Case report

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Clear cell meningioma of the filum terminale in a 44-year-old woman – case report

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Abstract

Clear cell meningioma (CCM) is a rare subtype of meningioma, especially unusual as a neoplasm of the filum terminale. Clear cell meningioma seems to have a more aggressive nature and a higher risk of recurrence than WHO grade I meningiomas.
A 44-year-old woman presented with lower back pain radiating to the left leg and mild weakness in the left leg. Magnetic resonance imaging (MRI) showed a well-demarcated, intradural lesion filling the spinal canal at the L3-S1 levels and compressing the cauda equina. The patient underwent laminectomy from L3 to S1. During the operation, the filum terminale was identified as a structure that was disappearing into the tumor. The filum terminale was cut and the tumor was totally removed in one piece.
Pathological findings were indicative of the diagnosis of clear cell meningioma, CNS WHO G2. Postoperative magnetic resonance imaging at 6 months showed no residual mass. Total surgical excision of the CCM of the spinal cord should be chosen as the optimal treatment. In addition, radiological follow-up is equally important due to the high risk of recurrence. Our case is unusual in that the tumor's location was the filum terminale.

Key words: *meningioma, clear cell meningioma, filum terminale, cauda equina tumor, spinal canal tumor.*

Introduction

Clear cell meningioma (CCM) is a rare subtype of meningioma, especially unusual as a neoplasm of the filum terminale [16,33]. Myxopapillary ependymoma is the commonest tumor at this location [16,31]. Clear cell meningioma is more often found intracranially and in younger patients [30]. The most common clinical symptom of filum terminale tumors is local back pain. In the past, about 66 cases of CCM have been reported in the literature, all of which were treated by surgical resection (Tables I and II) [1,3-9,11-14,17,18,19,21-29,32,34-38]. Here we report a very rare case of CCM of the filum terminale in a patient, which was successfully totally removed.

Presentation

A 44-year-old woman was referred to the Neurosurgical Department of the Institute of Psychiatry and Neurology in Warsaw from the outpatient clinic for surgical resection of a tumor located intradurally extending from vertebrae L3 to S1. The patient had back pain radiating to the left leg. The patient also reported numbness especially of her left foot. A month before admission, the patient began to notice a progressive weakness in the left leg. She denied bladder and anal sphincter dysfunction. Her symptoms had started approximately a year ago. Motor neurological examination at the admission to the neurosurgical department revealed mild left lower extremity weakness, 4/5 points

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Table I. Overview of all reported cases of clear cell meningioma (CCM) tumors located in the spinal canal. The location, recurrences and adjuvant therapy are presented in individual patients

