Eur Spine J (2017) 26:1917–1928
DOI 10.1007/s00586-016-4939-9

ORIGINAL ARTICLE

Adult-onset intradural spinal teratoma: report of 18 consecutive cases and outcomes in a single center

Wei Wan[1] · Cheng Yang[1] · Wangjun Yan[1] · Tielong Liu[1] · Xinghai Yang[1] · Dianwen Song[2] · Jianru Xiao[1]

Received: 6 November 2015 / Revised: 6 December 2016 / Accepted: 25 December 2016 / Published online: 21 January 2017

Abstract
Study design Eighteen consecutive patients with adult-onset intradural spinal teratoma underwent surgical treatment in our center from 1998 to 2013.
Background and purpose Teratoma is defined as a neoplasm composed of elements derived from three germ cell layers (ectoderm, endoderm and mesoderm). Intraspinal teratoma is extremely rare and accounts for 0.2–0.5% of all spinal cord tumors. Moreover, teratoma occurs primarily in neonates and young children. Adult-onset intradural spinal teratoma is even rare. The aim of this study was to discuss the clinical characteristics, diagnosis and therapeutic strategies of adult-onset intradural spinal teratoma.
Methods This retrospective study included 18 consecutive adult patients with intradural teratoma who were surgically treated in our center between 1998 and 2013. The clinical features, pathogenesis, diagnostic strategies and surgical outcomes were discussed. Neurological function outcomes were evaluated by the JOA scoring system.
Results Of the 18 included patients, 4 patients received subtotal resection and the other 14 patients received total resection. All the 18 cases were diagnosed with mature teratoma. The mean follow-up period was 79.7 (median 60.5; range 27–208) months. Local recurrence occurred in two of the four patients who underwent subtotal resection and in no patient who underwent total resection. The neurologic status improved in 16 cases and remained unchanged in the other two patients.
Conclusions Adult-onset intradural spinal teratoma is extremely rare. To the best of our knowledge, this is the largest series of patients with this disease. Despite the slow-growth and indolent nature, radical resection remains the recommended treatment to reduce tumor recurrence.

Keywords Adult-onset · Intradural spinal teratoma · Subtotal resection · Total resection · Local recurrence

Introduction

Teratoma is defined as a neoplasm composed of a variety of parenchymal cell types derived from three germinal layers (ectoderm, endoderm, and mesoderm) [1], and only 2% occur in the central nervous system [2]. Intraspinal teratoma was first described by Virchow in 1863 [3], and the first confirmed case of intradural spinal teratoma was reported by Gowers et al. in 1888 [4]. It is extremely rare and accounts for 0.2–0.5% of all spinal cord tumors [2, 5]. Furthermore, teratoma predominantly occurs in infants and young children [1, 2, 6, 7]. Surgery is the foundational treatment strategy for intradural spinal teratoma. Although adult-onset intradural spinal teratoma is usually mature and benign, and the recurrence rate was extremely similar between total and subtotal resection (9 vs. 11%) [8]. To avoid the immature tissue or residual malignancy, complete resection is strongly recommended [9].

W. Wan and C. Yang contributed equally to this work and should be considered as first authors.

✉ Dianwen Song
songdw@sh163.net

✉ Jianru Xiao
1210012900@qq.com

[1] Department of Bone Tumor Surgery, Changzheng Hospital, Second Military Medical University, 415 Fengyang Road, Shanghai 200003, China

[2] Department of Orthopedics, Shanghai Jiao Tong University, Shanghai First People's Hospital, No.100 Haining Road, Shanghai 200080, China

There is little published information about the diagnosis and prognosis of adult-onset intradural spinal teratoma due to rarity. Most of these reports described isolated cases or fewer than three cases in a series (Table 2), and the only two series which included seven and eight adult patients older than 18 years emphasized the clinico-pathological features of the disease without discussing the diagnosis and prognosis [1, 30]. In this study, we retrospectively reviewed 18 patients with intradural teratomas who were surgically treated in our spine tumor surgery center, and compared them with the 84 reported cases in the literature since 1888 [1–63]. To the best of our knowledge, this is the first largest series of adult-onset intradural spinal teratoma.

Materials and methods

Patients

From March 1998 to April 2013, 18 adult patients with intradural spinal teratomas were surgically treated in our center. They included eight males and ten females ranging from 18 to 67 years with a mean of 37.6 years. Of them, two (case #3 and #18) patients had a history of surgical treatment in other local hospitals 25 and 15 years ago, respectively, and were re-admitted in our center for local recurrence. The remaining 16 patients were considered as "intact" cases (Table 1). This study was approved by the Ethic Committee of Changzheng Hospital affiliated to the Second Military Medical University (Shanghai, China),and informed consent was obtained individually from the patients or their family members.

Radiologic studies and surgical strategies

Plain X-ray radiography, CT and MRI were performed in all patients before surgery. Radiological examinations were also required to estimate whether there was local recurrence and to observe the stability of the spine during the follow-up periods (Fig. 1). A posterior approach was performed for all patients. Finally, stability reconstruction was accomplished with posterior fixation in 13 patients of the 18 patients. The example images of reconstructed cases are presented in Fig. 1.

Pathologic studies

The finally pathologic diagnosis was confirmed postoperatively by two experienced pathologists independently. To avoid misdiagnosis of "teratoid cyst", "epidermoid cyst" or "dermoid cyst", the sections for pathologic evaluation included the entire extent of the specimen to disclose the presence of the elements from different germinal layers (Fig. 2).

Neurologic evaluations

The pre- and postoperative neurologic status was evaluated by the Japanese Orthopaedic Association Scoring System (JOA score) [64]. The postoperative recovery rate was calculated by Hirabayashi's method as follows: recovery rate = (postoperative JOA score − preoperative JOA score)/(11 − preoperative JOA score) × 100%, and classified as excellent (100–75%), good (74–50%), fair (49–25%), unchanged (24–0%), and deteriorated (decrease in score ≤0%) [64].

Follow-up evaluations

The follow-up evaluation was taken regularly through clinical visits and telephone interviews in the 3rd, 6th and 12th months after surgery in the first year and every 6 months for the next 2 years, and then annually. The recurrence status was confirmed on the basis of the clinical manifestations and imaging findings.

Literature review

The literature review was made on PubMed to sum up the clinical features of adult-onset intradural spinal teratomas from 1888 to the present. Due to the various terms assigned to the same lesion, the search was performed with the key words including "spinal'', "intraspinal", "intradural", "teratoma", "teratoid", "teratoid cyst", "cystic teratoma", "teratomatous cyst", and "cystic teratoid tumor". "Adult" was defined as patients older than 18 years. Altogether 84 cases were filtrated and the clinical data were summarized in Table 2.

