelioid cysts, epidermoid cysts or teratomas can be demonstrated by MR imaging. MRI images reveal that intramedullary ependymomas often have T1-iso-intense or slightly hypo-intense signal, T2-hyperintense signal, homogeneous enhancement, and some of the tumor poles form cavities. In contrast, the heterogeneous enhancement can be caused by cyst formation or necrosis [11]. Malignant peripheral epithelioid schwannomas are usually observed as isointense on the T1-weighted MRI image and hyperintense on the T2-weighted image with well-defined margins and strong contrast enhancement and polycystic changes are present within the lesions [12, 13]. The radiographic findings of intraspinal PNET are usually lack of specificity, and radiographic diagnosis may often indicate astrocytoma, ependeoma or schwannoma. Although imaging data is nonspecific, the possibility of PNET should be considered in pediatric patients when MRI reveals large, well-defined margin intraspinal space-occupying lesions [14].

The surgical resection is still the primary treatment of pediatric intraspinal space-occupying lesions. Early surgical resection and rehabilitation at early stages after surgery are beneficial to the preservation of spinal cord and recovery of neurological functions [15]. In this study, the clinical symptoms were significantly improved in 20 out of 25 patients with surgical resection. The challenges of surgical resection are mainly to avoid spinal cord injury and maintain spinal stability. Because children are at a special stage of growth and development, the incidence of postoperative spinal deformity is significantly higher than that of adults. Therefore, under the premise of adequate decompression, the destruction of bone should be minimized and the composite structure of muscle and ligament should be preserved. It has been suggested that removal of lamina, spinous process and posterior ligament complexes and reconstruction of the lamina after resection of the lesion may be beneficial to maintain spinal stability. However, some reports demonstrate this procedure has no significant impact [16]. Gross-total resection of spinal cord ependymoma should be performed without postoperative radiotherapy and chemotherapy, as the incomplete resection is the main cause of postoperative recurrence of ependymoma [17]. The presence or absence of intraoperative tumor planes is the most important factor in determining the extent of resection. For malignant tumors, postoperative radiotherapy and chemotherapy can improve the quality of life and prolong the survival time of children. However, due to the rareness of intraspinal tumors, there is still a large amount of research on the efficacy of radiotherapy and chemotherapy in need [18, 19].

Conclusions

The intraspinal occupying-lesions are relatively rare in children. The first symptoms or clinical manifestations of intraspinal non-neoplastic lesions in children are sometimes very similar to those with intraspinal neoplastic lesions, but the treatment and prognosis are different from those with tumor lesions. Diagnosis of these diseases depend on histopathologic findings. Microsurgical resection of the intraspinal space-occupying lesions is the mainstay of treatment. Further

studies with larger sample sizes, surgical resection combined with adjuvant radiotherapy and chemotherapy, longer follow-up periods and multicenter prospective database should yield stronger conclusions.

Declarations

Ethics approval and consent to participate

This study was approved by the hospital's Ethics Committee and the informed consent was obtained from each child's parent or guardian prior to inclusion in this study.

Consent for publication

Written, informed consent was obtained from the patients for published images of this article.

Availability of data and materials

Data used to support the findings of this study are available from the corresponding author upon request.

Competing interests

The authors declare no conflict of interest.

Acknowledgement and Funding resource

We gratefully acknowledge the following funding sources for this study: The Key Research and Development Program of Ningxia (2018BFG02007) and Youth Foundation of the First Affiliated Hospital of Xixiang Medical University (QN-2017-B008).

Authors' contributions

CHG, WYX and YJK carried out the experimental work and the data collection and interpretation. HL, LY and MPJ participated in the design and coordination of experimental work, and acquisition of data. JZ and WF participated in the study design, data collection, analysis of data and preparation of the manuscript. CHG and YJK carried out the study design, the analysis and interpretation of data and drafted the manuscript. All authors read and approved the final manuscript.