No.	Author, year	Sex	Age	Location	Treatment	Recurrence	Follow-up	Radiotherapy
1	Zorludemir *et al*., 1995 [38]	F	23	L5	GTR	–	36	–
2	Zorludemir *et al*., 1995 [38]	F	36	L2-L5	GTR	–	12	–
3	Zorludemir *et al*., 1995 [38]	F	17	L4-L5	GTR	–	36	–
4	Zorludemir *et al*., 1995 [38]	M	34	L4-S1	GTR	12	156	–
5	Zorludemir *et al*., 1995 [38]	F	9	L3-L5	GTR	6	24	–
6	Zorludemir *et al*., 1995 [38]	M	47	L3-L4	GTR	36	60	+/–
7	Prinz *et al*., 1996 [28]	M	38	Sacrococcygeal	GTR	4	NR	–
8	Holtzman and Jormark, 1996 [12]	M	32	L3-L4	GTR	–	1	–
9	Cancès *et al*., 1998 [5]	F	9	L1-L4	GTR	5	8	+
10	Dubois *et al*., 1998 [9]	F	10	L1-L4	GTR	6	6	+
11	Matsui *et al*., 1998 [22]	F	9	Th12, L2, L5	GTR	–	12	–
12	Maxwell *et al*., 1998 [23]	F	31	L3	GTR	NR	NR	–
13	Maxwell *et al*., 1998 [23]	M	24	L1	GTR	NR	NR	–
14	Alameda *et al*., 1999 [1]		42	L-S	GTR	–	24	–
15	Pimentel *et al*., 1998 [27]	M	55	C	GTR	–	NR	–
16	Pimentel *et al*., 1998 [27]	F	21	L	GTR	–	NR	–
17	Heth *et al*., 2000 [11]	F	7	L4-L5	GTR	–	13	–
18	Park *et al*., 2000 [25]	F	1.2	Th12-L2	GTR	8	8	–
19	Jallo *et al*., 2001 [14]	F	8	L3-L5	GTR	6	11	+
20	Jallo *et al*., 2001 [14]	F	1.8	C3-C5	STR	2.5	38	+/–
21	Yu *et al*., 2002 [34]	F	1.2	Th12-L1	GTR	4	15	–
22	Carra *et al*., 2003 [6]	M	1.8	Th11-L4	GTR	60	60	–
23	Boet *et al*., 2004 [4]	M	34	L4-S3	STR	–	15	+
24	Dhall *et al*., 2005 [8]	F	33	Th12-S1	GTR	–	11	–
25	Liu *et al*., 2005 [19]	M	2.2	Th10-L1	GTR	60	60	–
26	Oviedo *et al*., 2005 [24]	M	7	L2-L3	GTR	–	12	–
27	Park *et al*., 2005 [26]	F	65	Th9-Th10	GTR	–	24	–
28	Jain *et al*., 2006 [13]	F	26	L2-S1	GTR	–	36	–
29	Vural *et al*., 2007 [37]	F	4	C1-C2	GTR	–	6	–
30	Colen *et al*., 2009 [7]	F	13	L4-L5	GTR	–	24	+
31	Tong *et al*., 2010 [32]	F	35	C7	GTR	–	3	–
32	Mallya *et al*., 2012 [21]	F	50	Sacro-coccygeal	NR	NR	NR	–
33	Balogun *et al*., 2013 [3]	M	3	Th12-L5	GTR	9	9	–
34	Schollenberg *et al*., 2013 [29]	F	54	L3-L4	NR	NR	NR	NR
35	Wang *et al*., 2014 [35]	M	79	C1-C2	GTR	NR	NR	–
36	Wang *et al*., 2014 [35]	M	18	C6-Th2	NR	20	60	NR
37	Wang *et al*., 2014 [35]	M	35	L5-S1	GTR	–	7	+
38	Li *et al*., 2016 [17]	M	21	L5	GTR	–	5	–
39	Li *et al*., 2016 [17]	F	43	L3-S3	GTR	–	10	–
40	Li *et al*., 2016 [17]	F	7	Th11-L1	GTR	–	9	–
41	Li *et al*., 2016 [17]	F	7	L2-L4	GTR	–	2	–

No.	Author, year	Sex	Age	Location	Treatment	Recurrence	Follow-up	Radiotherapy
42	Li *et al*., 2016 [17]	M	4	Th11-Th12	GTR	–	2	–
43	Li *et al*., 2016 [17]	F	20	L4/L5	GTR	–	2	–
44	Li *et al*., 2018 [18]	M	50	C3-Th1	GTR	–	38	–
45	Li *et al*., 2018 [18]	M	28	Th10	GTR	24	27	–
46	Li *et al*., 2018 [18]	F	20	L3	GTR	–	57	–
47	Li *et al*., 2018 [18]	F	15	L3	GTR	–	16	–
48	Li *et al*., 2018 [18]	F	34	L4	GTR	–	70	–
49	Li *et al*., 2018 [18]	F	37	L5-S3	GTR	–	10	–
50	Li *et al*., 2018 [18]	M	16	L5-S1	GTR	–	93	–
51	Li *et al*., 2018 [18]	M	53	L4-S1	GTR	–	69	–
52	Li *et al*., 2018 [18]	F	27	C1-C2	STR	120	130	–
53	Li *et al*., 2018 [18]	F	16	Th12-L1	STR	120	192	–
54	Li *et al*., 2018 [18]	F	14	L4	STR	24	56	–
55	Li *et al*., 2018 [18]	M	35	L3-S2	GTR	12	70	–
56	Wu *et al*., 2019 [36]	F	7	Th11-L1	GTR	–	46	–
57	Wu *et al*., 2019 [36]	F	7	L2-L4	GTR	–	36	–
58	Wu *et al*., 2019 [36]	M	4	Th11-Th12	GTR	–	70	–
59	Wu *et al*., 2019 [36]	F	50	Th11	GTR	–	112	–
60	Wu *et al*., 2019 [36]	M	23	L5-S3	GTR	–	100	–
61	Wu *et al*., 2019 [36]	F	52	C5	GTR	–	36	–
62	Wu *et al*., 2019 [36]	F	43	L3-S3	STR	–	56	–
63	Wu *et al*., 2019 [36]	M	21	L5	GTR	–	54	–
64	Wu *et al*., 2019 [36]	F	20	L4-L5	GTR	–	48	–
65	Wu *et al*., 2019 [36]	F	16	Th12-L1	GTR	–	126	–
66	Present study	F	44	Film terminale, L3-S1	GTR	–	6	–