Results

A total of 18 patients with adult-onset intradural spinal teratoma were included in our series, who ranged in age from 18 to 67 years with a mean of 37.6 years. The ratio of male to female was 0.8:1 vs. 1.47:1 in the literature (50 male and 34 female; age range 18–85 years, mean 40.02 years) [1–63]. The long diameter of tumor ranged from 1.5 to 12 cm with a mean of 5.1 cm. In our series, tethered cord syndrome (TCS) caused by intradural teratoma occurred in two patients (case #7 and #14), and spinal bifida was observed in one patient (case #11). No other congenital anomalies such as anomalous vertebra, diastematomyelia, myelomeningocele, hemivertebrae and butterfly vertebrae, and hypoplasia of the spinous process were detected.

Intraoperative blood loss ranged from 200 to 1200 mL (mean 422.2 mL). Neither surgical complication occurred

Table 1 Summarized data for all the cases in our study

Case No.	Age (y)/Sex	Symptoms/DT (mo)	Location	AA	JOAS Pre	Excision model	Instrumentation	IBS (ml)	TS (Long Diameter)	Pathologic	AT	JOAS Post	JOA RR	F-U (mo)	LR (mo)	LS
1	35/M	N, W, D, Dy/6	T10-12/EM	None	5	Subtotal	PF	500	7 cm	Mature	None	9	66.7%/G	208	Yes (180)	AWD
2	30/M	W/36	L3-4/EM	None	9	Subtotal	TR	300	4 cm	Mature	None	10	50%/G	188	Yes (168)	AWD
3#	60/M	P, N, W, SD, D/12	L1-5/EM	None	2	En bloc total	PF	600	12 cm	Mature	None	6	44.4%/F	117	No	NED
4	27/F	P, N, Dy/108	L2-S1/EM	None	7	Piecemeal total	PF	1200	10 cm	Mature	None	9	50%/G	108	No	NED
5	48/M	W, D/108	L2-3/EM	None	5	En bloc total	PF	400	4 cm	Mature	None	9	66.7%/G	101	No	NED
6	31/F	P, N/0.5	L3/EM	None	9	En bloc total	TR	300	4 cm	Mature	None	11	100%/E	94	No	NED
7	26/M	W, D, SD/3	L1-2/IM	TCS	6	Subtotal	PF	300	4.5 cm	Mature	None	8	40%/F	92	No	AWD
8	18/F	P/2	T12-L3/EM	None	10	Piecemeal total	PF	400	9 cm	Mature	None	11	100%/E	84	No	NED
9	43/F	P, N, W, D, Dy/7	L3-4/EM	None	6	Piecemeal total	TR	400	3 cm	Mature	None	10	80%/E	62	No	NED
10	67/F	P, W, D, Dy/360	L2/EM	None	1	En bloc total	TR	400	3 cm	Mature	None	7	60%/G	59	No	NED
11	24/F	P/8	T12-L2/IM	Spina bifida	9	Piecemeal total	PF	400	6 cm	Mature	None	11	100%/E	55	No	NED
12	26/F	P, N, Dy/3	L1-2/EM	None	7	En bloc total	TR	300	1.5 cm	Mature	None	10	75%/E	50	No	NED
13	19/M	N, W, D/18	L1-2/EM	None	6	Piecemeal total	PF	400	4 cm	Mature	None	10	80%/E	47	No	NED
14	32/F	P, N, W/120	L2-3/IM	TCS	7	Subtotal	PF	300	4 cm	Mature	None	7	Unchanged	40	No	AWD
15	48/F	P, N/84	L2-3/EM	None	7	En bloc total	PF	200	4 cm	Mature	None	10	75%/E	39	No	NED
16	58/M	P/2	L4-S1/EM	None	10	Piecemeal total	PF	300	5 cm	Mature	None	11	100%/E	33	No	NED
17	34/F	N, W, Dy/24	L1-2/IM	None	8	En bloc total	PF	500	2.5 cm	Mature	None	10	66.7%/G	30	No	NED
18#	50/M	P, W, D, Dy/0.5	L1-2/EM	None	3	Piecemeal total	PF	400	4 cm	Mature	None	8	62.5%/G	27	No	NED
$\bar{X}$	37.6				6.5			422.2	5.1 cm			9.3		79.7		

#: With surgical treatment history

M male, *Fe* female, *DT* duration time since the initial symptom occurred, *N* numb, *W* weakness, *D* dysuria, *Dy* dyschezia, *P* pain, *SD* sexual dysfunction, *EM* extramedullary, *IM* intramedullary, *AA* associated anomalies, *TCS* Tethered Cord Syndrome, *JOAS* JOA Scores, *Pre* preoperation, *Post* postoperation, *PF* posterior fixation, *TR* tumor resection, *IBS* intraoperative blood loss, *TS* tumor size, *AT* adjuvant therapy, *RR* recovery rate, *F-U* follow-up, *LR* local recurrence, *LS* last status, *AWD* alive with disease, *NED* no evidence of disease

$\bar{X}$: Mean

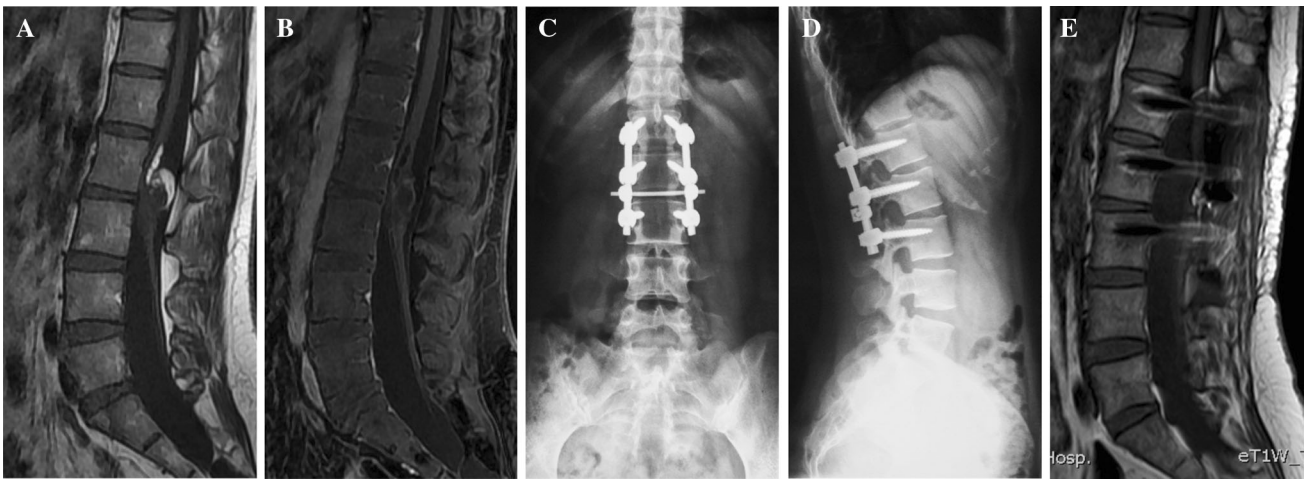

Fig. 1 Images of the intradural spinal teratoma of a 32-year-old woman (Case #14). An ovoid shape, intramedullary space-occupying lesion with heterogeneous signal was presented on T1WI (**a**). The neoplasms extrude the conus medullaris from caudally, and tethered cord syndrome (TCS) was confirmed. Sagittal contrast-enhanced MRI showed just slightly enhancement (**b**). Postoperative radiographs performed 3 years after surgery and manifested that the posterior internal fixation from L1 to L3 was stable (**c**, **d**). In addition,none of local recurrence was detected through MRI (**e**)