References

1. Hsu W, Gl. Jallo. Pediatric spinal tumors. In: O. Dulac, M. Lassonde, H.B. Sarnat, Editors. Handbook of Clinical Neurology. Elsevier B.V. ; 2013. Vol. 112 (3rd series); p. 959–965. <https://doi.org/10.1016/b978-0-444-52910-7.00016-7>.
2. Joaquim AF, Ghizoni E, MGC V, Appenzeller S, SDS A, Tedeschi H. Spinal tumors in children. *Rev Assoc Med Bras* (1992). 2017; 63(5): 459–465. doi: 10.1590/1806-9282.63.05.459.
3. Nakashima K, Koga Y, Sakai Y, Takada H, Harimaya K, Ohga S, et al. Radiotherapy for Langerhans cell histiocytosis with paraplegia: A rare oncologic emergency case report in infancy and literature review. *Brain Dev*. 2018;40(10):952–5. doi. 10.1016/j.braindev.2018.05.016.
4. Calloni SF, Huisman TA, Poretti A, Soares BP. Back pain and scoliosis in children: When to image, what to consider. *Neuroradiol J*. 2017;30(5):393–404. doi. 10.1177/1971400917697503.
5. Shweikeh F, Quinsey C, Murayi R, Randle R, Nuño M, Krieger MD, et al. Treatment patterns of children with spine and spinal cord tumors: national outcomes and review of the literature. *Childs Nerv Syst*. 2017; 33(8): 1357–65. <https://doi.org/10.1007/s00381-017-3433-y>.
6. Duong LM, McCarthy BJ, McLendon RE, Dolecek TA, Kruchko C, Douglas LL, et al. Descriptive Epidemiology of Malignant and Nonmalignant Primary Spinal Cord, Spinal Meninges, and Cauda Equina Tumors, United States, 2004–2007. *Cancer*. 2012;118(17):4220–7. doi. 10.1002/cncr.27390.
7. Merlot I, Francois J, Marchal JC, Joud A, Guerbouz R, Chastagner P, et al. Spinal cord tumors in children: A review of 21 cases treated at the same institution. *Neurochirurgie*. 2017;63(4):291–6. doi. 10.1016/j.neuchi.2017.01.008.
8. Choi GH, Oh JK, Kim TY, You NK, Lee HS, Yoon DH, et al. The clinical features and surgical outcomes of pediatric patients with primary spinal cord tumor. *Childs Nerv Syst*. 2012; 28(6): 897–904. <https://doi.org/10.1007/s00381-012-1718-8>.
9. McCormick PC, Torres R, Post KD, Stein BM. Intramedullary ependymoma of the spinal cord. *J Neurosurg*. 1990;72:523–32.
10. Spacca B, Giordano F, Donati P, Genitori L. Spinal tumors in children: long-term retrospective evaluation of a series of 134 cases treated in a single unit of pediatric neurosurgery. *Spine J*. 2015;15(9):1949–55. doi. 10.1016/j.spinee.2015.04.012.
11. Bandopadhyay P, Silvera VM, Ciarlini PD, Malkin H, Bi WL, Bergthold G, et al. Myxopapillary ependymomas in children: imaging, treatment and outcomes. *J Neurooncol* 2016; 126(1): 165–74. <https://doi.org/10.1007/s11060-015-1955-2>.
12. Rekhil B, Kossemehmetoglu K, Tezel GG, Dervisoglu SC. Clinicopathologic features and immunohistochemical spectrum of 11 cases of epithelioid malignant peripheral nerve sheath tumors, including INI1/SMARCB1 results and BRAF V600E analysis. *APMIS*. 2017;125(8):679–89. doi. 10.1111/apm.12702.
13. Jo VY, Fletcher CD. Epithelioid malignant peripheral nerve sheath tumor: clinicopathologic analysis of 63 cases. *Am J Surg Pathol*. 2015; 39(5): 673–82. <https://doi.org/10.1097/PAS.0000000000000379>.

14. Qi W, Deng X, Liu T, Hou Y, Yang C, Wu L, et al. Comparison of Primary Spinal Central and Peripheral Primitive Neuroectodermal Tumors in Clinical and Imaging Characteristics and Long-Term Outcome. *World Neurosurg.* 2016;88:359–69. doi. 10.1016/j.wneu.2015.12.033.
15. Kose N, Muezzinoglu O, Bilgin S, Karahan S, Isikay I, Bilginer B. Early rehabilitation improves neurofunctional outcome after surgery in children with spinal tumors. *Neural Regen Res.* 2014;9(2):129–34. doi. 10.4103/1673-5374.125340.
16. McGirt MJ, Garcés-Ambrossi GL, Parker SL, Sciubba DM, Bydon A, Wolinsky JP, et al. Short-term progressive spinal deformity following laminoplasty versus laminectomy for resection of intradural spinal tumors: analysis of 238 patients. *Neurosurgery.* 2010;66(5):1005–12. doi. 10.1227/01.NEU.0000367721.73220.C9.
17. Safaee M, Oh MC, Mummaneni PV, Weinstein PR, Ames CP, Chou D, et al. Surgical outcomes in spinal cord ependymomas and the importance of extent of resection in children and young adults. *J Neurosurg Pediatr.* 2014;13(4):393–9. doi. 10.3171/2013.12.PEDS13383.
18. Ahmed R, Menezes AH, Awe OO, Torner JC. Long-term disease and neurological outcomes in patients with pediatric intramedullary spinal cord tumors. *J Neurosurg Pediatr.* 2014; 13(6): 600–12. <https://doi:10.3171/2014.1.PEDS13316>.
19. Ahmed R, Menezes AH, Torner JC. Role of resection and adjuvant therapy in long-term disease outcomes for low-grade pediatric intramedullary spinal cord tumors. *J Neurosurg Pediatr.* 2016;18(5):594–601. doi. 10.3171/2016.5.PEDS15356.

Tables

Table 1. Clinical characteristics of 27 children with intraspinal space-occupying lesions.