No – number, GTR – gross total removal, NR – not reported, STR – subtotal resection

on the Lovett scale. Sensory neurological examination revealed a decrease in touch sensation in the left lower limb, mainly distal, which was consistent with the presented symptoms. There were no abnormalities in the examination of pain, temperature, position and vibration. The patient had no history of trauma to the lumbosacral region. Medical history was uneventful. On radiological investigation, magnetic resonance imaging (MRI) showed a well-demarcated, intradural lesion filling the spinal canal at the L3-S1 levels compressing the cauda equina. The tumor signal was heterogeneously iso- to hypointense in T2-weighted images and isointense in T1-weighted images (Fig. 1). Based on imaging appearances and clinical examination, surgical resection of the tumor was planned. Differential diagnoses included typical meningioma, neurofibroma, and ependymoma.

The patient underwent surgery in the prone position. Laminectomy from L3 to S1 was performed. After performing laminectomy from L3 to L5 the tense dura mater was ascertained. A dural incision was made in the midline under the operative microscope. The tumor was solid, greyish, well demarcated, and was lying between the spinal nerve roots. At the L3 level, the filum terminale, which was disappearing into the tumor, was identified. The lesion was carefully dissected from the nerve roots and totally removed in one piece. The filum terminale was cut and total tumor resection was achieved (Fig. 2).

Neuropathological examination revealed that the tumor was composed of glycogen-rich cells surrounded by bands of collagen (Fig. 3). Neoplasm cells had round- to oval-shaped central nuclei, inconspicuous nucleoli, abundant, clear, periodic acid-Schiff (PAS)-positive cytoplasm, and lacked significant mitotic activity. Immunohistochemistry showed weak, membranous, positive immunoreactivity for epithelial membrane antigen (EMA), and negative immunoreactivity for S-100, glial

Table II. Clinical characteristics of patients with diagnosis of clear cell carcinoma in the spinal canal. Recurrences occurred in 19 patients (31.1%) at a mean follow-up of 25.5 months (range 1-192 months). The follow-up was missing in 8 patients operated on for clear cell carcinoma in the spinal canal

Clinical characteristic		Number (%)
Age (years)	Mean	25.0 ±18.1
	Range	1.2-79
Sex	F	41 (63.1)
	M	24 (36.9)
Operation	GTR	57 (90.4)
	STR	6 (9.6)
Follow-up (months)	Median	25.5
	Range	1-192
Location	Cervical	7 (10.6)
	Cervico-thoracic	2 (3)
	Thoracic	5 (7.6)
	Thoraco-lumbar	11 (16.7)
	Lumbo-sacral	41 (62.1)
Tumor size	1 level	12 (18.2)
	> 1 level	44 (81.8)

fibrillary acid protein (GFAP), and chromogranin A. Pathological findings were indicative of the diagnosis of clear cell meningioma, CNS WHO G2.

Postoperative course

The postoperative course was uneventful and the neurological deficits completely resolved. The patient was discharged home on the second day after surgery without any neurological deficits. No adjuvant radiotherapy or chemotherapy was recommended by the consultant neurooncologist. The patient was further followed on an out-patient basis. Follow-up MR examinations were scheduled for the patient. An early postoperative follow-up MRI performed six months after tumor resection showed no residual mass (Fig. 4).