in any of our patients, nor was the incidence of spine instability observed during the follow-up periods. Histological diagnosis of mature teratoma was confirmed in all our cases, and pathologic evaluation showed well differentiated elements derived from all three germinal layers (Fig. 2). Of the 84 reported cases, 78 (92.86%) were mature teratoma, 2 cases developed with carcinoid tumor [22, 35], 2 were confirmed as malignant [30, 57], and one was immature [58].

The mean follow-up period was 79.7 months (median 60.5 months; range 27–208 months). Neurologic symptoms were improved to varying degrees postoperatively. The preoperative JOA score was 2–10 (mean 6.5) vs. 6–11 (mean 9.3) postoperation. The neurologic prognosis was excellent in 8 cases, good in 7, fair in 2, and unchanged in the remaining one. Local recurrence was only detected in 2 (11.1%) patients (case #1 and #2), who received subtotal resection and then experienced local recurrence 180 and 168 months after surgery, respectively. But both of them refused reoperation. All the four patients who received subtotal resection have still been symptomless despite the existence of the tumor until now. Meanwhile, none of local recurrence or tumor progression was detected in the rest 14 patients during the follow-up period.

Discussion

Teratoma is classified as mature, immature and malignant based on the degree of differentiation [35, 57]. There are controversies over the pathogenesis of intradural spinal teratoma. Currently, the most widely accepted hypotheses are dysembryogenic theory [31] and misplaced primordial germ cells theory [16, 30]. The former believes that teratoma originates from chaotically differentiated pluripotent cells in primitive streak or caudal cell mass due to dysfunction of several factors involved in gene function and cellular inductive interactions [31]. The latter considers that certain pluripotent primordial germ cells of the neural tube are misplaced into the dorsal midline during their normal migration from the primitive yolk sac to the gonadal ridges [16, 30], resulting in intradural teratoma formation. Associated dysraphic malformations are regarded as the strong evidence to support the dysembryogenic theory [65]. Especially in India and Turkey, the incidence of congenital anomaly or spinal dysraphism is significantly high [65–67]. Whereas, significant dysraphism is absent in greater percentages of adult intradural teratomas. In our study, we only found 2 cases of TCS and 1 case of spinal bifida (16.7%) vs. 35 cases (41.7%) in the literature. Therefore, some researchers insisted that adult intradural spinal teratoma without dysraphism supports the misplaced germ cell theory [1, 30, 39, 56]. However, neither of the theories can accurately elaborate the pathogenic mechanism. In our opinion, the hypothesis proposed by Makary and his colleagues [45] is most acceptable at present. They proposed that there may be a cause-and-effect relationship between spinal cord malformation and the migration of primordial germ cells. Pluripotential cells may get entrapped in an abnormal environment, resulting in teratoma formation due to chaotic differentiation induced by the dysfunction of genetic and cellular inductive interactions during embryogenesis [31].

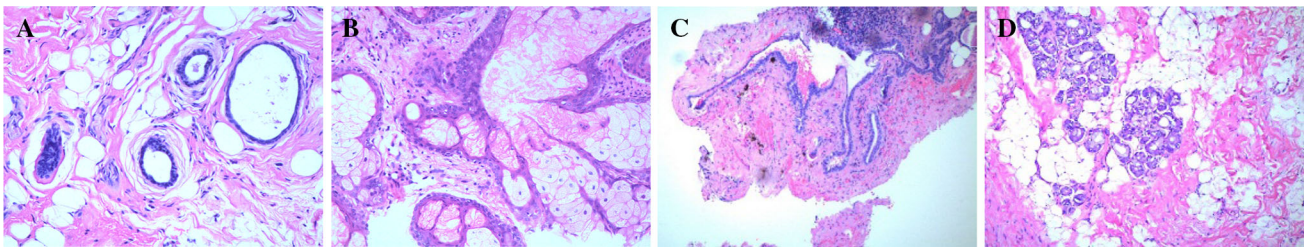

Fig. 2 Histopathology of the specimens. **a** (case #14) showing mature hair follicles, sweat glands, salivary glands, nervous tissues, adipose and connective tissues (Hematoxylin and eosin 200×). **b** (case #17) showing keratinized stratified squamous epithelium cells, hair follicles, and sebaceous glands (200×). **c** (case #15) revealing the cyst wall consist of ciliated columnar epithelium and squamous epithelium cells, and mature glial tissues (100 ×). **d** (case #10) depicting superficial layer of mature ciliated columnar epithelia, nervous, adipose, and connective tissues; adnexal structures, salivary glands, striated muscle are also present (100×)

The tumors in our series were predominantly located near the conus medullary, including 4 intramedullary cases and 14 extramedullary cases. Of the 84 cases reported in the previous literature, 59 (70.2%) cases were in the thoracolumbar junction and lumbar spine, and most of the reported cases were intramedullary ($n = 53$, 63.1%). The reason why the incidence of intramedullary lesions in our series was much lower than the literature was that we had classified some exophytic intramedullary teratomas (case #1, 8, 11, 13) into extramedullary cases. If these patients were categorized into intramedullary cases,the incidence of our study would be 44.4%. We believe these differences are mostly because of the limited sample size of our series or racial differences of Chinese population.

The preoperative definite diagnosis of adult-onset intradural spinal teratoma is usually difficult, because it is quite similar to other space-occupying intradural tumors, especially when the congenital anomaly is not associated. The clinical history of the adult-onset intradural spinal teratoma is usually lengthy and progressive or intermittent [29]. In our series, the duration dates from the initial symptoms ranged from 0.5 to 360 months with a mean of 50.1 months. The main presenting symptoms in our patients were pain, weakness and numbness of the lower extremities, and sphincter dysfunction, depending upon the location of the lesion. Remission of the symptoms during the course of disease may be best explained by varying activity of glandular elements or resorption or leakage of the cystic contents [9, 54]. The leakage of keratin, cholesterol and lipoid may cause recurrent aseptic meningitis [68]. This specific syndrome usually indicates the existence of intradural teratoma. Occasionally, various cutaneous markers such as skin stigmata, hairy patch and dimple, and dermal sinus can be helpful to disclose the presence of teratoma. Thus, a thorough physical examination is necessary.