Case no.	Age (years)	Sex	Clinical symptoms and signs	Duration (months)	Neuroimaging features (Level/location)	Surgical treatments	Pathology	MMCS		Outcome	Follow-up (months)
								Pre	Post		
1	5	M	LBP, unstable walking	6	L4-L5/ Intradural extramedullary	GTR	Epidermoid cyst	2	2	0	36.4
2	14	F	LBP	24	L5-S2/ Intradural extramedullary	GTR	Epidermoid cyst	1	1	0	84
3	14	M	Lt L/Ex weakness, incontinence	12	L1-L2/ Intramedullary	STR	Epidermoid cyst	3	2	↑	68.9
4	12	M	LBP, muscular dystrophy	0.7	L1/ Intramedullary	STR	Lipoma	2	2	↑	13.6
5	13	F	L/Ex weakness, incontinence	12	L5-S1/ Extradural	STR	Lipoma	3	3	↑	21.7
6	14	M	LBP, incontinence	12	L1-L2/ Intramedullary	GTR	Cystic teratoma	3	3	↑	19.2
7	12	F	Low back pain	0.5	L2-L3/ Intradural extramedullary	GTR	Myxopapillary ependymoma	1	1	ECRE	11
8	10	M	LBP, headache (hydrocephalus)	11	T12-L3/ Intramedullary	GTR	Myxopapillary ependymoma	1	1	ECRE	12
9	11	F	Lumbosacral pain, L/Ex weakness	0.7	L2/ Intradural extramedullary	GTR	Myxopapillary ependymoma	3	2	↑	57.2
10	14	F	Lt L/Ex pain	1	T10-L1/ Intramedullary	GTR	Ependymoma(WHO Ⅱ)	1	1	0	9.3
11	4	F	Hyperhidrotic, neck pain	1	T3-T8/ Intramedullary	GTR	Ependymoma(WHO Ⅱ)	1	1	RE	36
12	6	F	L/Ex weakness, dysuria, incontinence	1	L3-S2/ Extradural	GTR	PNET	4	3	dead	7.5
13	7	M	Abdominal pain	6	T6-L1/ Intradural extramedullary	STR	PNET	1	1	0	20.3
14	9	F	LBP, unstable walking	0.3	T6-T12/ Intradural extramedullary	GTR	PNET	3	3	dead	2
15	13	M	L/Ex numbness and weakness, unstable walking,	6	T6-T7/ Extradural	GTR	Schwannoma	3	3	↑	35.6
16	4	M	L/Ex weakness, chest pain, dysuresia	0.2	T7-T11/ Intramedullary	B	Malignant peripheral epithelioid schwannoma	3	2	DI	16.5
17	14	F	Neck and shoulder pain, weakness, incontinence	24	C7-T1/ Intramedullary	GTR	Ganglion neurofibroma	3	2	↑	48.2
18	6	F	L/Ex weakness, dysuresia	0.1	T2/ Intradural extramedullary	GTR	Ewing's sarcoma	3	2	0	8.6
19	8	M	L/Ex numbness and weakness	0.2	T3-T6/ Extradural	GTR	B cell non Hodgkin lymphoma	2	2	0	10.7
20	14	M	LBP, L/Ex numbness and weakness	0.3	T8-T10/	B	Classical Hodgkin lymphoma	3	2	DI	7.3

						Intradural extramedullary						
21	11	F	LBP, muscular dystrophy, Lt lower limb weakness	2	T10-T11/ Intradural extramedullary	GTR	Mesenchymal chondrosarcoma	3	2	↑		11.6
22	4	M	Upper limbs weakness, buttock pain	12	C4-T12/ Intramedullary	STR	Ganglioglioma	2	2	0		49.6
23	4	F	Limp, muscle strength reduction	3	T11-L5/ Intramedullary	PR	Glioma	2	2	↑		10.3
24	14	M	LBP, both L/Ex weakness	3	T7-T8/ Extradural	GTR	Inflammatory granuloma	2	2	0		31.6
25	11	M	Thorax-back pain, L/Ex weakness	1	T1-T3/ Extradural	GTR	Inflammatory granuloma	3	2	↑		36.5
26	14	F	L/Ex numbness and weakness	2	T9-T10/ Intradural extramedullary	GTR	Tuberculous granuloma	3	2	0		42.8
27	11	F	Rt weakness	60	C4-T1/ Epidural	GTR	Nerve root sleeve cyst	2	1	0		60.3

F: female, M: male, Pre: preoperative, Post: postoperative, Lt: left, Rt: right, L/Ex: lower extremity, LBP: lower back pain, PNET: primitive neuroectodermal tumor, GTR: gross-total resection, STR: subtotal resection, B: biopsy, PR: partial resection, MMCS modified McCormick scale, ↑ improved, ↓ deteriorated, 0 remained unchanged, ECRE: ectopic recurrence, RE: recurrence, DI: distant invasion

Table 2. Modified McCormick scale for Clinical/functional classification scheme

Grade	Definition
1	Neurologically intact, ambulates normally, may have minimal dysesthesia
2	Mild motor or sensory deficit, maintains functional independence
3	Moderate deficit, limitation in function, independent with external aid
4	Severe motor or sensory deficit, dependent with external aid
5	Paraplegia or quadriplegia

