MEDICAL SCIENCE MONITOR

CLINICAL RESEARCH

e-ISSN 1643-3750
© Med Sci Monit, 2018; 24: 7072-7089
DOI: 10.12659/MSM.910447

Received: 2018.04.07
Accepted: 2018.05.10
Published: 2018.10.05

Pediatric Spinal Ependymomas

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Source of support: Departmental sources

Background: The aim of this study was to assess the clinical and radiological outcomes of surgical treatment for primary spinal ependymoma in children.

Material/Methods: Medical records of 46 primary spinal ependymoma patients who underwent surgery in BRSHH hospital during a 12-year period from 2004 to 2015 were retrospectively reviewed. All pediatric patients (patient age <18 years) were selected as the core sample used for this study.

Results: This series included 1 female and 2 male patients between the ages of 9 and 17 years with mean age 13.3±3.9 years. The mean preoperative course was 9.1±10.5 months. The most common location was the lumbar spinal cord (n=2). The most common presenting symptoms was lower-limb weakness and numbness. Two tumors were located intradural-intramedullary and 1 was located intradural-extramedullary. Gross-total resection (GTR) was achieved in 2 patients, and a near-total resection was performed in 1 patient. No adjuvant treatment was received. The mean follow-up duration was 51.3±37.6 (17–98) months. No complications were recorded. Functional assessment of all patients by the latest follow-up evaluation showed good progress even though the patient is not fully recovered. At 6.3 years after the first operation, 1 patient presented with drop-seeding metastasis. No patients had neurofibromatosis type 2.

Conclusions: Laminoplasty and intraoperative neurophysiological monitorization are essential in surgical treatment of pediatric spinal ependymomas. GTR and recovery in pediatric spinal ependymoma are more likely than in adults. Despite the GTR, the risk of drop metastasis remains. Therefore, close clinical and radiological follow-up is recommended.

MeSH Keywords: Ependymoma • Glioma • Laminectomy

Full-text PDF: https://www.medscimonit.com/abstract/index/idArt/910447

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Background

Primary spinal ependymoma are very rare in the pediatric population. Population-based data indicate that the overall incidence rate per 100 000 person-years was significantly higher in male than in female patients (males 0.227±0.029, females 0.166±0.03) [1,2]. Recently, a population-based study found that only 31 spinal pediatric ependymomas (SPEs) were reported in the USA during a 31-year period from 1973 to 2003 [1]. In children, ependymomas are the most common spinal cord tumors [1,2]. Spinal ependymomas are usually well-circumscribed. Histopathological classification includes myxopapillary ependymoma (MPE) and subependymoma, which are classified as WHO grade I, classic (typical), and papillary ependymoma classified as WHO grade II, and anaplastic ependymoma referred to as WHO grade III. Low-grade (grade I and II) lesions are more common than anaplastic ependymoma.

MPEs occur most commonly in the lumbosacral region and originate from within the terminal filum or the conus medullaris. Such lesions account for 13% of all spinal cord ependymomas between all age groups. These lesions appear to be common in patients who present in their third or fourth decade of life [3]. There is a limited number of pediatric cases have been reported with details. In the pediatric population, there has been ongoing debate about the characteristics that distinguish WHO grade II and grade III ependymomas [4].

In adults, spinal ependymomas vary greatly in size and typically have a long prodrome. They frequently cause nonspecific symptoms such as numbness, nighttime back, and limb pain, which may be misinterpreted as radicular pain caused by degenerative disease [5]. This is not true for children. Thanks to their small-diameter spinal canal compared to adults, pediatric SPEs have a short prodrome (<12 months). Intramedullary ependymomas in the cervical or thoracic spine can coexist with syrinx cavities, which may lead to motor, sensory, urinary, and gait abnormalities.

Primary PSEs may be associated with neural axis dissemination, although this complication occurs with low-grade lesions rather than anaplastic ependymomas. Anaplastic ependymomas are more likely to be reported with metastasis to lung, skin, and kidney rather compared to low-grade ependymomas [6]. Spinal ependymomas have an unusual propensity to spread extraneurally. This is particularly true for subcutaneous myxopapillary tumors that arise over the sacrococcygeal region [6,7]. It has been reported that pediatric patients are more likely to experience leptomeningeal dissemination and extraneural metastasis, and their disease course may be more aggressive than in adult patients [5].

Cases appear to be rare; most are reported as case reports or retrospective studies, and different histological grades of SPEs are often reported together, making the analysis of data on these tumors in pediatric patients difficult and complicating efforts to establish a standard protocol for management of SPEs. Gross-total resection (GTR) is the main treatment for SPEs, but several studies pointed out the impact of adjuvant treatment on progression of these tumors and survival rate; however, this is still debatable [4]. Here, we present the surgical treatment outcomes of 3 consecutive pediatric spinal ependymoma cases.

Material and Methods

Patiant population

This retrospective study was approved by the Medical Ethics Committee of our hospital under decision number 507/2015. Written informed consent was obtained from both parents of all patients for publication of their cases and accompanying images.

Medical records were retrospectively reviewed and we selected 46 patients with spinal ependymoma who were diagnosed and surgically treated at the Department of Neurosurgery of our hospital between the years 2004 and 2015. All the spinal ependymoma cases of children (patients age of under 18 years) are included in the current study (n=3). The patient characteristics, clinical presentation, surgical findings, and pre- and postoperative neurological functional assessments were evaluated using a modified neurological scoring system (Table 1) [8], and long-term outcome are discussed. The relevant literature concerning ependymomas in children is also reviewed.

Statistical analysis

Statistics presented hereafter are expressed as the mean ± standard deviation values together with the range in parentheses. Differences among groups were assessed with the one-way analysis of variance (ANOVA) using the SPSS 21.0 statistical package. Significance in the multivariate model was determined using a p value <0.05. All p values are given with odds ratios (OR). OR are presented with the 95% confidential interval (CI). All tests were 2-tailed.

Surgery

Under general anesthesia and using intraoperative neurophysiological monitoring (IONM), the patients were positioned prone using a supporting roll on each side. A paramedian vertical midline incision was made. After dissecting the paraspinal muscles, laminectomy or laminotomy was completed. Bilateral lamiotomy was done using high-speed drills or kerrison rongeurs. Then, ligamentum flavum and the adipose tissue were removed. The laminectomy or laminotomy were performed up to the tumor spanned levels. The operative microscope was

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Table 1. Neurological scoring system [8].

Score	Pain intensity	Sensory disturbance, dysesthesias	Motor weakness	Gait ataxia	Sphincter function
5	None	Normal	Full power	Normal	Normal
4	Slight, no medication	Present, not significant	Movement against resistance	Unsteady, no aid	Slight disturbance, no catheter
3	Tolerable w/ medication	Significant, function not restricted	Movement against gravity	Mobile w/ aid	Residual in urodynamic studies, no catheter*
2	Insufficient control w/ medication	Some restriction of function	Movement w/o gravity	Few steps w/ aid	Rarely incontinent
1	Severe despite medication	Severe restriction of function	Contraction w/o movement	Standing w/ aid	Frequent catheter
0	Incapacitating	Incapacitated function	Paralysis	Paralysis	Permanent catheter

* Modified. w/ – with; w/o – without.

brought in over operation field. The thecal sac was opened in the midline and tacked up bilaterally using strong sutures. After the dura was opened, a dorsal midline myelotomy was performed on any intramedullary ependymomas to reach the intramedullary region. After exposuring all nerve roots, spinal cord/filum terminale and arachnoid bands, the neurosurgeons distinguished neural tissues from tumoral tissue using the microscope and probe of IONM. In the intramedullary region, a cavitron ultrasonic surgical aspirator was used. The tumoral tissue was completely resected in 3 cases (2 primary and 1 drop seeding metastasis lesions). Although is thought to be easy to separate neural tissues from tumoral tissue in the syrinx cavity, it seems that in the syrinx cases it is difficult to achieve GTR. Tumor tissue was removed and, to preserve neurological functions, we were careful to avoid injuring the spinal cord. After hemostasis using serum physiologic water, duraplasty was performed using 5.0 absorbable sutures (particularly, in the last reported 10 cases in this series, to avoid CSF fistula after tight closure of dura, the surgeons used fibrin sealant products). The operation field was kept clean and the CSF circulation between the neural elements was preserved. In the laminotomy cases, the laminae were placed using strong non-absorbable suture to fix them again. All layers were closed appropriately with their anatomy.