X-ray radiography often demonstrates erosion of the vertebral bodies, widening of the interpedicular space, and thinned laminae, with or without significant associated vertebral anomalies (Fig. 3), such as spina bifida, vertebral body fusion, incomplete segmentation of vertebraes, diastematomyelia, butterfly vertebrae, etc. It is usually performed as the first diagnostic procedure with limited information provided. CT scanning is available to reveal mixed-density lesion that occupied the intradural space or intralesional calcifications, and assess the spinal bony structure. But it is incapable of visualizing the demarcation between tumor and spinal cord parenchyma. The variation of MRI signal is heterogeneous because of the teratoma's multicomponent nature and dependency on the degree of cystic and solid components [63]. Lipomatous tissue presents hyperintense signal on T1 sequences while hypointense on T2 and fat suppression sequences. Calcification presents hypointense both on T1 and T2. Furthermore, contrast-enhanced MRI usually shows no enhancement. However, it is difficult to distinguish teratoma from the other intradural tumor types by CT and MR images [24, 27], except when the intratumoral calcification and the adipose tissue are present simultaneously (Fig. 4). The final diagnosis must be confirmed via histopathology [52].

Undoubtedly, surgical resection is an essential treatment to relieve the nerve compression in adult-onset intradural spinal teratoma at present [52, 53, 60]. It can prevent neurologic function from deteriorating effectively, and most patients have achieved preferable outcomes after surgery [1–63]. Although, the recurrence rates were extremely similar for total and subtotal resection (9 vs. 11%) in the published literature [8]. In addition, adult-onset intradural spinal teratoma occurs more frequently as intramedullary. Therefore, subtotal resection is usually performed to remove the tumor as much as possible, while the integrity of neural function is preserved. Nevertheless, no patient who underwent total excision experienced recurrence. In contrast, 3 of the patients who received subtotal resection suffered relapse as reported in the literature [8, 22, 58]. In our series, two patients (50%) who received subtotal excision experienced local recurrence. To avoid misdiagnosis and decrease the incidence of relapse,

Table 2 Review of adult intradural teratoma cases reported in the literature, from 1888 to date

References	Number of cases	Sex	Age (year)	Location	AA	Resection model/AT	Pathologic	Outcome
Gowers et al. [4]	1	M	47	T2-3/Intradural	No	Total	Mature	Satisfied
Hosoi [9]	1	M	24	L2-3/IM	Yes	Subtotal	Mature	Satisfied
Sullivan [10]	1	F	32	L1-3/Intradural	No	Total	Mature	Satisfied
Furtado and Marques [11]	1	F	42	T4-T7/EM	No	Subtotal	Mature	Not specified
Dereymaker [9]	2	M:1, F:1	34	M:L1/IM	No	Subtotal	Mature	Poor
			43	F:C5-T2/IM	Yes	Total	Mature	Satisfied
Bakey [12]	1	F	65	L1-2/IM	Yes	Subtotal	Mature	Satisfied
Teng and Gordon [13]	1	F	44	T12-L1/IM	No	Total	Mature	Satisfied
Sloof et al. [5]	2	M:2	20	T11-L1/IM	No	Total	Mature	Satisfied
			67	T11/IM	Not specified	Autopsy	Not specified	Not specified
Rewcastle and Francoeur [14]	1	F	34	T10/IM	No	Subtotal	Mature	Poor
Hansebout and Betrand [15]	1	M	47	L1-L3/IM	Yes	Total	Mature	Satisfied
Enestorm and Von Essen [16]	1	M	36	T11-L1/IM	No	Subtotal	Mature	Satisfied
Rosenbaum et al. [17]	1	M	49	T9/IM	Yes	Total	Mature	Satisfied
Garrison and Kasdon [7]	1	M	23	L2/IM	No	Total	Mature	Satisfied
Padovani et al. [18]	1	F	21	C6-T1/IM	No	Subtotal	Mature	Satisfied
Padovani et al. [19]	1	F	33	L1-2/IM	No	Total	Mature	Satisfied
Ironside et al. [20]	1	F	34	L2-4/Intradural	No	Subtotal	Mature	Poor, Recurred with Carcinoid tumor 8 years later
								Satisfied
Conti [21]	1	F	24	CM/IM	Yes	Subtotal	Mature	Satisfied
Smoker [6]	1	M	26	T1-4/EM	Yes	Subtotal	Mature	Satisfied
Ahmad Monajati et al. [22]	1	M	52	L3-L4/EM	Yes	Total	Mature	Satisfied
Giacomini et al. [23]	1	M	31	T12-L1/IM	Yes	Total	Mature	Satisfied
Pelissou-Guyotat et al. [24]	1	M	33	L4/IM	Yes	Subtotal	Mature	Satisfied
Masanobu Hamabuchi et al. [25]	1	M	54	T12-L1/EM	No	Subtotal	Mature	Satisfied
Nicoletti et al. [26]	1	M	47	T12-L4/IM	Yes	Subtotal	Mature	Satisfied
Caruso et al. [27]	1	M	41	CM/IM	No	Total	Mature	Satisfied
Al-Sarraj et al. [28]	7	M:4, F:3	41	M:CM/EM	No	Not specified	Mature	No recurrence(2 years)
			23	M:CM/EM	Yes	Not specified	Mature	No follow-up
			32	M:CM/EM	No	Not specified	Mature	No recurrence(9 months)
			35	M:CM/IM	No	Not specified	Mature	Recurrence after 10 years
			43	F:CM/EM	No	Not specified	Mature	No recurrence(5 years)
			32	F:C2-7/EM	Yes	Not specified	Mature	Recurrence 6 times in 6 years
			47	F:T10-11/EM	Yes	Not specified	Malignant	No recurrence(5 years)