Figures

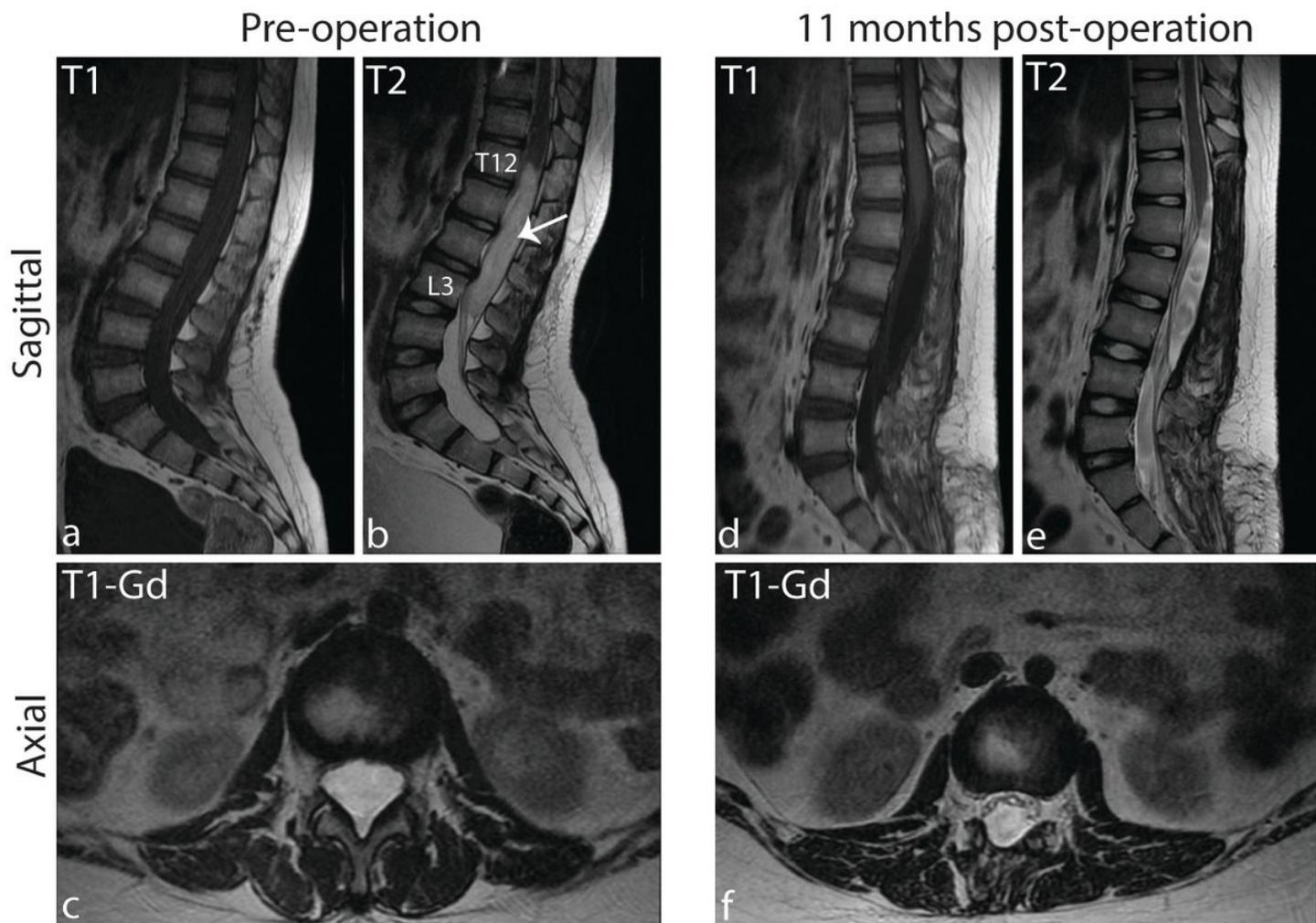


Figure 1
 MRI images of a male, 10 years old with myxopapillary ependymoma (patient 8) . (a-c) Pre-operative MRI showing an intramedullary lesion at the T12-L3 level with hypointensity on the sagittal T1-weighted image (a) and hyperintensity on the sagittal T2- weighted image (b),enhancement on the axial T1-weighted image (c), compressing and pushing conus medullaris and cauda equine. (d-g) Images taken 11 months after surgery showing a well-circumscribed fusiform lesion with T1-hypointensity (d) and T2-hyperintensity (e) from the upper edge of the lumbar L1 to the lower edge of the lumbar L3. No significant enhancement was observed (f and g).