Case reports (Table 2)

Case 1

An 11-year-old male was referred to our hospital with unsteady gait and left leg numbness of 7 weeks. His complaints were more severe at nights. His neurological examination was intact, except for global hypoesthesia on left leg. T1-weighted MR images demonstrated hyperintense areas, T2-weighted MR images showed iso-hyperintense areas, and there was a well-circumscribed intradural intramedullary lumbar (filum terminale) lesion measuring 12×24×33 mm at L1–L3 level (Figure 1A, 1B). The patient underwent GTR of the lesion using total L2 laminectomy. He recovered well and was discharged after 7 days without any complication. Histopathologically, the mass lesion was confirmed to be a MPE (WHO grade I). Yearly, MRI and neurological examinations were performed. On his doctor visit at 75 months, the patient presented with low back pain, bilateral lower-extremities numbness, and heavy walking. MRI showed a well-circumscribed intradural sacral lesion measuring 14×8.2×30.1 mm at S1–S2 level (Figure 2). The patient underwent GTR of lesion using total S1-2 laminectomy (Figure 3). He recovered well and was discharged after 3 days without any complication. Histopathologically, the mass lesion was confirmed to be a MPE (drop-seeding metastasis). The patient had complete resolution of his symptoms. The patient was doing well at his postoperative 98th/25th month doctor visit.

Case 2

A 16-year-old male presented to our out-patient clinic with low back and left leg pain of 2-year duration. His neurological examination was intact, except the straight leg raise (Lasegue) test was bilaterally positive at 60 degrees, and there was local tenderness on spinous processes of L1–L3. T2-weighted MR images were iso-hyperintense and demonstrated a well-circumscribed intradural extramedullary lumbar (cauda equine) lesion measuring 15x23x54 mm at L1–L3 level. The patient underwent GTR of the lesion using total L1–L3 laminoplasty. He recovered well and was discharged after 3 days without any complication. Histopathologically, the mass lesion was

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Table 2. Baseline clinical characteristics, clinical presentation and findings, surgical approaches, and outcomes of treatment in three operated pediatric patients.

No	Clinical presentation	Age/ sex	Location	Clinical findings	Surgery	Postoperative course; pathology	Survival after surgery
1	Unsteady gait, left leg numbness of 7 wks	11/M	Intradural-Intrame-dullary; Lumbar; L1–L3	Global left leg hypoesthesia	GTR using laminectomy and IONM	PO 7th day; recovered fully; MXE WHO grade I; PO 75th month presented with seeding metastasis (see below); No complication or local recurrence was recorded	Alive; 98 mns
	Low back pain, bilate-ral lower extremities numbness and heavy walking of 2 mns	17/M	Intradural-Extrame-dullary; Sacral; S1–S2	Bilateral straight leg test at 60 degrees	GTR using laminectomy and IONM	PO 3th day; recovered fully; MXE WHO grade I (drop seeding metastasis); No complication, local recurrence or neuroaxis dissemination was recorded	Alive; 25 mns
2	Low back and left leg pain of 2 yrs	16/M	Intradural-Extrame-dullary; Lumbar; L1–L3	Bilateral straight leg test at 60 degrees, local tenderness on L1–L3	GTR using laminoplasty and IONM	PO 3th day; recovered fully; MXE WHO grade I; No complication, local recurrence or neuroaxis dissemination was recorded	Alive; 63 mns
3	Difficulty of urination and breathing of 3 dys; heavy walking, low back and left leg numbness and weakness of 35 dys	9/F	Intradural-Intrame-dullary; Thoracic; T8–T10; (+Syrinx)	Urinary retention, 4/5 strength and global hypoesthia on the left leg, left Babinski reflex was no response and DTR of left leg were hyperactive	NTR using laminoplasty and IONM	PO 7th day; improved; Classic Ependymoma WHO grade II; She recovered with transient glob, and was discharged after rehabilitation she was doing well; No complication, local recurrence or neuroaxis dissemination was recorded	Alive; 17 mns

GTR – gross total resection; NTR – near total resection; dys – days; wks – weeks; mns – months; yrs – years; F – Female; M – Male; PO – postoperative; DTR – deep tendon reflexes; MXE – myxopapillary ependymoma; IONM – intraoperative neurophysiological monitorization.

confirmed to be a MPE (WHO grade I). Yearly, MRI and neurological examinations were performed. No recurrence was observed. The patient had complete resolution of his symptoms and was doing well on his postoperative 63th month doctor visit.

Case 3

A 9-year-old female was referred to our Emergency Department with urinary retention, difficulty of breathing (3-day duration), heavy walking, low back and left leg numbness and weakness lasting 35 days. Her neurological examination revealed 4/5 strength and global hypoesthesia on the left leg, left Babinski test was no response, and deep tendon reflexes of left side lower extremity were hyperactive. T1-weighted MR images demonstrated hyperintense and T2-weighted MR images showed an iso-hyperintense, irregular, diffuse, intradural, intramedullary thoracic lesion with several septae, measuring 24×33×46 mm at T8-10 level. The lesion up and down the side of the syrinx cavity extended from C5 to L1 levels (Figure 4). The patient underwent near-total resection (NTR) of the lesion using T8–T10 laminoplasty. She recovered with transient glob, and when discharged after rehabilitation was doing well. Histopathologically, the mass lesion was confirmed to be a classic ependymoma (WHO grade II). No adjuvant treatment was carried out for syrinx. The 3rd and 6th postoperative MRI (Figure 5) showed regress of syrinx cavities, and no progression or seeding metastasis were detected. On her 3-month doctor visits, she had complete resolution of her symptoms and was doing well on her postoperative 17-month doctor visit.

Results

This series included 1 female (33.3%) and 2 male patients (66.7%) between the ages of 9 and 17 years, with mean age 13.3±3.9 years. The basline clinical characteristics, location,

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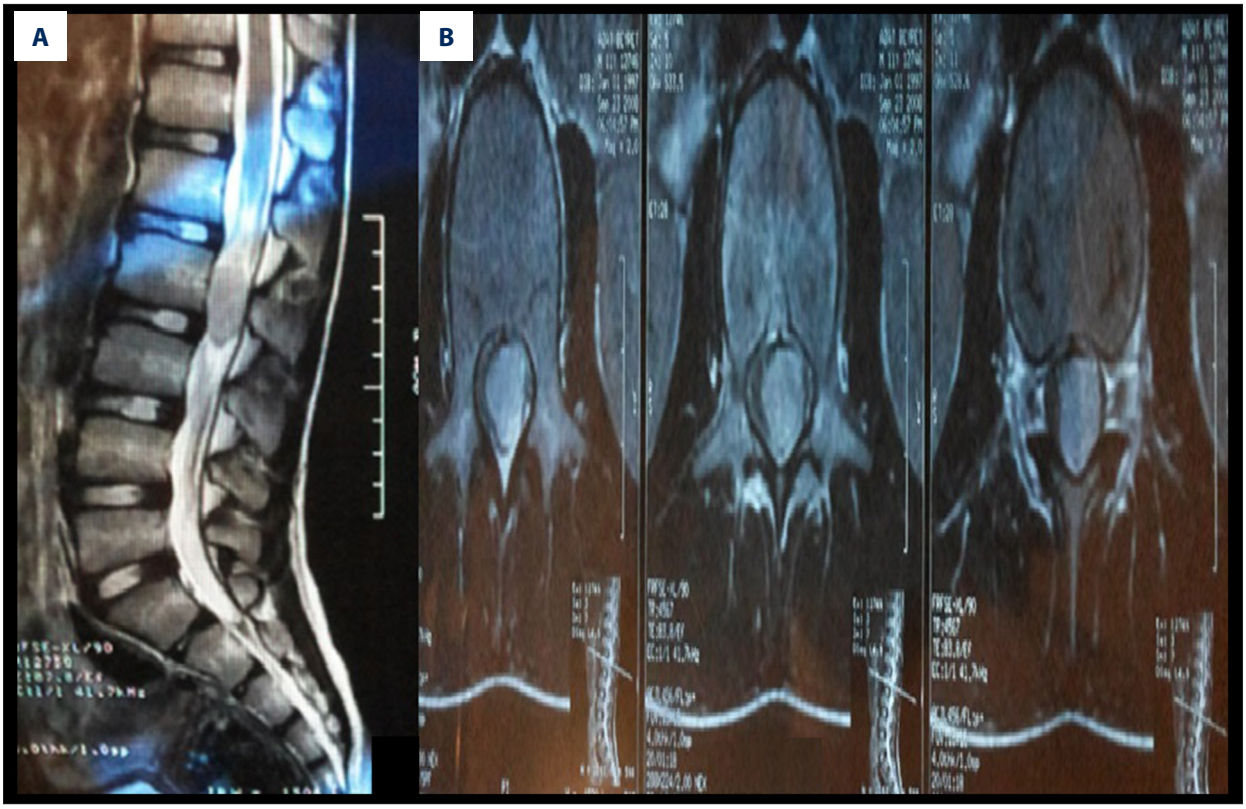