Discussion

Clear cell meningioma is a rare histopathological subtype of meningioma, classified by the World Health Organization classification as a grade 2 tumor [20]. Li *et al*. reported the largest series to date, with 55 spinal CCM. The most common location of spinal CCM is the spinal lumbar region [18]. To the best of our knowledge, this is the first case report of CCM of the filum terminale. In this location, the most common tumor found is myxopapillary ependymoma [31].

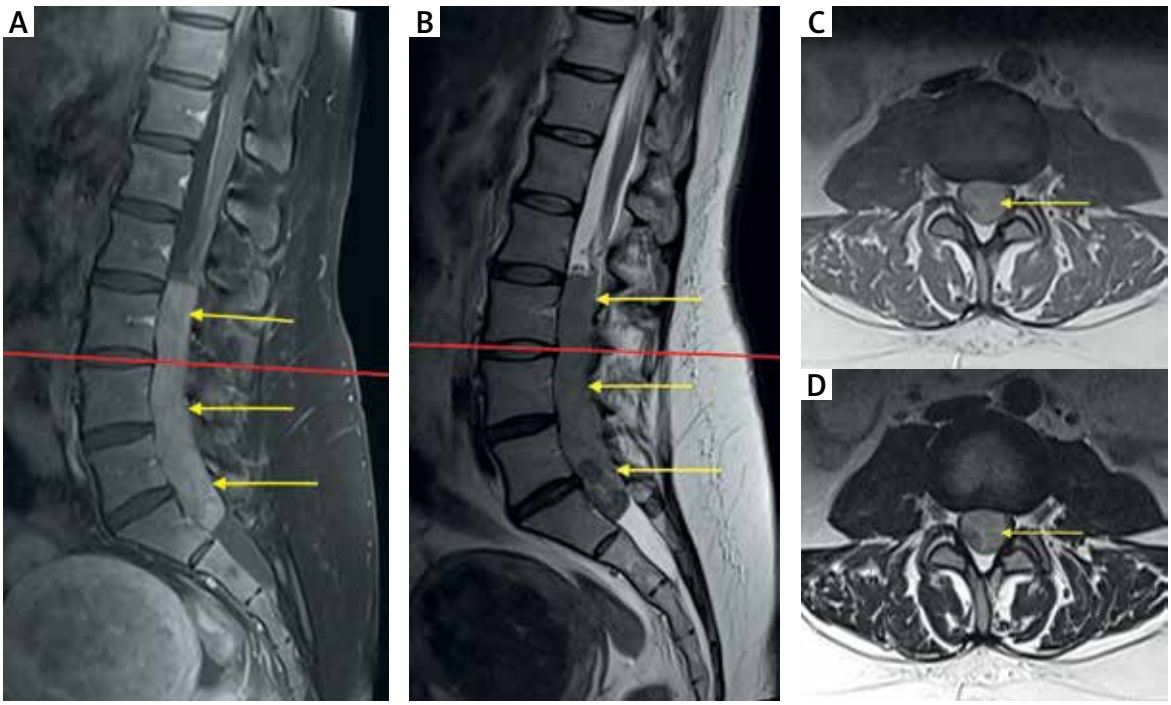

Fig. 1. Magnetic resonance imaging (MRI) of a mildly heterogeneously intensity clear cell meningioma (CCM). Preoperative MRI showed a lesion (yellow arrow) located at L3-S1 with heterogeneous intensity on (**A**) T1-weighted gadolinium-enhanced and (**B**) T2-weighted sagittal images. Transaxial (**C**) T1-weighted gadolinium-enhanced and (**D**) T2-weighted MR image at the L3-L4 level (red line).