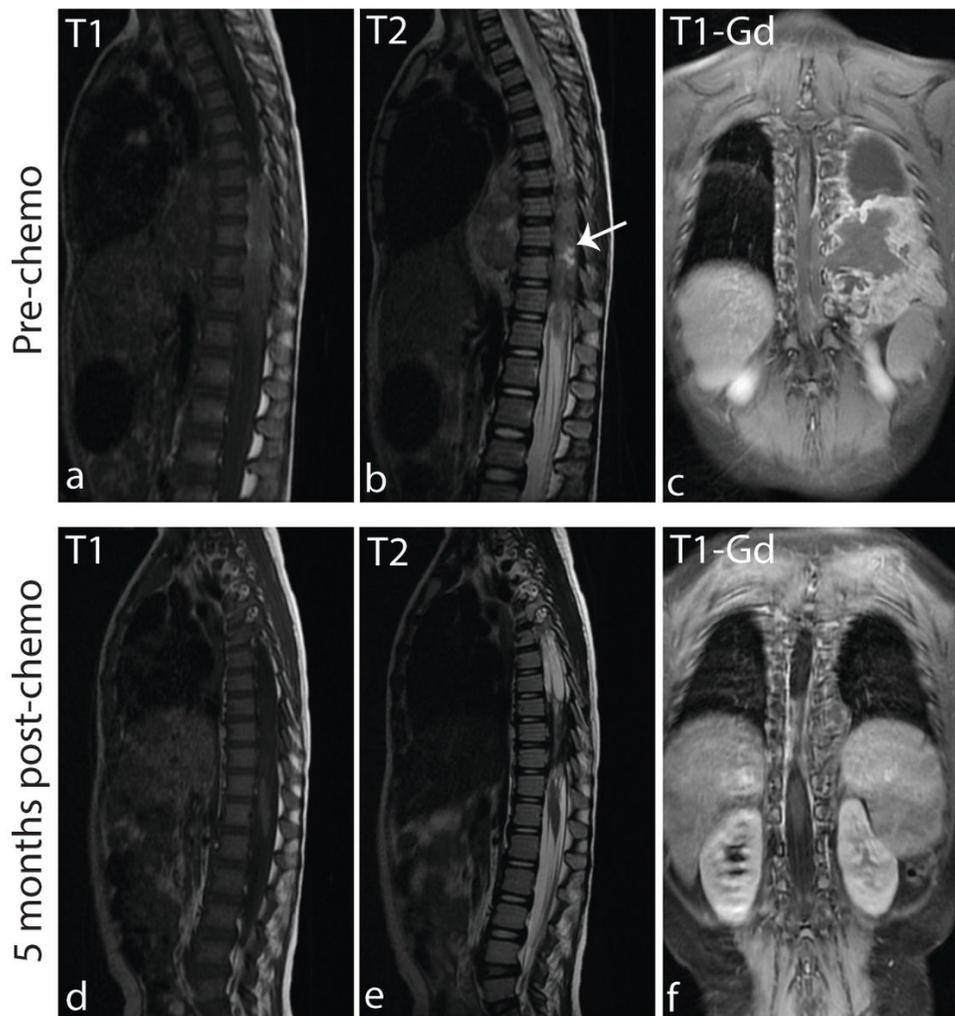


Figure 2

MRI images of a male, 4 years old with malignant peripheral epithelioid schwannoma. (a) T1-weighted image showing the isodense lesion and (b) T2-weighted image showing variable density of the lesion. (c) T1-weighted image shows the tumor inhomogeneous enhancing after gadolinium enhancement. The lesions spread through the intervertebral foramen to the spinal canal. (d-f) Five months after chemotherapy on schedule, T1-weighted image, T2-weighted image and contrast enhanced MRI image of the thoracic spine showing the lesion in the posterior mediastinum was reduced than before, and the extent of lesions in the spinal canal increased slightly.