Figure 1. An 11-year-old male was referred to our hospital with unsteady gait and left leg numbness of 7-week duration; (**A**) Preoperative T2-weighted sagittal MRI demonstrated an iso-hyperintense, well-circumscribed, intradural, intramedullary, lumbar lesion measuring 12×24×33 mm at L1–L3 level; (**B**) T2-weighted axial MRI, note that the tumor is located intramedullary.

treatment, outcomes, and complications of treatment for all cases are given in Table 2. The mean preoperative course (prodrome) was 9.1±10.5 months (range, 35 days to 24 months). The most common location was the lumbar spinal cord (n=2). The most common presenting symptoms were lower-limb numbness and ataxic gaits, which were seen in 3 of 4 cases per each (including the drop-seeding metastasis case). Two tumors were located intradural-intramedullary and 1 was located intradural-extramedullary. GTR was achieved in 2 patients, and NTR was performed in 1 patient. No adjuvant treatment was received. The mean follow-up duration was 51.3±37.6 months, with a range of 17 to 98 months. No complications were recorded. According to the modified neurological scoring system, functional assessment of all patients at the latest follow-up evaluation showed that were good (Table 3). Two MPEs and 1 classic ependymoma were histopathologically diagnosed. At 6.3 years after the first operation, 1 patient presented with drop-seeding metastasis. The 45- and 51-month progression-free survival rates were 100% and 75%, respectively, and the 4-year survival rate was 100%. The mean length of hospital stay was 5.3±3.2 (range, 3–10) days. No recurrence, morbidity, or mortality was recorded. No patient had neurofibromatosis type 2. All our pediatric patients attended a normal schooling system.

Table 4 shows a comprasion between our spinal primary ependymoma adult and child patients who were treated surgically in the same institute and during the same time period. Two symptoms were significantly different between children and adults who had spinal ependymomas; 'gait impairment' symptom was more likely to appear in children than in adults (OR 15.4, P=0.026), while 'radicular (extremity) pain' symptom was more likely to appear in adults than in children (OR 0.01 P=0.026). The mean prodrome period was shorter in child than in adult patients (9.1±10.5 and 25.0±43 months, respectively) [P=0.006].

Although most studies report both children and adults, small numbers of children were included, and these numbers are insufficient to perform a meaningful review or statistical analysis, even if data are pooled. All distinguished SPE cases and series are given in Table 5.

Discussion

The occurrence of ependymomas is very rare in children younger than 10 years of age [9,10]. A review of the literature reveals that the most commonly reported primary SPEs

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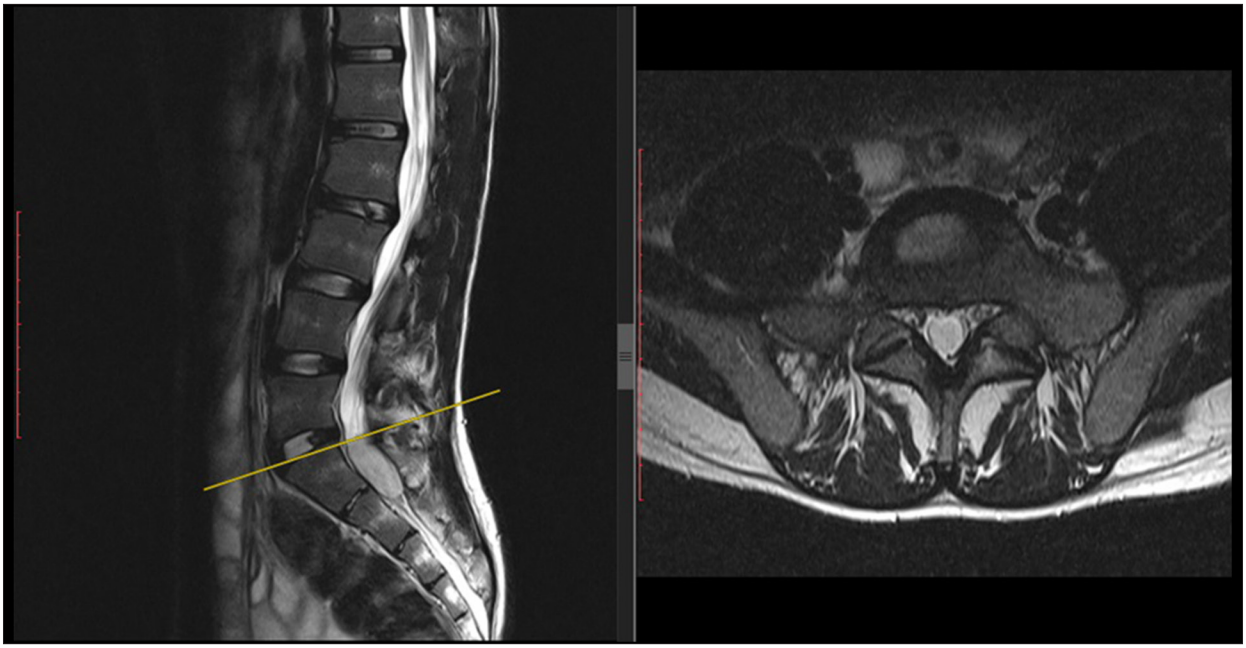

Figure 2. The same patient as in Figure 1. At 75 months after surgery, he presented with low back pain, bilateral lower-extremities numbness, and heavy walking. T2-weighted MRI showed a well-circumscribed intradural sacral lesion measuring 14×8.2×30.1 mm at S1–S2 level. Note that there is no local recurrence or residue at L1–L3 level; defects of L2 laminectomy.

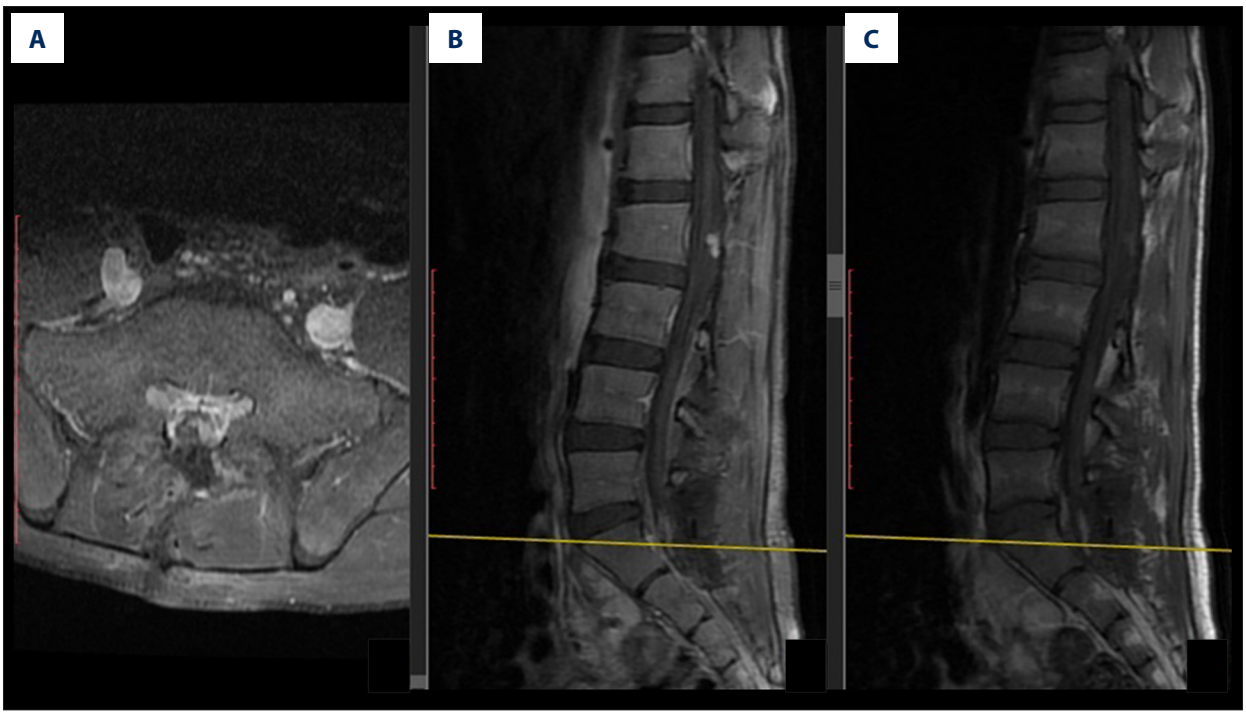

Figure 3. Second operation early postoperative contrast-enhanced T1-weighted MRI showed that GTR was achieved, as well as postoperative changes and defect of S1 laminectomy; (**A**) Axial contrast-enhanced T1-weighted MRI demonstrated postoperative changes and S1 laminectomy; (**B**) Sagittal contrast-enhanced T1-weighted MRI; (**C**) Sagittal T1-weighted MRI.