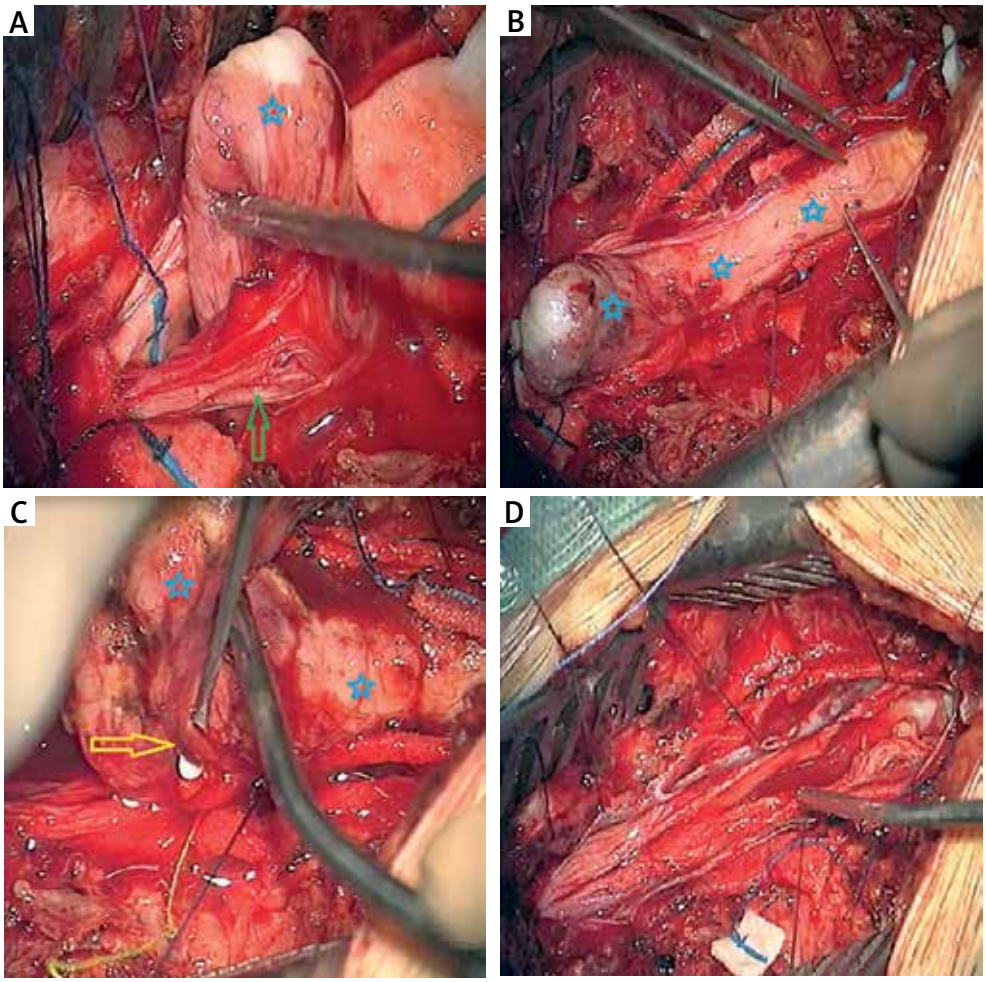

Fig. 2. Intraoperative images after L3-S1 laminectomy. **A**, **B**) The intradural tumor (blue star) covering and compressing the nerve roots (green arrow). **C**) The filum terminale (yellow arrow) which was disappearing into the tumor. **D**) Intradural space after tumor gross total resection.

Clear cell meningioma has no specific radiological features to distinguish it from other meningiomas, which makes histopathological examination the basis for diagnosis [2,34]. Typically, the tumor is composed of cells with clear, glycogen-rich cytoplasm and prominent perivascular and interstitial collagen deposits.

A review of previous reports showed that gross total resection is the primary goal of surgical treatment [10,15,18]. One of the main features that distinguishes CCM from WHO grade 1 meningiomas seems to be a more aggressive nature and a higher risk of recurrence. According to Liu *et al*. the local recurrence rate after gross tumor resection is 63% and the mean time to recurrence is 18.3 months [19]. In our case, MRI showed no signs of recurrence at the postoperative follow-up, but further follow-up is required with scheduled MRI examinations.

