EN BLOCK RESECTION OF THE GIANT INVASIVE SCHWANNOMA IN THE THORACOLUMBAR SPINE

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The paper presents a rare clinical case of surgical treatment of a patient with a giant invasive schwannoma of the thoracolumbar spine. A single-stage en block resection of the tumor through a combined posteroanterior approach was performed followed by replacement of post-resection interbody diastasis with a carbon implant and by posterior instrumental fixation of the spine. The pain syndrome regressed from VAS scores 7 and 8 (back, lower limbs) to scores 4 and 1, respectively. The follow-up examination was conducted at 6 and 12 months after surgery: there were no signs of relapse. Publications on giant invasive spinal schwannomas were analyzed.

Key Words: tumor, giant invasive spinal schwannoma, en block resection.

Please cite this paper as: *Kuklin DV, Naumov DG, Belyakov MV, Sovpenchuk IA, Serdobintsev MS. En block resection of the giant invasive schwannoma in the thoracolumbar spine. Hir. Pozvonoc. 2019;16(1):70–76. In Russian.*

DOI: http://dx.doi.org/10.14531/ss2019.1.70-76.

Schwannoma is a slow-growing benign tumor that is morphologically represented by Schwann cells associated with axons of the peripheral nerve fibers [1]. Schwannoma accounts for 25 % of primary intradural tumors and is most often located in the lumbosacral spine [2]. According to a 10-year population-based study [3], 3 % of patients with neurofibromatosis type 2 (NF2) develop schwannoma due to abnormal synthesis of a tumor growth suppressor, the merlin protein (schwannomin). The rate of schwannoma malignant transformation into neurofibrosarcoma is not more than 1 % [4].

Giant invasive spinal schwannoma is an extremely rare benign neoplasm that affects two or more spinal motion segments with destruction of the adjacent vertebrae and extends into paraspinal tissues [5]. Giant invasive schwannomas account for no more than 2.5 % of primary intradural tumors, with the lumbar spine being most often affected [6]. An asymptomatic course and a long-term absence of neurological deficit underlie a prolonged diagnostic pause resulting in late diagnosis and significant tumor growth [7]. En bloc resection of the pre-, para- and intravertebral components using a combined (anterior and posterior) approach is the gold standard for surgical treatment of schwannomas, which not only enables spinal canal decompression but also ensures a recurrence-free postoperative period [8]. Technical difficulties (intimate anatomical relationships of the vascular structures, spinal cord, and its roots; high risk of intraoperative bleeding and liquorrhea; large tumor size) cause high interest in the features of surgery for giant invasive schwannomas.

The purpose of this study is to describe and analyze surgical treatment of a rare giant invasive schwannoma of the thoracolumbar junction.

A 60-year-old male patient P. underwent chest radiography due to acute bronchitis in 2011, which revealed a lesion at the T12 level. According to CT and MRI findings, the lesion of 8.0 × 10.1 × 6.0 cm in size was located extraperitoneally and invaded the T12 vertebral body and related nerve root on the right (Fig. 1).

For morphological verification, a puncture biopsy of the lesion was performed through the longissimus dorsi muscles on the right. An immunohistochemical study revealed that this lesion was schwannoma (Fig. 2). Given the asymptomatic course and accidental detection of the tumor, oncologist follow-up at the place of residence was recommended to the patient. The patient had no complaints for 6 years.

In 2017, he noted pronounced pain in the back and lower extremities, scored 7 and 8 (VAS), respectively; neurological status: Frankel grade E. Contrast-enhanced CT and MRI (Fig. 3) revealed a space-occupying lesion at the T10–L3 level, which extended paravertebrally with T12 body destruction (sectors 8–12 according to WBB [9]) and compressed the lateral wall of the dural sac. An intraspinal component of the tumor was 6.2 × 1.5 cm, and that located in the longissimus dorsi muscles (within *m. erector spinae dex.*) – 11.3 × 12.4 × 5.5 cm. A tumor component located paravertebrally on the right was 8 × 8.4 × 3.0 cm in size, with the segmental vessels passing through the component's nodes.