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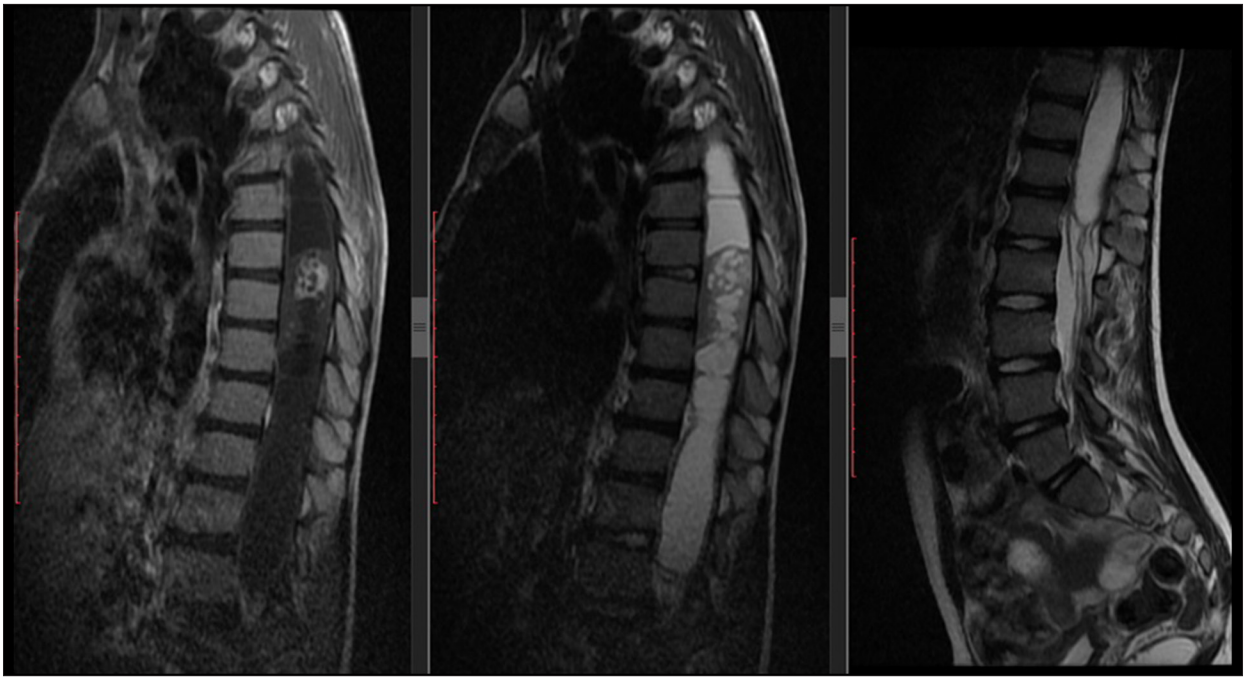

Figure 4. A 9-year-old female was referred to our Emergency Department with urinary retention, difficulty breathing, low back and left leg pain, numbness, and weakness. Preoperative T1-weighted MRI demonstrated hyperintense areas, T2-weighted MRI showed an iso-hyperintense irregular diffuse intradural intramedullary thoracic lesion with several septae, measuring 24×33×6 mm at T8–T10 level. The lesion up and down side syrinx cavities extends from C5 to L1 levels.

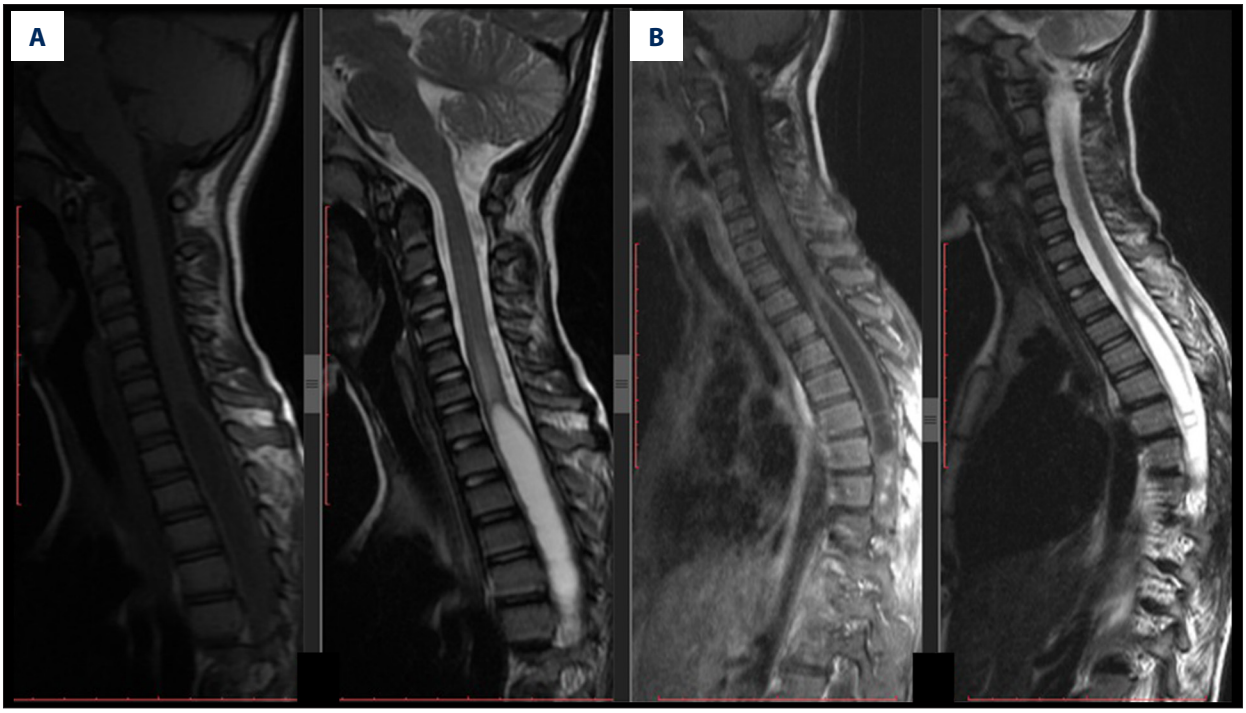

Figure 5. Postoperative 3- and 6-month MRIs of the same patient as in Figure 4. She underwent NTR of the lesion using T8-10 laminoplasty. No adjuvant treatment was carried out for the syrinx. Both MRIs showed regress of syrinx cavities, and no progression or seeding metastasis was detected; (**A**) Postoperative 3-month MRI, T1-weighted MRI in left side and T2-weighted MRI in right side; (**B**) Postoperative 6-month MRI, T1-weighted MRI in left side and T2-weighted MRI in right side. Note that the syrinx cavities were regressed without additional therapy.

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Table 3. Clinical outcomes according to neurological scoring system.

No*	Leg ± back pain intensity Preop/postop	Sensory disturbance, dysesthesias Preop/postop	Motor weakness Preop/postop	Gait ataxia Preop/postop	Sphincter function Preop/postop	Surgical outcome
1	5/5	3/4	5/5	3/5	5/5	Recovered good
	2/5	4/5	5/5	4/5	5/5	Recovered good
2	3/5	5/5	5/5	5/5	5/5	Recovered good
3	5/5	3/4	4/5	4/5	3/4	Improved**

* Case No in this table referes to the same case No in Table 2. ** Improved: if the patient's complaints decreased but did not mean full recovery.

Table 4. Comparison between our spinal primary ependymoma adult and child patients who were treated surgically in same institute and during the same period.