The role of adjuvant radiotherapy in spinal CCM is controversial [7]. Most authors after gross tumor resection do not use adjuvant radiotherapy [10]. However, radiotherapy could be considered for patients who have incomplete resection or local recurrence [7,15,33]. On the other hand, Tao *et al*. used radiotherapy as an adjuvant treatment after surgery in all 9 patients and no recurrence was observed [30]. There is general agreement that patients diagnosed with CCM require long-term follow-up with MRI to pick up clinically silent recurrences [15,19]. These spinal CCM recurrences are usually treated with adjuvant radiotherapy [10].

Conclusions

Clear cell meningioma is a rare histological subtype of meningioma. Total surgical excision of the CCM of the spinal cord should be chosen as the primary and

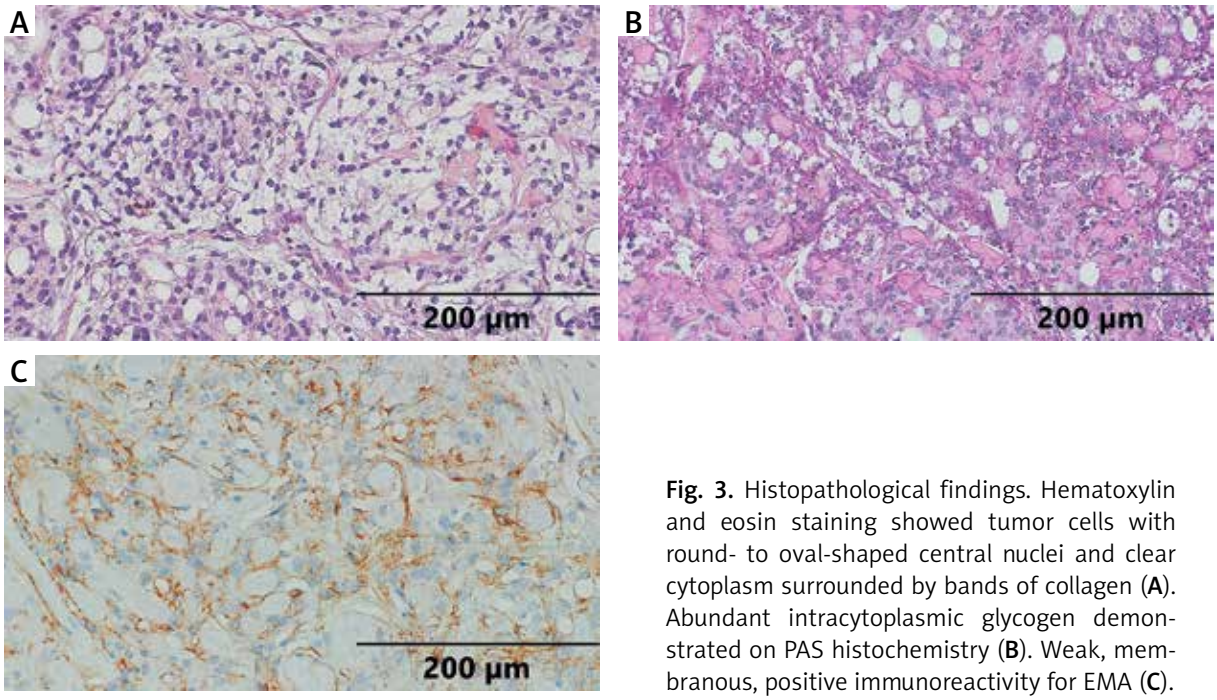

Fig. 3. Histopathological findings. Hematoxylin and eosin staining showed tumor cells with round- to oval-shaped central nuclei and clear cytoplasm surrounded by bands of collagen (**A**). Abundant intracytoplasmic glycogen demonstrated on PAS histochemistry (**B**). Weak, membranous, positive immunoreactivity for EMA (**C**).