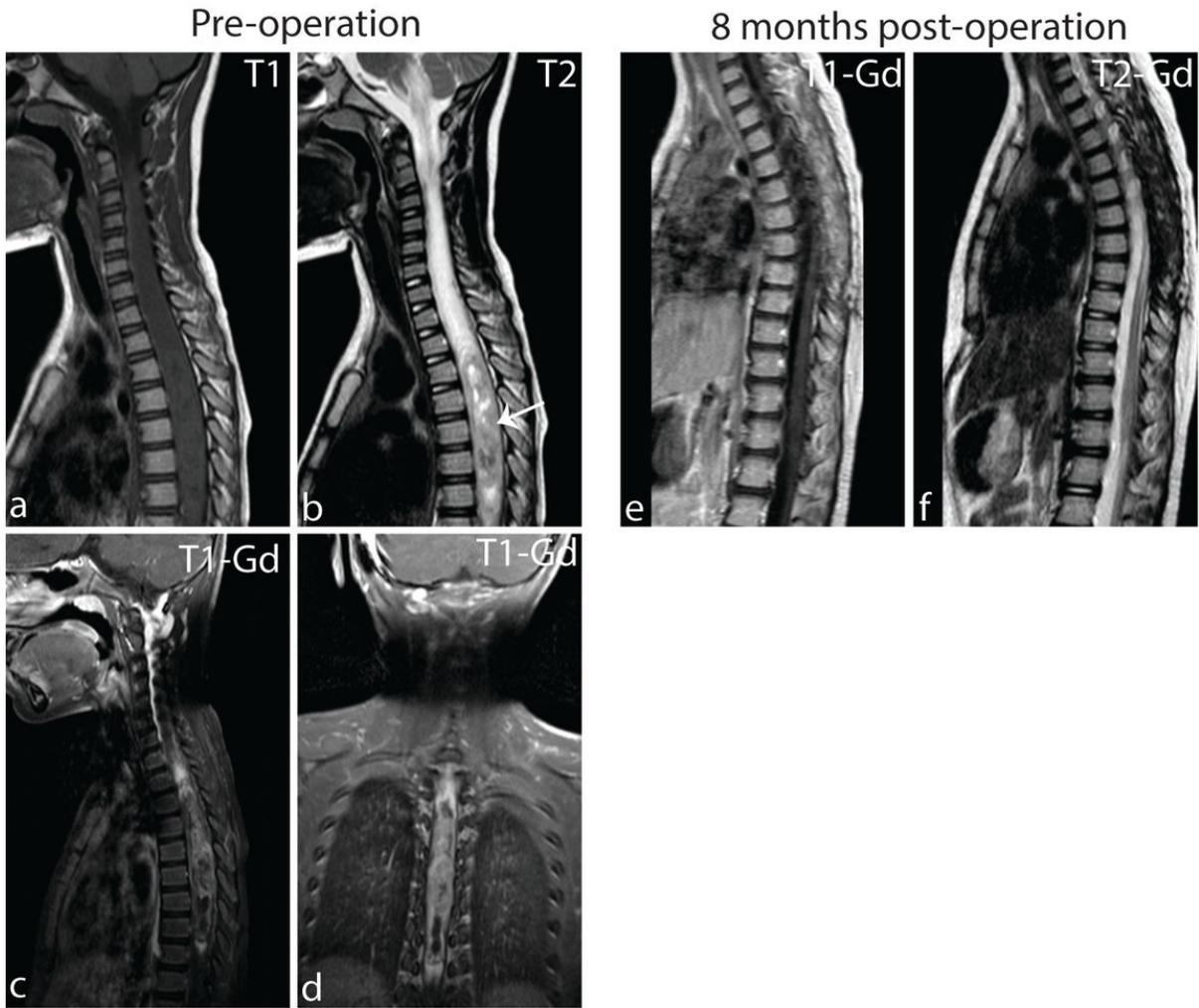


Figure 3
 MRI images of a female, 4 years old with ependymoma (patient 11). (a-d) Pre-operative MRI revealing an intramedullary lesion at T3-T8 with abnormal signal intensity. The lesion is heterogeneously hypointense on T1-weighted imaging (a), heterogeneously hyperintense on T2-weighted imaging (b), and has less clear boundaries on enhancing T1-weighted images (c and d). (e and f) Eight months post surgery, contrast enhanced MRI images of thoracic spine showing no tumor recurrence.