	Children	Adults	Comments
Numbar of Patients	3 (1F; 2M)	43 (20F; 23M)	OR 1.74, CI=0.15–20.65; P=0.57
Age	13.3±3.9 (9–17) yrs	39.2±12.0 (22–67) yrs	–
F:	9 yrs	40.3±13.2 (22–67) yrs	–
M:	14.67±3.2 (11–17) yrs	38.3±11.1 (22–55) yrs	–
Complaints			
– Radicular (extremity) pain	25% (1/4*)	83.7% (36/43)	OR 0.01, CI=0.01–0.72; *P=0.026*
– Local pain	50% (2/4)	79% (34/43)	OR 0.25, CI=0.03–2.02; P=0.21
– Loss of sensation	75% (3/4)	41.9% (18/43)	OR 4.17, CI=0.4–43.4; P=0.28
– Motor deficit	25% (1/4)	27.9% (12/43)	OR 0.86, CI=0.1–9.1; P=0.7
– Gait impairment	75% (3/4)	16.3% (7/43)	OR 15.4, CI=1.4–170.7; *P=0.026*
– Urination disturbance	25% (1/4)	4.7% (2/43)	OR 6.8, CI=0.47–98.8; P=0.24
– Difficulty of breathing	25% (1/4)	0	P=0.085
– Neuropathic pain	0	4.7% (2/43)	P=0.84
– Headache	0	2.3% (1/43)	P=0.91
– Neck pain	0	2.3% (1/43)	P=0.91
Mean prodrome	9.1±10.5 mn (35 dy–24 mn)	25.0±43 mn (10 dy–20 yr)	*P=0.006*
Location			
– Cervical (all Int)	0	10 (7F; 3M) [23.3%]	–
– Thoracic [Int (4); Ext (2)]	1 (F) [33.3%]	5 (3F; 2M) [11.6%]	–
– Lumbar [Int (3); Ext (25)]:	2 (M) [66.7%]	26 (10F; 16M) [60.5%]	–
– Sacral	0	0	–
– Multiple (all Ext)	0	2 (M) [4.6%]	–

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Table 4 continued. Comparison between our spinal primary ependymoma adult and child patients who were treated surgically in same institute and during the same period.

	Children	Adults	Comments
Treatment choice			
– GTR (n=25)	2 [Int (1); Ext (1)] 66.7%	23 [Int (5); Ext (18)] 53.5%	OR 1.58, CI=0.13–18.8; P=0.6
– NTR (n=12)	1 [Int (1)] 33.4%	11 [Int (4); Ext (7)] 25.6%	
– STR (n=5)	0	5 [Int (4); Ext (1)] 11.6%	
– GTR+RT (n=1)	0	1 [Int (1)] 2.3%	
– STR+RT (n=3)	0	3 [Int (2); Ext (1)] 7.0%	
Surgical outcome			
– Recovered	2 (GTR) [66.7%]	27 (17: GTR; 6: NTR; 4: STR) [62.8%]	OR 1.1; P=0.72
– Improved	1 (NTR) [33.3%]	8 (3: GTR; 4: NTR; 1: STR) [18.6%]	
– Unchanged	0	4 (2: GTR; 2: STR) [9.3%]	
– Worsened	0	3 (2: GTR; 1: STR) [7.0%]	
– Dead	0	1 (NTR) [2.3%]	
WHO grading			
– Grade I	2 MPE(2M) [66.7%]	18 MPE (7F; 11M) [41.9%]	–
– Grade II	1 (F) [33.3%]	23 (11F; 12M) [53.5%]	
– Grade III	0	2 (F) [4.6%]	
Recurrence	0 (1F; 1M) [5.1%]		
Neuroaxis dissemination	(n=1) 33.3%	0	
Mean LOS (dys)	5.3±3.2 (3–10)	8.7±9.6 (2–64)	P=0.36
Mean follow-up (mns)	51.3±37.6 (17–98)	90.2±50.9 (14–156)	–
4-year PFS rate	66.7% (1 Dis case)	90.5% (4 recurrent cases)	–
4-year OS rate	100%	97.7% (related surgery)	–

* Cases in children included drop seeding metastasis case. F – Female; M – Male; Int – intradural-intramedullary; Ext – intradural-extramedullary; dy – days; mn – months; yr – years; GTR – gross total resection; NTR – near total resection; STR – subtotal resection; RT – postoperative radiotherapy; LOS – length of hospital stay; PFS – progression-free survival; OS – overall survival.

are MPEs (WHO grade I) followed by classic/typical ependymomas (WHO grade II) and anaplastic ependymomas (WHO grade III). Out of 441 SPE cases, 197 MPE were reported in the literature (included all cases with sufficient details between 1945 and 2016). Anaplastic ependymomas are extremely rare in pediatric patients (only 24 cases were reported). In 1932, Kernohan [11] described MPE as a separate histopathological subgroup of ependymoma, and since then several case and serial reports have been reported in the pediatric population (Table 5). Kernohan found that areas of mucinous degeneration within the vascular connective tissue cores of papillary tumors helped to distinguish MPE from other subgroups of ependymoma [11]. Ependymomas may occur intra- or extramedullary. In adults, intramedullary ependymomas are most commonly observed in the cervical spine but there is no such data on pediatric patients. Lumbar spine ependymomas mostly consist of those originating from the filum terminale (FT) or cauda equine (CE). MPEs are common in the lumbar region and are extramedullary tumors. However, ependymomas grade II and grade III are most common in the cervical region followed by the thoracic region. In the present study, 3 cases (included the drop-seeding metastasis) were located in the lumbosacral

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Table 5. Reported cases of pediatric (<18 years) spinal ependymoma in the literature (to the best of our knowledge).

No	Lead Author; year	Mean age/sex	No of Pts	Location	Prodrome (mns)	Treatment	Pathology and postoperative course	Mean of FU (mns)
1	Nisenson [24]; 1945#	12/M	1	N.M	N.M	STR+RT	Ependymoma (NOS); No Rec or Dis	42
2	Dereymaeker [24]; 1962#	14/F	1	N.M	N.M	GTR	Ependymoma (NOS); No Rec or Dis	60; Dead
3	Hendren [25]; 1963*#	16/M	1	SC	108	Resection*+RT + ChT	Ependymoma (NOS); Rec+Dis (Extraneural met: inguinal lymph nodes, pelvis)	72
4	Sloof [17]; 1964	14/M	1	L (CE)	N.M	STR+RT	MPE; 3 times Rec at 2, 4 and 8 years. Dis(+)	150; Dead
5	Anderson [6]; 1966	9/1M: 2F	3	SC (3)	N.M	GTR (2); STR+RT (1)	MPE (3); Rec (1): in STR+RT; no PO details for 1 pt.	45
6	Probhaker [25]; 1969*#	0.83/M	1	SC	Since birth	Resection*	Ependymoma (NOS); Rec (+)	24
7	Rubinstein [26]; 1970	17/F	1	L (FT)	N.M	STR+RT	Ependymoma (NOS); Rec afte 3 yrs, Dis after 6 yrs; Extraneural metastasis (Lungs, pleural and lymph nodes): DOD after 29 yrs	348; Dead
8	Wolf [27]; 1972**#	4/M	1	SC	N.M	Resection*	Ependymoma (NOS); Rec+Dis (extraneural metastas: Lungs and inguinal lymph nodes)	228
9	Payne [28]; 1973	11/F	1	L (CE)	Acute; Pos-ttrauma	Resection*+RT	MPE; No Rec or Dis	36
10	Scharrer [25]; 1974*#	7/F	1	SC	N.M	Resection*	Ependymoma (NOS); Rec/Dis (N.M)	N.M
11	Scott [29]; 1974	17/M	1	L	N.M	STR+RT	MPE; Rec (after 5 yrs); Dis (No)	276
12	Ammerman [24]; 1975#	16/F	1	N.M	N.M	GTR	MPE; No Rec or Dis	24
13	Cameron [30]; 1976	13/F	1	Multiple (supra-, infratentorial and spinal)	N.M	Resection* (4 operations in 4 yrs)	MPE; Rec? and Dis? Or multiple at presentation	78
14	Fisher [24]; 1977#	(6-14)/4M: 4F	8	N.M	N.M	GTR (5); STR+RT (3)	Ependymoma grade I (5); Ependymoma grade II (2); Ependymoma NOS (1); Dead at PO 36 mn	43.5
15	Mavroudis [31]; 1977	7/M	1	L (CE)	N.M	GTR	MPE; Rexploration after 2yrs (No lesion); Rec (after 24 yrs); Extraneural metastasis to lungs after 29 yrs)	348
16	Mork [10]; 1977	13/2M: 3F	5	N.M	N.M	NOS	MPE and Classic ependymoma (NOS)	N.M
17	Bale [25]; 1980*#	4/F	1	SC	1	Resection*	Ependymoma (NOS); Rec/Dis (No)	168
18	Mork [32]; 1980	12/M	1	TL	6	STR+RT	Anaplastic ependymoma; Rec (after 2 yrs), Dis (after 6 yrs)	78

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Table 5 continued. Reported cases of pediatric (<18 years) spinal ependymoma in the literature (to the best of our knowledge).