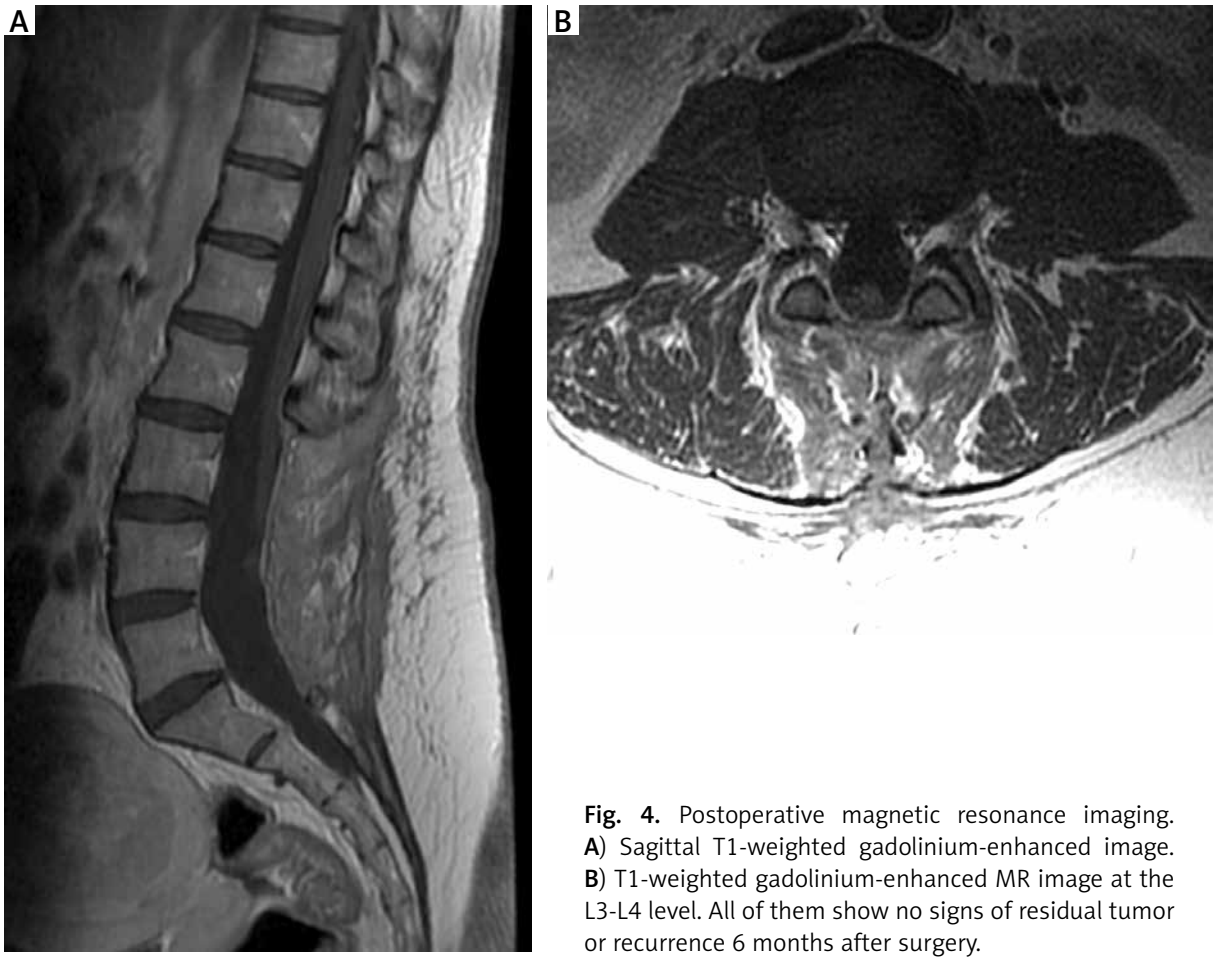

Fig. 4. Postoperative magnetic resonance imaging. **A**) Sagittal T1-weighted gadolinium-enhanced image. **B**) T1-weighted gadolinium-enhanced MR image at the L3-L4 level. All of them show no signs of residual tumor or recurrence 6 months after surgery.

optimal treatment. Adjuvant radiotherapy should be considered only in selected cases of recurrence. Because of the high recurrence rate, close radiological and clinical follow-up is strongly recommended.

Disclosure

The authors report no conflict of interest.

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