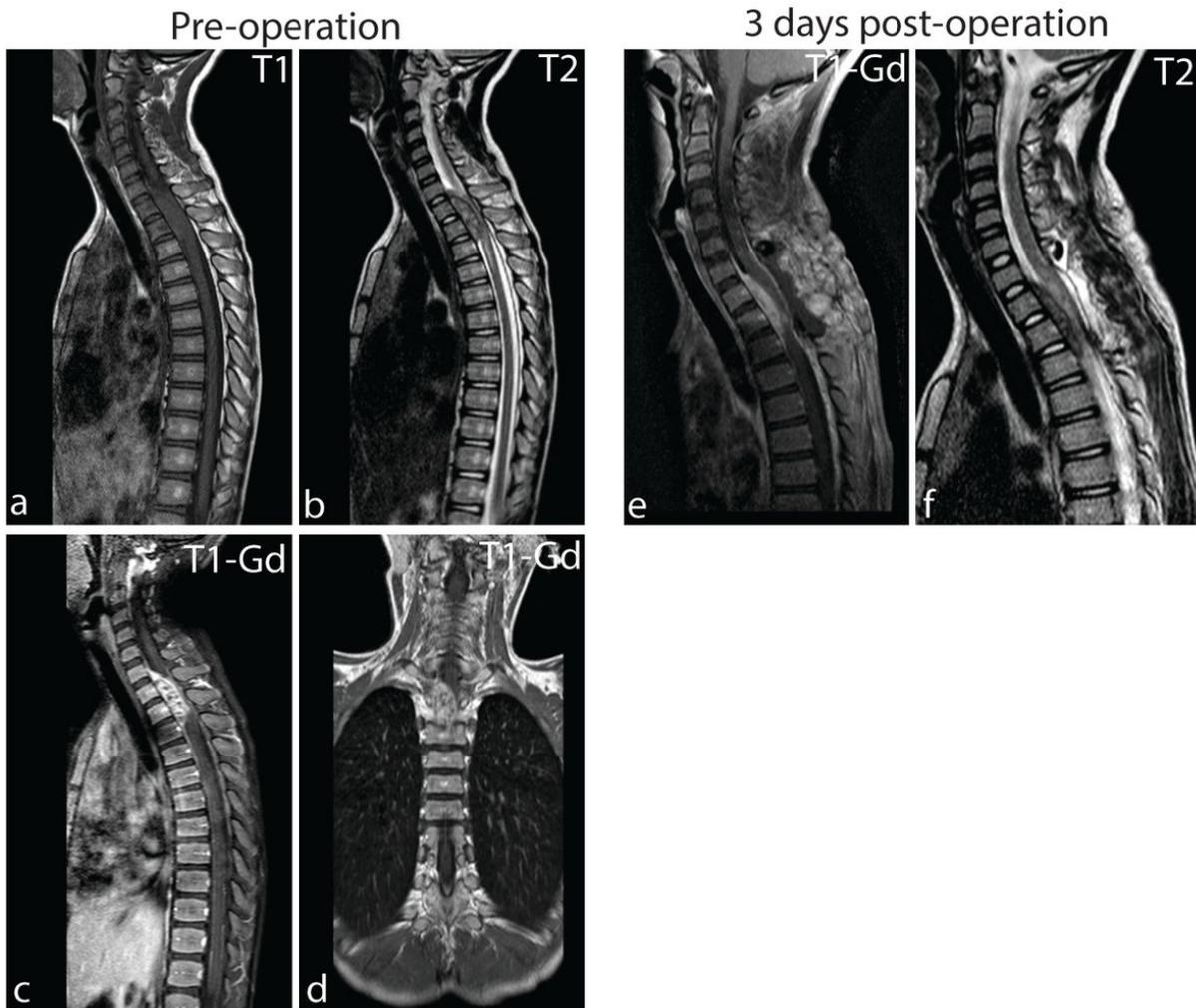


Figure 4

MRI images of a male, 11 years old (patient 25). (a-d) Pre-operative MRI revealing an intradural extramedullary lesion on the right with slight hypointensity on the sagittal T1-weighted image (a) and mixed hyperintensity on the sagittal T2-weighted image (b). (c and d) Sagittal T1-weighted image after gadolinium administration: heterogeneous contrast enhancement, without enhancement in multiple small class flakes. After the administration of gadolinium, this image shows the spinal dura close to the lesion with intense enhancement. At the same level, the thoracic spinal cord was obviously compressed and displaced, and the T3 and 4 levels of central canal dilated. (e and f) Three days after surgery, T2-weighted sagittal image (e) and T1-weighted sagittal image (f) of the lesion after gadolinium administration showing the reduced lesion.