No	Lead Author; year	Mean age/sex	No of Pts	Location	Prodrome (mns)	Treatment	Pathology and postoperative course	Mean of FU (mns)
19	Morris [33]; 1983	10/F	1	L (CE)	60	NTR+RT	Ependymoma grade I; Rec and Dis (Extraneural metastasis: lungs and hip) after 6 yrs	144
20	Chan [34]; 1984	10.6/5M: 2F	14*	L (7) [4: CE+ 3: FT]	10.6	GTR (5); STR+RT (2)	MPE (7); Rec in 2 GTR;Dis in 2 pts	105.6
21	Helwig [35]; 1984	17/F	1	SC	4	Resection*	MPE; Rec+Dis after 6 mns (DOD)	60; Dead
22	Matsuo [36]; 1985	11/F	1	SC	N.M	GTR	MPE; Rec/Dis (No)	7
23	Sonneland [18]; 1985	N.M/10M: 5F	15	L (15)	N.M	NOS	MPE (15); Rec (5 pts); 3 pts DOD	205.1
24	West [37]; 1985	7.9/4M	4	T (2); L (CE) (1); Extrasacral (CS) (1)	N.M	STR+RT+ChT (1); STR+RT (1); Bx+RT (2)	Ependymoma (Low grade) (4); No Rec or Dis	102.5
25	Ciraldo [38]; 1986	0.83/2M: 3F	5	SC (5)	Since birth	GTR (5)	MPE (5); Rec/Dis (No)	26
26	Chou [27]; 1987**#	12.5/1M: 1F	2	SC (2)	N.M	Resection* (2)	MPE (2); Rec or Dis (N.M)	N.M
27	Murphy [27]; 1987**#	0.09/M	1	SC	Since birth	N.M	MPE; Rec/Dis (No)	N.M
28	Di Marco [39]; 1988	17/F	1	L	N.M	STR+RT	Low grade ependymoma; Rec after 169 months (treated)	182
29	Kramer [25]; 1988	15/M	1	SC	N.M	GTR	MPE; Rec (twice)+Dis (extraneural: inguinal, lymph nodes and skin)	240
30	Pulitzer [40]; 1988	0.6/3M: 1F	4	SC (4)	Since birth	Resection*	MPE (4); Rec/Dis (No)	7-40
31	Naidu [41]; 1989	14/M	1	L (CE)	6	Resection*+RT	Ependymoma (NOS); Dis after one yr, Rec after two yrs	24
32	Fujiyama [42]; 1990	6/F	1	TL	6	GTR+RT	Anaplastic Ependymoma; Dis and Rec after 3 yrs	89
33	Le Marc'hadour [43]; 1991	14/F	1	SC	6	Resection*	MPE; Rec/Dis (No)	24
34	Wen [44]; 1991	12/NOS	2	L (2)	18 (Median)	STR+RT (2)	MPE (2); Both had Dis in spine at 32 and 48 mns; both DOD at 209 and 110 mns.	159; Dead
35	Gupta [45]; 1992	1.5/M	1	SC	Since birth	NOS	MPE; since birth, Dis (metastasis to inguinal lymph nodes) PO aggressive tm	18
36	Serour [46]; 1992	11/M	1	SC	N.M	GTR	MPE; Rec/Dis (No)	12
37	Clover [47]; 1993	16/2M: 1F	3	Multiple [LS+CE] (2); LS (1)	24 (Median)	STR+RT (2); STR (1)	MPE (3); Rec (1) after 3.2 yrs in STR+RT (F-U: 6 yrs -AWD)	52
38	Gagliardi [24]; 1993	13.5/3M: 1F	4	L [FT] (4)	12	GTR (2); STR+RT (1); STR (1)	MPE (4); 1 Rec in STR	93.3

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Table 5 continued. Reported cases of pediatric (<18 years) spinal ependymoma in the literature (to the best of our knowledge).

No	Lead Author; year	Mean age/sex	No of Pts	Location	Prodrome (mns)	Treatment	Pathology and postoperative course	Mean of FU (mns)
39	Lunardi [48]; 1993	10-16/ NOS	9	Int [NOS] (9)	NOS	GTR+RT (3); GTR (4); STR+RT (2)	Low grade ependymoma (NOS) (9); Rec (2)	48 [Median]
40	Ross [16]; 1993	10.7/2M: 1F	3	N.M	N.M	GTR+RT (3)	MPE (3); Rec/Dis (No)	93.3
41	Waldron [23]; 1993	11.5/NOS	4	LS (4)	N.M	Resection*+RT (4)	MPE (1), Classic Ep (1), Anaplastic (1), NOS (1); Rec (1); Dis (3): 2 of Dis were DOD; 1 Reopere+RT (30 yrs: NED); 1 Reopere (5 yrs: NED)	114.8
42	Botti [49]; 1994	10.5/N.M	1	SC	4	GTR	MPE; Rec/Dis (No); but the pt was presented with Dis.	N.M
43	Do-Dai [50]; 1995	12/F	1	CT	5 dys	Bx+ChT	MPE; Rec: (+), Dis (+); Dis at prersentation	6
44	Mottl [51]; 1997	N.M/F	1	L (FT)	N.M	Partial resection+RT	WHO grade I Ependymoma; Rec: No, Dis: No; alive	N.M
45	Nagib [52]; 1997	9.3/1M: 2F	3	TL (FT) [1]; L (CE) [2]	1.7	GTR (3)	MPE (3); Rec: No, Dis (1)	42.7
46	Ilhan [53]; 1998	8/M	1	SC	3	GTR	MPE; Rec/Dis (No)	20
47	Lonjon [54]; 1998	14/11M: 9F##	32##	CM (2); C (4); CT (5); T (8); TL (1); 12 MPEs were located in LS	5–84	GTR+RT (1); GTR (13); STR+RT (5); STR (1); No details about treatment of MPEs	MPE (12), low grade ependymoma (17), high grade ependymoma (3); Rec (2) after 2 and 3 yrs (both were treated with STR+RT; Dis (4): 2 of them were anaplastic (In 1 GTR+R, 1 GTR and 2 STR+RT). 11 presented with syrinx, 3 pts had NF Type 2. 3 operations for kyphoscoliosis, 1 operation for shunting hydrocephalus	67 [Median] (25–177)
48	Graf [55]; 1999	15/M	1	TL (CE)	3	GTR	MPE; Rec (+): after 32 yrs; Neuroaxial Dis to Lungs and Liver (+) after 37 yrs and dead	480; Dead
49	Johnson [56]; 1999	7/M	1	SC	18	GTR	Grade II ependymoma; No Rec/Dis	96
50	Ohata [57]; 1999	16/2M	2	CM (1); CT (1)	N.M	GTR (1); STR+RT (1)	Grade II (1) and grade III (1); No Rec/Dis.	86.2
51	Aktug [27]; 2000	5/M	1	SC	6	GTR	MPE; Rec/Dis (No)	36
52	Chinn [13]; 2000	11/2M: 1F	3	Multiple (2); L (1)	11.3	GTR+RT (1); GTR (1); STR+RT (1)	MPE (3); Rec (1) in GTR; Dis in all pts (3): 2 at presentation, one after 7 mns of GTR	24
53	Constantini [9]; 2000	<21/NOS	26	Int (NOS)**	11.6**	NOS**	Ependymoma (19), MPE (7); 10-yrs survival rate is 86%; 4-yr PFS rate is 75%**	85.1**

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Table 5 continued. Reported cases of pediatric (<18 years) spinal ependymoma in the literature (to the best of our knowledge).