Table 2 continued

References	Number of cases	Sex	Age (year)	Location	AA	Resection model/AT	Pathologic	Outcome
Koen et al. [29]	1	F	31	L3-4/EM	Yes	Subtotal	Mature	Satisfied
Poeze et al. [9]	1	M	23	T12-L1/IM	No	Subtotal	Mature	Satisfied
Bloch et al. [30]	1	M	56	T12-L1/IM	No	Subtotal	Mature	Poor
Allsopp et al. [8]	2	M:1, F:1	64	M:T11-L1/EM	Yes	Total	Mature	Poor/Recurred 3 years later
			57	F:L1-2/EM	Yes	Total	Mature	Poor
Arai et al. [31]	1	F	54	L2-5/Intradural	Yes	Total	Mature	Satisfied
Elmaci et al. [32]	1	M	42	L4-5/EM	Yes	Not specified	Mature	Poor
Fan et al. [33]	1	F	43	L2/IM	No	Total	Mature with Carcinoid Tumor	Satisfied
Nonomura et al. [2]	2	M:1, F:1	56	M:T12-L2/IM	No	Subtotal	Mature	Poor
			33	F:T12-L1/IM	Yes	Subtotal	Mature	Satisfied
Hejazi and Witzmann [3]	2	M:1, F:1	20	M:L2-4/IM	No	Total	Mature	Satisfied
			45	F:T11-L3/IM	No	Total	Mature	Satisfied
Fernandez-Cornejo et al. [34]	1	M	46	L1-2/IM	No	Total	Mature	Satisfied
Ates et al. [35]	1	M	30	L3-4/EM	No	Total	Mature	Satisfied
Ak et al. [36]	1	F	43	C2-3/IM	Yes	Total	Mature	Satisfied
Paterakis et al. [37]	1	M	63	C2-5/IM	Yes	Subtotal	Mature	Poor
Caruso et al. [38]	2	M:1, F:1	41	M: L1-2/IM	No	Subtotal	Mature	Satisfied
			40	F: L1-2/IM	No	Subtotal	Mature	Poor
Kahilogullari et al. [39]	1	F	42	L1/IM	No	Total	Mature	Satisfied
Stevens et al. [40]	1	M	85	L1-2/EM	No	Subtotal	Mature	Satisfied
Tsitsopoulos et al. [41]	1	F	44	T8-10/IM + EM	Yes	Subtotal	Mature	Satisfied
Raafat Makary et al. [42]	1	F	46	C1-2/IM	Yes	Total	Mature	Satisfied
Mut et al. [43]	1	F	34	L1-2/IM	Yes	Total	Mature	Satisfied
Mohindra [44]	1	M	35	L4/IM + EM	No	Total	Mature	Satisfied
Sung et al. [45]	1	M	38	T11-L1/EM	No	Subtotal	Mature	Satisfied
Han In-Ho et al. [46]	1	F	35	T12-L2/IM	No	Total	Mature	Satisfied
Oh et al. [47]	1	M	44	L3-5/IM	No	Subtotal	Mature	Satisfied
Sharma et al. [1]	8	M:6, F:2	30	M:C2/EM	Yes	Not specified	Mature	Not specified
			28	M:L4-5/EM	Yes	Not specified	Mature	Not specified
			30	M:L1-3/IM	Yes	Not specified	Mature	Not specified
			32	M:T10/IM	Yes	Not specified	Mature	Not specified
			31	M:T11/IM	No	Not specified	Mature	Not specified
			32	M:T11/IM	Yes	Not specified	Mature	Not specified
			45	F:T1-3/EM	Yes	Not specified	Mature	Not specified
			21	F:T10-L2/IM	Yes	Not specified	Mature	Not specified

Table 2 continued

References	Number of cases	Sex	Age (year)	Location	AA	Resection model/AT	Pathologic	Outcome
Arvin et al. [48]	1	M	34	C4-6/IM	Yes	Total	Mature	Satisfied
Samer Ghostine et al. [49]	1	F	65	C1-2/IM	No	Subtotal	Mature	Satisfied
Ijiri-Kosei et al. [50]	1	F	68	L1-2/EM	No	Total	Mature	Satisfied
Benes et al. [51]	1	F	52	L2-5/IM	No	Subtotal	Mature	Satisfied
Gu et al. [52]	1	M	23	L2-3/EM	No	Subtotal	Mature	Satisfied
Jian et al. [53]	2	M:2	18	M:L2-4/IM	No	Total	Mature	Satisfied
			57	M:L1-2/IM	No	Total	Mature	Satisfied
Agrawal et al. [54]	3	M:1, F:2	35	M:L2-3/EM	No	Subtotal	Mature	Lost to follow-up.
			45	F:C4-6/IM + EM	No	Total	Mature	Satisfied
			34	F:C6/EM	No	Total + Radiotherapy	Mature with Malignant change	Satisfied
Moon et al. [55]	1	M	35	C6-T1/IM	No	Subtotal	Immature	Poor, regrowth 4 months later
Yu [56]	1	M	34	L1-2/IM + EM	Yes	Subtotal	Mature	Satisfied
Kalani et al. [57]	1	M	18	T10/EM	No	Subtotal	Mature	Satisfied
Jiang [58]	1	M	56	L4-S1/EM	Yes	Total	Mature	Satisfied
Li et al. [59]	1	F	22	T12-L2/IM + EM	No	Subtotal	Mature	Satisfied
Vanguardia et al. [60]	1	M	41	T12-L1/EM	No	Subtotal	Mature	Satisfied

M male, *Fe* female, *EM* extramedullary, *IM* intramedullary, *AA* associated anomalies, *AT* adjuvant therapy, *Subtotal* subtotal resection, *Total* total spondylectomy

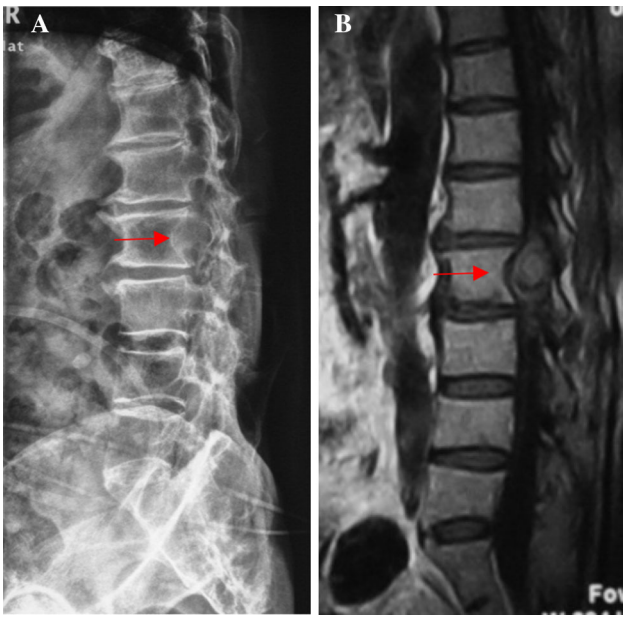

Fig. 3 The X-ray demonstrated the erosion of dorsal vertebral bodies indicating the existence of intraspinal lesion (**a**, case #10). Sagittal T1WI confirmed the ovoid intradural neoplasm located at the level of L2 with heterogeneous signal (**b**, case #10)

total tumor resection and thorough sample examination are strongly recommended, although total resection has higher technical requirements [9].