Given the tumor spreading, pronounced pain, and pattern of vertebral destruction, single-stage multistep surgery with participation of spinal surgeons and a vascular surgeon was decided.

The first step involved monolateral posterior transpedicular fixation of T10–L2 on the left, followed by identification of a tumor node in the *m. erector spinae* and T11–L1 laminotomy with exposure

of the intraspinal and foraminal tumor components.

At the second step, the paravertebral tumor component was identified via the thoraco-diaphragmatic approach, which enabled en bloc removal of the tumor with resection of the T12 vertebral body, followed by anterior fusion with a carbon mesh cage.

The third step included posterior transpedicular instrumented fixation of T10–L2 (Fig. 4).

The duration of surgery was 4 h 20 min; blood loss amounted to 1,200 mL.

In the postoperative period, the patient had a pronounced improvement with regression of pain on the 4th postoperative day in the back and lower extremities to a score of 4 and 1 (VAS), respectively. The patient remained neurologically intact (Frankel grade E). He was verticalized in an orthosis on the 4th postoperative day. Morphological findings: malignant transformation of schwannoma (Fig. 5).

Given a resistance of the tumor to chemotherapy, en bloc tumor resection, and oncologists' advice, adjuvant therapy was refused. The long-term outcomes were assessed at 6 and 12 months after surgery: complete pain relief; no signs of continued tumor growth (Fig. 6). The neurological status remained at the preoperative level.

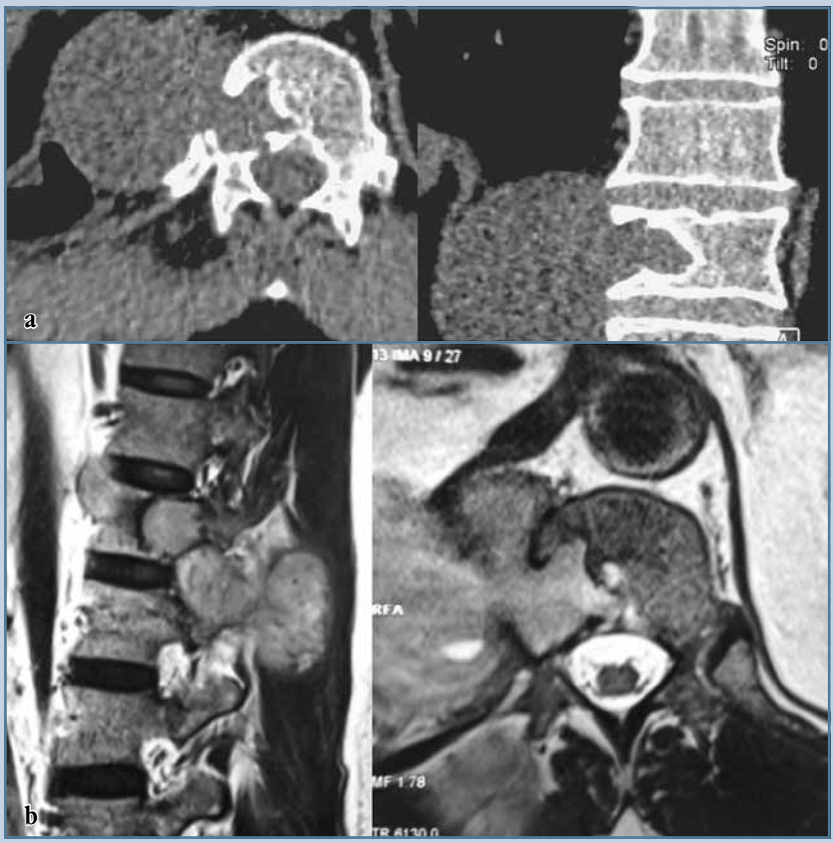

Fig. 1
CT **(a)** and MRI **(b)** scans of the thoracolumbar spine of a 60-year-old male patient P. at primary detection of a tumor (2011)

Discussion

The capabilities of schwannoma surgery are described in the literature in sufficient detail. Some papers focus on the features of this tumor.