No	Lead Author; year	Mean age/sex	No of Pts	Location	Prodrome (mns)	Treatment	Pathology and postoperative course	Mean of FU (mns)
54	Merchant [58]; 2000	8.6/3M: 5F	8	CM (1); CT (1); TL (3); L (1); LS (2)	N.M	GTR+RT+ChT (1); STR+RT+ChT (4); STR+RT (3)	MPE (4), ependymoma (4); Rec (2): in STR+RT+ChT both were ependymomas; Dis (4): 3 were MPEs which were treatd with STR+RT+ChT (2) and STR+RT (1), 1 ependymom which was treated as GTR+RT+ChT	85.9
55	Nishio [59]; 2000	15/2M	2	C+T (1); L (1)	12.5	GTR	Ependymoma grade II/III (NOS); Rec/Dis (No)	137
56	Gelabert-Gonzalez [60]; 2001	15/F	1	FT (2 tms: L+S)	2	GTR	MPE (both lesion); Rec/Dis (No)	6
57	Helseth [61]; 2001	8.76/3M: 2F	5	L (CE): 5	N.M	GTR (2); STR+RT (3)	MPE (3), grade II (2); Dis (No); Rec (2): 1 GTR, 1 STR+RT	164
58	Hirose [62]; 2001	14.1/7M: 2F	9	C (2); L (1); CE (6)	N.M	N.M	MPE (6), Ependymoma grade II (3); Rec and Dis: N.M (Genetic study)	N.M
59	Hanbali [63]; 2002	17/M	1	T+L (Multiple)	N.M	GTR (both tms)	Grade II ependymoma; Rec/Dis (No)	31
60	Goto[64]; 2003	12.5/2M	2	CM (1); CT (1)	13	GTR (1); STR+RT (1)	Ependymoma (NOS); Rec/Dis (No)	206.5
61	Hallacq [15]; 2003	13/M	1	L+S (Multiple)	5	GTR	MPE; Rec/Dis (No)	60
62	Wolf [65]; 2003	1.5/F	1	TL	0.5	GTR	MPE; Rec/Dis (No)	33
63	Peker [66]; 2004	9/F	1	C (Int)	4	GTR	Low grade ependymoma (NOS); Rec/Dis (No)	108
64	Sebire [67]; 2004	13/F	1	SC	N.M	Resection*	MPE; Rec/Dis (NOS).	N.M
65	Fassett [14]; 2005	11.2/4M: 1F	5	L (5)	18.7	GTR+RT (1); GTR (1); STR+RT (3)	MPE (5); Rec (1): STR+RT; Dis (4).	58.2
66	Lin [68]; 2005	12/M	1	C	N.M	GTR	Ependymoma grade II; Rec/Dis (No)	48
67	Tubbs [69]; 2005	2/F	1	S	6	GTR	MPE; Rec/Dis (N.M)	N.M
68	Akyurek [22]; 2006	15/NOS	2	TL (1); L (1)	N.M	GTR+RT (1); GTR (1)	MPE (2); Rec (2): GTR+RT at 15 mns and GTR at 20 mns	131
69	Bagley [7]; 2007	12.6/10M: 4F	14	T (1); TL (4); L (1); LS (5); S (1); Multiple (2)	23.2	GTR (7); STR+RT (2); STR (2); Resection* (1); Bx+RT (1); Bx (1)	MPE (14); Rec (11): in 4 GTR, 2 STR+RT, 2 STR, 1 Bx+RT, 1 Bx, 1 Resection*; Dis (6)	63.7
70	Mridha [70]; 2007	13/M	1	TL	18	GTR	MPE; Rec and Dis (1) (After 36 mns and 38 mns, repectively)	38
71	Jatana [71]; 2008	11/M	1	N/A	N/A	GTR	MPE; Rec and Dis (1); NF-2 pt	N/A

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No	Lead Author; year	Mean age/sex	No of Pts	Location	Prodrome (mns)	Treatment	Pathology and postoperative course	Mean of FU (mns)
72	Kabler [72]; 2008	16/M	1	L	N.M	GTR	MPE; No Rec or Dis	1.5
73	Cho [73]; 2009	16/M	1	C (Int)	N.M	Bx+RT (No response) thenGTR	Grrade III Ependymoma; No Rec or Dis. (After 6 mns follow-up had lost)	6
74	Al-Halabi [12]; 2010	15.4/5M: 2F	7	TL (1); L (5); LS (1)	12	GTR+RT (3); GTR (1); STR+RT (2); Bx then STR+RT after progression(1)	MPE (7); Rec after STR+RT (1); Rec+Dis (2): after GTR+RT; presentation with cranial and spinal ependymomas (2)	78
75	Benesch [4]; 2010	13.6 [median]/ 12M: 17F	29	CM (1); C (3); T (2); LS (6); 2 regions (14); >2 regions (3)	2 [Median]	GTR+RT+ChT (3); GTR+RT (5); GTR (9); STR+RT+ChT (2); STR+RT (2); STR+ChT (2); STR (3); Bx+RT+ChT (2); Bx+RT (1)	MPE (6), grade II (17), grade III (6); 1 pt grade III died PO65.mn; progressive disease or relapse: 2 GTR (both are MPEs) and 5 less than GTR (2: MPEs, 2: typical and 1: anaplastic ependymomas)	50.4 [Median]
76	Dulai [74]; 2010	8/F	1	T (Int)	N.M	STR then GTR	MPE; Rec (PO 2 yrs)+Dis (PO 5 yrs)	108
77	Kaner [75]; 2010	17/F	1	CT (1)	23.2	GTR	Classic Ependymoma; No Rec or Dis	14
78	Moon [76]; 2010	11/M	1	Multiple (L+S)	4	GTR+RT	Classic Ependymoma; No Rec or Dis	36
79	Boström [77], 2011	12/M	1	TL	N.M	GTR+RT	Anaplastic ependymoma; No Rec or Dis	30
80	Chakraborti [78], 2012	0.92/F	1	SC	1	GTR	MPE with anaplastic component; Rec and Dis after 6 weeks	13.5
81	Choi [79]; 2012	13.7/1M: 2F	3	L (1); LS (1); T2-S1 (1)	8.5	GTR (1); STR+RT (1); Bx+RT (1)	MPE (3); No Rec; Dis (1): Bx+RT	13
82	Stephen [80]; 2012	13/11M: 6F	17	C (1); C+T (1); T (3); L (9): [FT (5)+CE (1), Int (3)]; LS (2); S (1)	9.5	GTR+RT (5); GTR+Proton (1); GTR (9); STR+RT (2)	MPE (7), ependyomoma (9), anaplastic (1); Rec (4): all are MPEs (Rec after an average of 37.8 mns). 1 pt had NF Type 2 (Rec in primary site).	58.8 (4–203)
83	Agbahiwe [21]; 2013	14.3/9M	9	TL (2); L (4); LS (2); S (1)	N.M	GTR+RT (1); GTR (4); STR+RT (3); STR (1)	MPE (9); Dis in GTR+RT (1), and 2 Rec in GTR and STR	87 [Median] (9–317)
84	Becco de Souza [81]; 2013	13/M	1	L	3	GTR	MPE; Rec/Dis (No)	2
85	Liu [5]; 2013	11.5/3M: 1F	4	C (3); CT(1)	2.7	GTR+RT+CT (2); GTR (1); STR (1)	Anaplastic (4); Rec in 1 STR (dead after 23 mns)	50.7 for 3 pts, 4th (N/A)
86	Pedziwiatr [82]; 2013	13.3/15M: 13F	28	L (CE) (17); Other spinal (8); Multiple (3)	3-60	GTR+RT (18); GTR (2); STR+RT (2); Partial resection+RT (4); Bx+RT (2).Out of them 3 recieved ChT	MPE (13), Typical ependymoma (12), Anaplastic ependymoma (3); DOD (5): Rec (2), Rec+Dis (3): Out of them 2 anaplastic; DOC (1) after GTR	104 [Median] 36–300

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Table 5 continued. Reported cases of pediatric (<18 years) spinal ependymoma in the literature (to the best of our knowledge).