To reduce the secondary morbidity during surgery, we applied microsurgical techniques, with monitoring somatosensory evoked potentials (SEP) and electromyography (EMG) meticulously [69, 70]. Combination of SEP and EMG can effectively reduce the mechanical injuries of conus medullaris or nerve roots, increase the success rate of tumor resection, estimate the curative effect of surgery and predict post-operative status in real time [69, 70]. Needle aspiration can not only prevent the leakage of cystic fluid into the subdural space but also diminish the tumor's volume and be conducive to remove the neoplasm [59]. In our experience, when TCS is associated, the conus medullaris (CM) should be released sufficiently, including excising the thickening and shortening filum terminals, excising the tight tractive fibrovascular bridles and detaching the cauda equina from the teratoma and surrounding tissues. After removal of the tumor, the surgical field should be washed thoroughly with normal saline to prevent postoperative aseptic meningitis [71]. The dural sac should be closed in a watertight manner to prevent leakage of cerebrospinal fluid [72, 73].

Because of the slow-growth and indolent nature, the recurrence rate of adult-onset intradural spinal teratoma is low, and the prognosis is usually favorable. By far, adjuvant therapy for adult-onset intradural spinal teratomas remains controversial and obscure [8] due to the extremely low incidence and limited experience. Only two reported cases received radiotherapy [22, 57]. As long as malignant or immature elements are found by pathology, radiotherapy should be performed immediately even though the tumor is completely resected [39]. The dose of irradiation is always

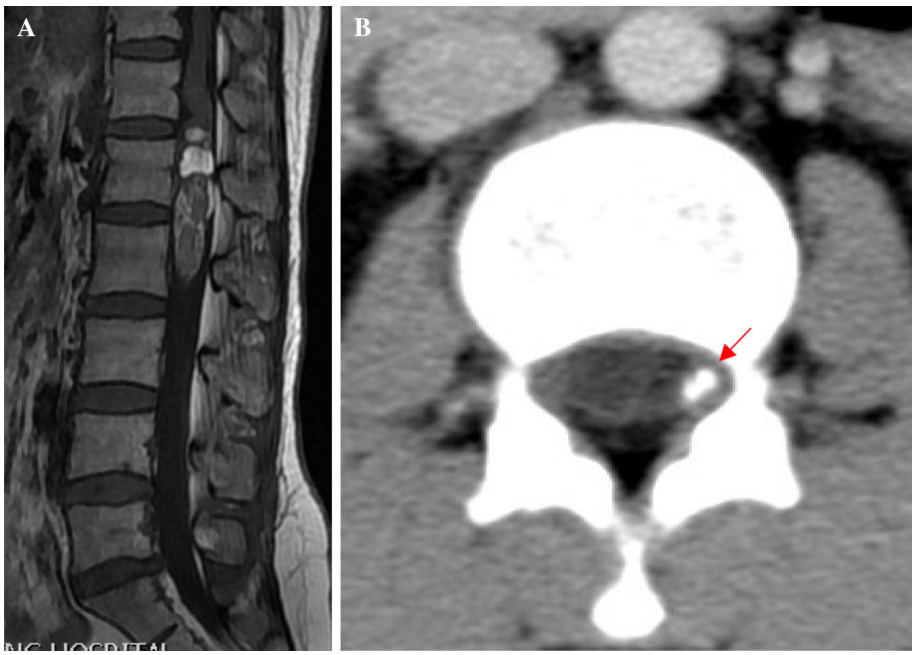

Fig. 4 T1WI showed a spindle shape, intramedullary space occupying lesion with heterogeneous signal (**a**, case #17). Axial CT scanning revealed the intratumoral calcification distinctly (**b**, case #17)

below the widely accepted spinal cord tolerance dose of 45–50 Gy [74]. Potential adverse effects of radiotherapy should also be evaluated [8, 62]. The more recent estimate of 57–61 Gy in conventional fractionation [75] may cause a 5% complication rate in 5 years. To the best of our knowledge, adjuvant chemotherapy has not been applied in adult-onset intradural spinal teratomas to date [8, 54, 62].

Conclusion

Adult-onset intradural spinal teratoma is extremely rare, and only 84 cases have been reported in the previous literature since 1888. To the best of our knowledge, this study including 18 cases is the first largest series so far. Due to the rarity and the lack of definite radiological criteria, preoperative diagnosis is usually difficult, provided intratumoral calcifications and adipose tissues are present simultaneously on CT and MR images. Recurrent aseptic meningitis or associated congenital anomaly is most valuable for diagnosis. Complete total resection is favored whenever possible [26, 35, 36, 38–40], but caution should be exercised when the tissue is intimately involved with the spinal cord parenchyma. The slow-growing nature of the mature forms allows for symptom-free survival even with incomplete resection [6], and malignant change is also extremely rarely seen [35]. Hence, we should not excessively pursue radical total resection, and neurologic function should be protected as much as possible. To avoid misdiagnosis, the whole excised specimen should be thoroughly sampled for histopathologic diagnosis. Once malignant or immature elements are found, radiotherapy should be performed promptly. The dose and protocol should be designed individually to avert radiation myelopathy.

Compliance with ethical standards

Conflict of interest None of the authors has any potential conflict of interest.