Fehlings et al. [10] analyzed surgical outcomes in 169 patients with schwannomas, among whom there were only 9 cases of postoperative recurrence, and identified potential predictors for recurrence risk: age (it was slightly higher in the recurrence group than in the recurrence-free group: 47.01 ± 15.19 years vs. 39.33 ± 14.58 years, respectively), lumbar localization, mean tumor size of more than 6.97 cm, polysegmental bone destruction, and intralesional resection.

The term "giant invasive spinal schwannoma" was introduced into clinical practice by Sridhar et al. [5] who improved the X ray-based classification of spinal schwannomas and presented their description (Fig. 7).

A distinctive feature of type V schwannomas is extradural extension, destruction of bone structures, and mandatory involvement of adjacent myofascial fibers. These clinical and morphological features dictate, on the one hand, the need for en bloc resection of the tumor and, on the other, make this task technically difficult.

To systematize cases of surgical treatment of giant invasive spinal schwannoma reported in the literature, we searched the PubMed, ClinicalKey, and eLibrary databases for the period between 2000 and 2018 using the following keywords: "giant invasive spinal schwannoma". We selected 13 publica-

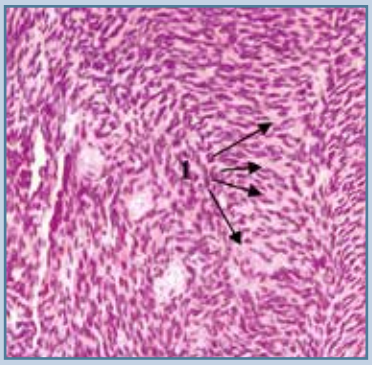

Fig. 2
Histologic structure of the tumor in the 60-year-old male patient P. (2011): *1* – multiple Verocay bodies forming the Antoni A pattern

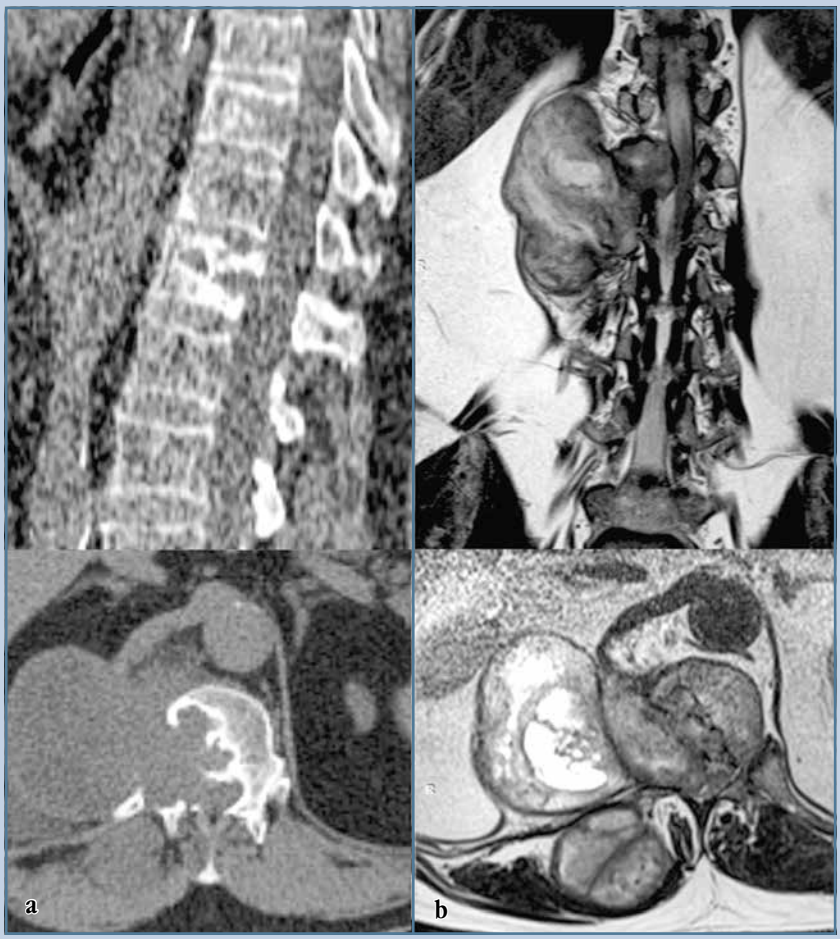

Fig. 3
Sagittal and axial CT scans **(a)** and coronary and axial MRI scans **(b)** of the thoracolumbar spine of the 66-year-old male patient P. at repeated admission (2017)