No	Lead Author; year	Mean age/sex	No of Pts	Location	Prodrome (mns)	Treatment	Pathology and postoperative course	Mean of FU (mns)
87	Cimino[83]; 2014	10.3/6M: 5F	11	Extradural (7): [SC (5)+ Lower back (1)+ Pelvis (1)]; L (2); LS (2)	N.M	GTR (11)	MPE (11); Rec in 2 Lumbar tms followed-up for 5 and 10 years; No Dis	N.M
88	Khalatbari [84]; 2014	15/M	1	TL	Acute	GTR	MPE; Rec/Dis (No)	48
89	Lundar [85]; 2014	10.9/7M: 3F	10	TL (1); L (7); LS (2)	N.M	GTR (6); STR+RT (4)	MPE (6), Ependymoma grade II (2), grade III (2); Rec (1): MPE/GTR; Dis (1): MPE/STR+RT; Rec+Dis (3): 1 MPE/STR +RT, 1 Ep gr II/GTR and 1 Ep gr III/GTR	268
90	Pencovich [86]; 2014	15.5/5M: 1F	6	C (1); T+TL (1); T+L+LS (1); L+S (3)	10	GTR (2); STR+RT (2); STR (2)	MPE (5); Ependymoma grade II (1); All pts presented with regional metastasis (Multiple); No Rec or Dis	55
91	Lin [87]; 2015	11.9/30M: 34F	64	Spinal cord (62); CE (2)	N.M	GTR+RT (4); GTR (17); STR+RT (11); STR (5); No resection [only RT] (23); Unknown (4)	Grade II ependymopma (64); No details about Rec or Diss. [The patients were reported here thought to be duplicated in previous published studies]	110.4 [Median]
92	Current study; 2018	13.3/2M: 1F	3	T (1); L (2)	9.1	GTR (2); NTR (1)	MPE (2) and Classic ependymoma (1); Dis (1): GTR (MPE)	51.3

AWD – alive with disease; Bx – biopsy; C – cervical; CE – *Cauda equine*; ChT – chemotherapy; CM – cervicomedullary; CT – cervicothoracic; Dis – neuroaxis dissemination; DOC – dead of other causes; DOD – dead of disease; dys – days; F – Female; FT – filum terminale; GTR – gross total resection; L – lumbar; LS – lumbosacral; M – male; mns – months; N/A – not available; NED – no evidence of disease; NF – neurofibromatosis; N.M – not mentioned; No – number; NOS – non otherwised specifised; PFS – progression-free survival; PO – postoperative; pts – patients; Rec – recurrence; Resection* – surgical resection borders had not been defined; RT – radiotherapy; S – sacral; SC – sacrococcygeal; STR – subtotal resection; T – thoracic; TL – thoracolumbar; wks – weeks; yrs – years. * Chan et al. [34] series contain 14 pediatric ependymoma patients, while only details of 7 patients of childhood MPE were reported. ** Constantini et al. [9] reported 164 patients 21 years of age and younger in whom intradural-intramedullary tumors were resected. Their study is not specific for ependymomas. # We could not reach to the original studies, the data included in this table about the studies, which were indicated with (#), were obtained from Gagliardi et al.'s study [24]. ## Lonjon et al. [54] series contain 32 pediatric ependymoma patients, while only details of 20 patients of childhood ependymomas were reported, 12 MPEs were excluded. *# The data of these studies were obtained from Kramer et al. study (1988) [25]. **# The data of these studies were obtained from Aktug et al. study (2000) [27].

region and all of their histopathological examinations showed that they were WHO grade I (MPEs). Only 1 case was histopathologically diagnosed as classic ependymoma WHO grade II and was located in the thoracic spine and coexisted with the syrinx cavity.

Mork and Loken published one of the first reported series of central nervous system ependymomas, which included ependymoma cases in Norway over a 22-year period from 1953 to 1974. They found few spinal cord ependymomas in children; of the 53 spinal ependymomas, only 5 patients (2 boys and 3 females, aged 3–19 years old) were reported [10]. The male preponderance in SPEs is well-established in all previously published reports [5,6,12–19]. The male predominance is demonstrated by the fact that 66.7% (n=2) of our patients were males, in line with the previous reports.

In our series, presentation symptoms of spinal primary ependymomas differed between children and adults (Table 4). In children, the most common presenting symptoms were lower-limb numbness and ataxic gait, which were seen in 3 of 4 cases (including the drop-seeding metastasis case). However, in adult

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patients, radicular and local pain were the most common presenting symptoms. Al-Halabi et al. [12] reported that the most common presenting symptom was low back pain, which was reportedin up to 100% of patients, followed by urination symptoms (reported in 3 out of 7 patients) and lower-limb weakness (reported in 2 patients). The mean prodrome period was shorter in child than in adults (9.1±10.5 and 25.0±43 months, respectively) [P=0.006]. It appears that the good surgical resection outcomes in our series were related to the short prodrome in pediatric patients. Logically, symptoms in children appear early because the space in the thecal sac is limited.

Maximal resection of the lesion for GTR to preserve neurological function is the optimal treatment for all spinal tumors such as schwannomas and ependymomas. The consensus among pediatric neurosurgeons that GTR of SPEs is the choice of treatment regardless of location, age, and WHO grade of the tumor. The extent of resection depends on the preservation of the neurological functions; therefore, especially in the pediatric population, IONM will guide neurosurgeons to maximal borders up to GTR. Nowadays, the adverse effects of radio- and chemotherapy are well known. To avoid such adverse effects, the general view is that early RT should be avoided in children with WHO grade II spinal ependymomas, irrespective of the extent of resection, but is indicated in anaplastic (WHO grade III) spinal ependymomas after STR, NTR, or GTR [9]. GTR (i.e., resection of the tumor until reaching a neural structure with absence of any residual tumor on early postoperative imaging) for intradural-intramedullary spinal ependymomas is difficult, especially for tumors coexisting with syrinx cavities. In these conditions, we recommend subtotal (STR) or NTR (i.e., decompressing the neural structures next to the tumor and excising more than 75% for STR or 95% for NTR of tumor mass, leaving parts of the tumor attached to the neural tissues or their neurovascular structures). A systematic review by Feldman et al. [20] suggested that STR with RT in children may be associated with better outcomes than is STR with RT in adults. In the present series, GTR was achieved in 3 cases (included the drop-seeding metastasis case), while NTR was achieved in 1 female patient who had syrinx coexisting with thoracic spine ependymoma.

Although almost all SPEs are histopathologically benign tumors, WHO grade I and II primary spinal ependymomas have high local recurrence and neuroaxis dissemination rates, in particular MPEs, even though they are benign tumors classified as WHO grade I. Therefore, in management of SPEs, use of RT, which has as definite impact on progression of these tumors as proven in larger series [12,19,21,22], is still controversial. Al-Halabi et al. suggested GTR alone provided suboptimal disease control in MPE, while RT resulted in control of residual, metastatic, and/or recurrent pediatric MPEs [12]. Akyurek et al. suggested that MPE patients who are less than 35 years old are at higher risk of local recurrence and distant metastasis compared counterparts older than 35 years [22]. Two studies concluded there is no significant influence on the extent of resection to control local recurrence or distant metastasis [22,23]. However, both studies reported serious postoperative RT complications. In our patients, no adjuvant treatment was received. At 6.3 years after the first operation, 1 patient presented with drop-seeding metastasis. Thus, 45- and 51-month progression-free survival rates were 100% and 75%, respectively, while the 4-year survival rate was 100%. No recurrence, morbidity, or mortality were recorded. All our pediatric patients attended a normal schooling system.

Surgical morbidity is a serious complication in SPEs. Surgical morbidity was reported in most relevant studies; for example, Constantini et al. [9] reported spinal deformity is a common and serious complication associated with surgical treatment of spinal cord tumors. Although our series was relatively small and we cannot make generalization, we did not have any serious complications in our pediatric patients. Our female patient had the same transit glob (retention of urination); however, she improved, as shown in Table 3. Our review of the literature found that several studies suggested age, histology, previous surgery, tumor size, location, tumor consistency, and the extent of resection are not correlated with surgical morbidity, while removing 2 or more laminae can lead to spinal deformity, especially in children [8,9].

Our data support the evolving literature which suggests that no accepted molecular prognostic factors that can be used to predict surgical outcome for spinal ependymomas. Age, histology grade, and the MIB-1 index, a marker of cell proliferation, do not correlate with outcome in patients with primary spinal ependymomas; however, spinal ependymomas in children may have better prognosis compared to their adult counterparts. This may be related to the short prodrome in children. Distant metastasis and local recurrence have poor prognosis.

Conclusions

GTR or NTR are sufficient to treat spinal ependymomas in children without needing adjuvant radio- or chemotherapy. Close patient follow-up, neurological assessment, and yearly craniospinal MRI are necessary to prevent serious complications in children. Timely treatment for local recurrence or distant metastasis may increase patient quality of life.

Conflicts of interest

The authors declare that they have no conflicts of interest.

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