References

1. Sharma MC, Jain D, Sarkar C, Suri V, Garg A, Singh M, Mahapatra AK, Sharma BS (2009) Spinal teratomas: a clinicopathological study of 27 patients. Acta Neurochir (Wien) 151:245–252. doi:10.1007/s00701-009-0212-x **(discussion: 252)**
2. Nonomura Y, Miyamoto K, Wada E, Hosoe H, Nishimoto H, Ogura H, Shimizu K (2002) Intramedullary teratoma of the spine: report of two adult cases. Spinal Cord 40:40–43
3. Virchow R (1863) Die Krankhaften Geschwulste, vol 1. Hirschwald, Berlin, p 514
4. Gowers WR, Horsley V (1888) A case of tumour of the spinal cord. Removal; Recovery. Med Chir Trans 71(377–430):11
5. Sloof JL, Kernohan JW, MacCarty CS (1964) Primary intramedullary tumors of the spinal cord and filum terminale. Philadelphia WB Saunders, pp 1–20
6. Smoker WR, Biller J, Moore SA, Beck DW, Hart MN (1986) Intradural spinal teratoma: case report and review of the literature. AJNR Am J Neuroradiol 7:905–910
7. Garrison JE, Kasdon DL (1980) Intramedullary spinal teratoma: case report and review of the literature. Neurosurgery 7:509–512
8. Allsopp G, Sgouros S, Barber P, Walsh AR (2000) Spinal teratoma: is there a place for adjuvant treatment? Two cases and a review of the literature. Br J Neurosurg 14:482–488
9. Poeze M, Herpers MJ, Tjandra B, Freling G, Beuls EA (1999) Intramedullary spinal teratoma presenting with urinary retention: case report and review of the literature. Neurosurgery 45:379–385
10. Hosoi K (1931) Intradural teratoid tumors of the spinal cord: report of a case. Arch Pathol 11:875–883
11. Sullivan BH (1948) Intraspinal teratoma with report of a case. Brooklyn Hosp J 6:142–145
12. Furtado D, Marques V (1951) Spinal teratoma. J Neuropathol Clin Neurol 10:384–393
13. Dereymacker A (1954) Medullar compression by congenital tumors or cysts. Acta Neurol Belg 54:874–889
14. Bakay L (1956) Case records of the Massachusetts General Hospital: case 42502. N Engl J Med 266:1153–1157
15. Teng P, Gordon J (1958) Teratoma of the conus medullaris: report of a case. J Neurosurg 15:569–571
16. Rewcastle NB, Francoeur J (1964) Teratomatous cysts of the spinal canal; with "sex chromatin" studies. Arch Neurol 11:91–99
17. Hansebout RR, Bertrand G (1965) Intraspinal teratoma simulating protruded intervertebral disc. J Neurosurg 22:374–379
18. Eneström S, von Essen C (1977) Spinal teratoma. Acta Neurochir (Wien) 39:121–126
19. Rosenbaum TJ, Soule EH, Onofrio BM (1978) Teratomatous cyst of the spinal canal. Case report. J Neurosurg 49:292–297
20. Padovani R, Tognetti F, Sanpaolo P, Pozzati E, Gaist G, Kuba I (1982) Intramedullary cystic teratoma. Acta Neurochir (Wien) 62:101–108
21. Padovani R, Tognetti F, Laudadio S, Manetto V (1983) Teratoid cyst of the spinal cord. Neurosurgery 13:74–77
22. Ironside JW, Jefferson AA, Royds JA, Taylor CB, Timperley WR (1984) Carcinoid tumour arising in a recurrent intradural spinal teratoma. Neuropathol Appl Neurobiol 10:479–489
23. Conti P, Conti R, De Luca G (1984) Observations on some rare cases of vertebro-medullar malformations associated with tumors. J Neurosurg Sci 28:81–87
24. Monajati A, Spitzer RM, Wiley JL, Heggeness L (1986) MR imaging of a spinal teratoma. J Comput Assist Tomogr 10:307–310
25. Giacomini P, Lunardi P, Liccardo G, Paris L, Sancesario G (1986) Cystic cervico-dorso-lumbar teratoma. Riv Neurol 56:1–8
26. Pelissou-Guyotat I, Sindou M, Pialat J, Goutelle A (1988) Intramedullary mature teratoma associated with an attached cord and an intradural lipoma. Apropos of a surgically treated case. Review of the literature. Neurochirurgie 34:205–209
27. Hamabuchi M, Hasegawa R, Murase T (1989) Teratoma of the spinal cord. A case report with CT scans. J Bone Joint Surg Br 71:390–392
28. Nicoletti GF, Passanisi M, Platania N, Lanzafame S, Albanese V (1994) Intramedullary spinal cystic teratoma of the conus medullaris with caudal exophytic development: case report. Surg Neurol 41:106–111
29. Caruso R, Antonelli M, Cervoni L, Salvati M (1996) Intramedullary teratoma: case report and review of the literature. Tumori 82:616–620
30. al-Sarraj ST, Parmar D, Dean AF, Phookun G, Bridges LR (1998) Clinicopathological study of seven cases of spinal cord teratoma: a possible germ cell origin. Histopathology 32:51–56