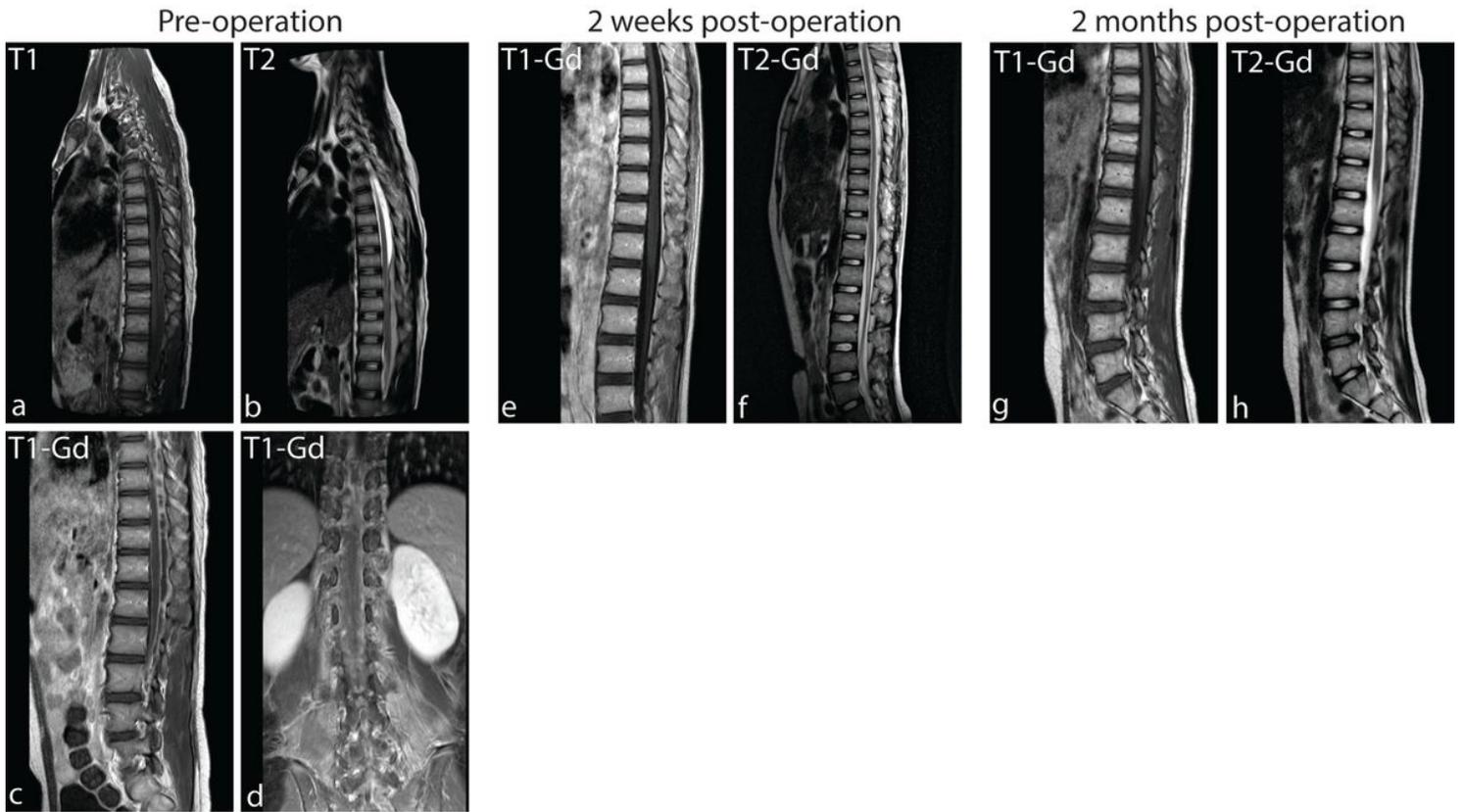


Figure 5
 MRI images of a male, 14 years old (patient 24). (a-d) Pre-operative MRI revealing an extradural lesion at T8-L4 with hypointensity and hyperintensity on the sagittal T1- (a) and T2 (b)-weighted image. (c and d) Sagittal T1-weighted image after gadolinium administration: heterogeneous contrast enhancement. (e-h) Two weeks (e and f) and two months (g and h) after surgery, postoperative contrast enhanced magnetic resonance imaging of thoracic spine revealed the lesion reduction and no lesion recurrence.

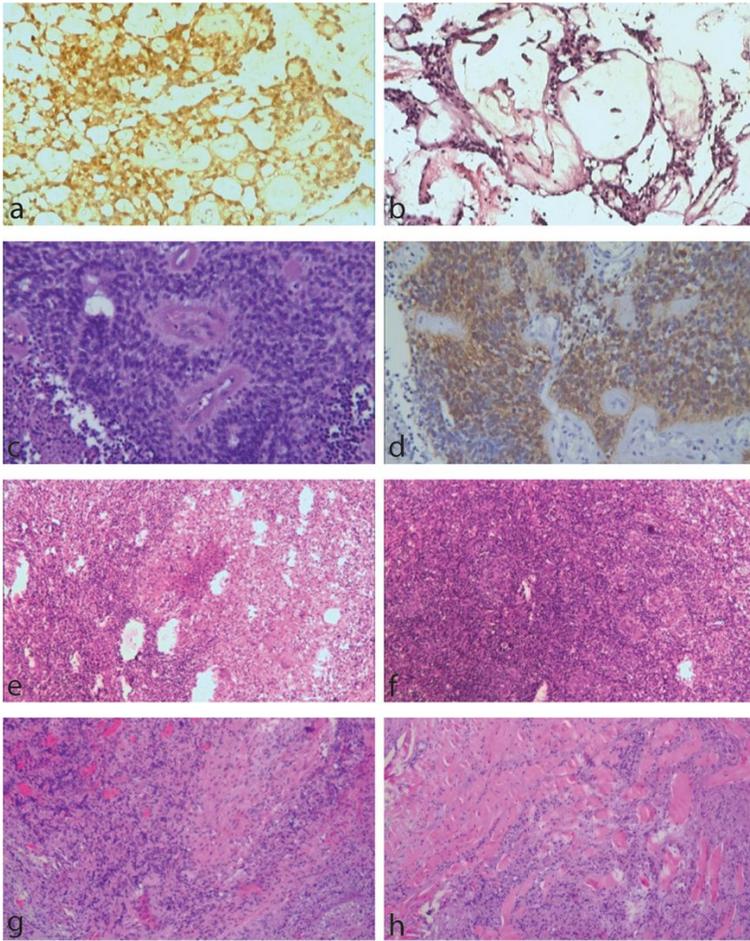


Figure 6
 Histopathological examination. (a and b) Representative H&E staining images of a myxopapillary ependymoma (patient 8). The tumor tissue was papillary and the surface of the nipple was covered with cuboidal epithelium. The connective tissue axis was mucoid and the immunohistochemical examinations revealed that the tumors were positive for S-100 (+), glial fibrillary acidic protein (GFAP) (+), Vimentin (+) and negative for epithelial membrane antigen (EMA) (-), CK-P (-), and less than 1% of cells are positive for Ki-67, which confirmed a myxopapillary ependymoma (WHO I grade). (c and d) Representative H&E staining images of an ependymoma (patient 11). The oval cells were uniform in size, and the tumor cells were arranged in pseudo-chrysanthemum-like clusters around the blood vessels, with hemorrhage and necrosis. Immunohistochemical examination reveals that the tumor is positive for GFAP (+), Vimentin (+), CD34 (vascular endothelial +), S100 and EMA were partly positive. About 5% of cells are positive for Ki67, which confirmed an ependymoma (WHO grade II). (e and f) Representative H&E staining images of an inflammatory granuloma (patient 25). A large number of lymphocytes infiltrated and proliferating cells and multinucleated giant cells in proliferative fibrous connective tissue, which indicated chronic granulomatous inflammation. (g and h) Representative H&E staining images of an inflammatory granuloma (patient 24). Fibrous connective tissue, rhabdomyosarcoma, focal vascular hyperplasia, visible infiltration of inflammatory cells, indicated the chronic inflammatory response.