31. Koen JL, McLendon RE, George TM (1998) Intradural spinal teratoma: evidence for a dysembryogenic origin. Report of four cases. J Neurosurg 89:844–851
32. Bloch I, Varga Z, Benini A (1999) Teratoma of the conus medullaris. J Clin Neurosci 6:249–252
33. Arai Y, Takahashi M, Takeda K, Shitoto K (2000) Adult-onset intradural spinal teratoma in the lumbar spine: a case report. J Orthop Surg 8:69–74
34. Elmaci I, Dagcinar A, Ozgen S, Ekinci G, Pamir MN (2001) Diastematomyelia and spinal teratoma in an adult. Case report. Neurosurg Focus 10:ecp
35. Fan X, Turner JE, Turner TM, Elrod JP, Clough JA, Howell EI, Johnson MD (2001) Carcinoid tumor development in an intramedullary spinal cord mature teratoma. AJNR Am J Neuroradiol 22:1778–1781
36. Hejazi N, Witzmann A (2003) Spinal intramedullary teratoma with exophytic components: report of two cases and review of the literature. Neurosurg Rev 26:113–116
37. Fernandez-Cornejo VJ, Martinez-Perez M, Polo-Garcia LA, Martinez-Lage JF, Poza M (2004) Cystic mature teratoma of the filum terminale in an adult. Case report and review of the literature. Neurocirugia (Astur) 15:290–293
38. Ates O, Cayli SR, Koçak A, Alkan A, Onal C, Usta U (2005) Mature spinal teratoma associated with thickened filum terminale. Neurol Med Chir (Tokyo) 45:375–378
39. Ak H, Ulu MO, Sar M, Albayram S, Aydin S, Uzan M (2006) Adult intramedullary mature teratoma of the spinal cord: review of the literature illustrated with an unusual example. Acta Neurochir Wien 148:663–669 **(discussion 669)**
40. Paterakis KN, Karantanas AH, Barbanis S, Hadjigeorgiou GM, Karavelis A (2006) Cervical spinal cord intramedullary teratoma. Clin Neurol Neurosurg 108:514–517
41. Caruso R, Colonnese C (2006) Intramedullary teratomas: two case reports and a review of the literature. Zentralbl Neurochir 67:213–218
42. Kahilogullari G, Erdem A, Heper AO, Erden E (2006) Intramedullary mature cystic teratoma of the conus medullaris. A case report. J Neurosurg Sci 50:55–58
43. Stevens QE, Kattner KA, Chen YH, Rahman MA (2006) Intradural extramedullary mature cystic teratoma: not only a childhood disease. J Spinal Disord Tech 19:213–216
44. Tsitsopoulos P, Rizos C, Isaakidis D, Liapi G, Zymaris S (2006) Coexistence of spinal intramedullary teratoma and diastematomyelia in an adult. Spinal Cord 44:632–635
45. Makary R, Wolfson D, Dasilva V, Mohammadi A, Shuja S (2007) Intramedullary mature teratoma of the cervical spinal cord at C1-2 associated with occult spinal dysraphism in an adult. Case report and review of the literature. J Neurosurg Spine 6:579–584
46. Mut M, Shaffrey ME, Bourne TD, Jagannathan J, Shaffrey CI (2007) Unusual presentation of an adult intramedullary spinal teratoma with diplomyelia. Surg Neurol 67:190–194
47. Mohindra S, Chhabra R, Gupta R, Dass Radotra B (2008) Cystic, exophytic teratoma of conus medullaris presenting with chronic renal failure. Surg Neurol 69:81–83 **(discussion 84)**
48. Sung KS, Sung SK, Choi HJ, Song YJ (2008) Spinal intradural extramedullary mature cystic teratoma in an adult. J Korean Neurosurg Soc 44:334–337
49. Han IH, Kuh SU, Chin DK, Kim KS, Jin BH, Cho YE (2008) Surgical treatment of primary spinal tumors in the conus medullaris. J Korean Neurosurg Soc 44:72–77
50. Oh JS, Im SB, Kim BT, Shin WH (2009) Surgical findings of a lumbar mature teratoma accompanying the preoperative intracranial dissemination of Fatty droplets. J Korean Neurosurg Soc 46:409–412
51. Arvin B, Pohl U, David K (2009) Intramedullary cervical teratoma in an adult. Spine J 9:e14–e18
52. Ghostine S, Perry E, Vaynman S, Raghavan R, Tong KA, Samudrala S, Johnson JP, Colohan A (2009) The rare case of an intramedullary cervical spinal cord teratoma in an elderly adult: case report and literature review. Spine (Phila Pa) 34:E973–E978. doi:10.1097/BRS.0b013e3181ba0fe5
53. Ijiri K, Hida K, Yano S, Iwasaki Y (2009) Huge intradural ossification caused by a mature spinal teratoma: case report. Neurosurgery 64:E1200–E1201. doi:10.1227/01.NEU.0000345939.06275.9A **(discussion E1201)**
54. Benes V 3rd, Barsa P, Mikulastik J, Suchomel P (2009) Exophytic intramedullary mature teratoma of the conus medullaris: case report and review of the literature. Cent Eur Neurosurg 70:154–160
55. Gu W, Shang H, Jin X, Xie J, Zhao W (2010) Intradural lumbar mature teratoma with neuronal and glial tissue component in an adult. Case report. Neurol Med Chir (Tokyo) 50:1112–1115
56. Jian W, Ying W, Chao Y (2010) Intramedullary spinal teratoma of the conus medullaris: report of two cases. Acta Neurochir (Wien) 152:553–554
57. Agrawal M, Uppin MS, Patibandla MR, Bhattacharjee S, Panigrahi MK, Saradhi V, Rani JY, Purohit AK, Challa S (2010) Teratomas in central nervous system: a clinico-morphological study with review of literature. Neurol India 58:841–846. doi:10.4103/0028-3886.73740
58. Moon HJ, Shin BK, Kim JH, Kim JH, Kwon TH, Chung HS, Park YK (2010) Adult cervical intramedullary teratoma: first reported immature case. J Neurosurg Spine 13:283–287. doi:10.3171/2010.3.SPINE09461
59. Yu J, Qu LM, Li Y, Huang H (2012) Coexistence of spinal teratoma of the conus medullaris and arteriovenous malformation in an adult: a case report. Turk Neurosurg 22:510–514. doi:10.5137/1019-5149.JTN.3961-10.1
60. Kalani MY, Iyer S, Coons SW, Smith KA (2012) Spinal intradural teratomas: developmental programs gone awry? Neurosurg Focus 33:E1. doi:10.3171/2012.8.FOCUS12230
61. Jiang H, Xiao Z, Zhan X, Chen Q, He M, Lu L (2013) Unusual association of intraspinal extramedullary teratoma with congenital scoliosis in an elderly adult: case report and literature review. Eur Spine J 22(Suppl 3):S306–S310. doi:10.1007/s00586-012-2385-x
62. Li Y, Yang B, Song L, Yan D (2013) Mature teratoma of the spinal cord in adults: an unusual case. Oncol Lett 6:942–946
63. Vanguardia MK, Honeybul S, Robbins P (2014) Subtotal resection of an intradural mature teratoma in an adult presenting with difficulty initiating micturition. Surg Neurol Int 5:23. doi:10.4103/2152-7806.127759
64. Matsuyama Y, Sakai Y, Katayama Y, Imagama S, Ito Z, Wakao N, Yukawa Y, Ito K, Kamiya M, Kanemura T, Sato K, Ishiguro N (2009) Indirect posterior decompression with corrective fusion for ossification of the posterior longitudinal ligament of the thoracic spine: is it possible to predict the surgical results? Eur Spine J 18:943–948. doi:10.1007/s00586-009-0956-2
65. Park SC, Kim KJ, Wang KC, Choe G, Kim HJ (2010) Spinal epidural teratoma: review of spinal teratoma with consideration on the pathogenesis: case report. Neurosurgery 67:E1818–E1825. doi:10.1227/NEU.0b013e3181f846ca
66. Onrat ST, Seyman H, Konuk M (2009) Incidence of neural tube defects in Afyonkarahisar, Western Turkey. Genet Mol Res 8:154–161
67. Devi AR, Rao NA, Bittles AH (1987) Inbreeding and the incidence of childhood genetic disorders in Karnataka, South India. J Med Genet 24:362–365

68. Larbrisseau A, Renevey F, Brochu P, Decarie M, Mathieu JP (1980) Recurrent chemical meningitis due to an intraspinal cystic teratoma: case report. J Neurosurg 52:715–717
69. Devlin VJ, Schwartz DM (2007) Intraoperative neurophysiologic monitoring during spinal surgery. J Am Acad Orthop Surg 15:549–560
70. Kim SM, Kim SH, Seo DW, Lee KW (2013) Intraoperative neurophysiologic monitoring: basic principles and recent update. J Korean Med Sci 28:1261–1269. doi:10.3346/jkms.2013.28.9.1261
71. Mpayo LL, Liu XH, Xu M, Wang K, Wang J, Yang L (2014) Extramedullary spinal teratoma presenting with recurrent aseptic meningitis. Pediatr Neurol 50:655–657. doi:10.1016/j.pediatrneurol.2014.02.005
72. Bokhari I, Rehman L, Farooq G, Qureshi A (2016) Postoperative functional outcome of intradural extramedullary spinal tumors through posterior approach. J Coll Phys Surg Pak 26:117–120. doi:02.2016/JCPSP.117120
73. Mehta AI, Adogwa O, Karikari IO, Thompson P, Verla T, Null UT, Friedman AH, Cheng JS, Bagley CA, Isaacs RE (2013) Anatomical location dictating major surgical complications for intradural extramedullary spinal tumors: a 10-year single-institutional experience. J Neurosurg Spine 19:701–707
74. Emami B, Lyman J, Brown A, Coia L, Goitein M, Munzenrider JE, Shank B, Solin LJ, Wesson M (1991) Tolerance of normal tissue to therapeutic irradiation. Int J Radiat Oncol Biol Phys 21:109–122
75. Schultheiss TE, Kun LE, Ang KK, Stephens LC (1995) Radiation response of the central nervous system. Int J Radiat Oncol Biol Phys 31:1093–1112