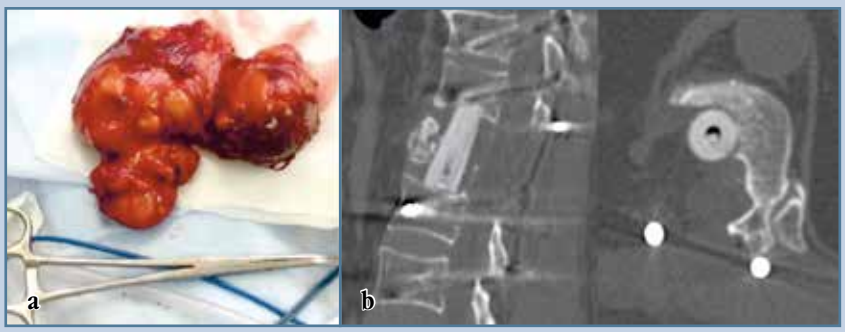

Fig. 4
Gross specimen **(a)** of a 15.0 × 12.0 × 15.0 cm en bloc resected tumor with an intact capsule; sagittal and axial CT scans of the spine **(b)** 7 days after surgery (360° reconstruction: anterior spinal fusion of T11–L1, posterior instrumentation (T10–L2)

tions; the information on them is summarized in the Table.

The largest clinical series [8] included data on 14 patients who underwent three types of surgery: en bloc resection (n = 11), partial resection without removal of the foraminal component (n = 2), and partial resection of the extravertebral component (n = 1). The most frequent complication that significantly complicated surgery was intraoperative bleeding from the vertebral destruction zone (n = 9) or tumor tissue (n = 3). In five cases, removal of the intradural component required opening of the dural sac, which was complicated by liquorrhea in the postoperative period in one patient. The best outcomes (improvement of the neurological status and no recurrence) were achieved in the en block resection group. For postoperative follow-up, the authors recommend annual MRI and analysis of the Ki-67 antigen expression level. The overwhelming majority of publications (9 of 13) were limited to the description of individual cases with the best postoperative outcome in the case of total en bloc resection of the tumor.

Conclusion

This clinical case is of interest for the following reasons:

1) the domestic literature lacks similar publications;

2) the described technical features of surgery (single-stage, combined approach, involvement of a multidisciplinary team of surgeons) enabled en bloc resection, ensuring regression of pain and an 1-year recurrence-free period;

3) according to the literature, malignant transformation of the tumor, which was revealed by morphological analysts of surgical material, does not exceed 1 %. However, tumor growth and transition from an asymptomatic to symptomatic course are not considered in the literature as clear signs of malignant transformation.

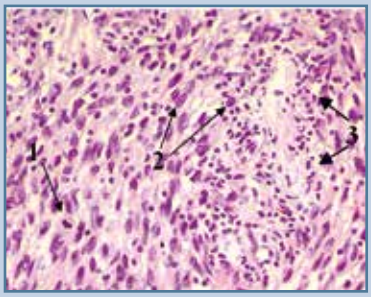

Fig. 5
Histologic structure of the malignant tumor of the 66-year-old male patient P. (2017): *1* – mitotic figure; *2* – closely arranged ovoid cells; *3* – areas of perineural differentiation

The authors are grateful to Prof. A.Yu. Mushkin for assistance in manuscript preparation and to V.V. Shlomin for participation in surgery.
The study was conducted without financial support.
The authors declare no conflict of interest.

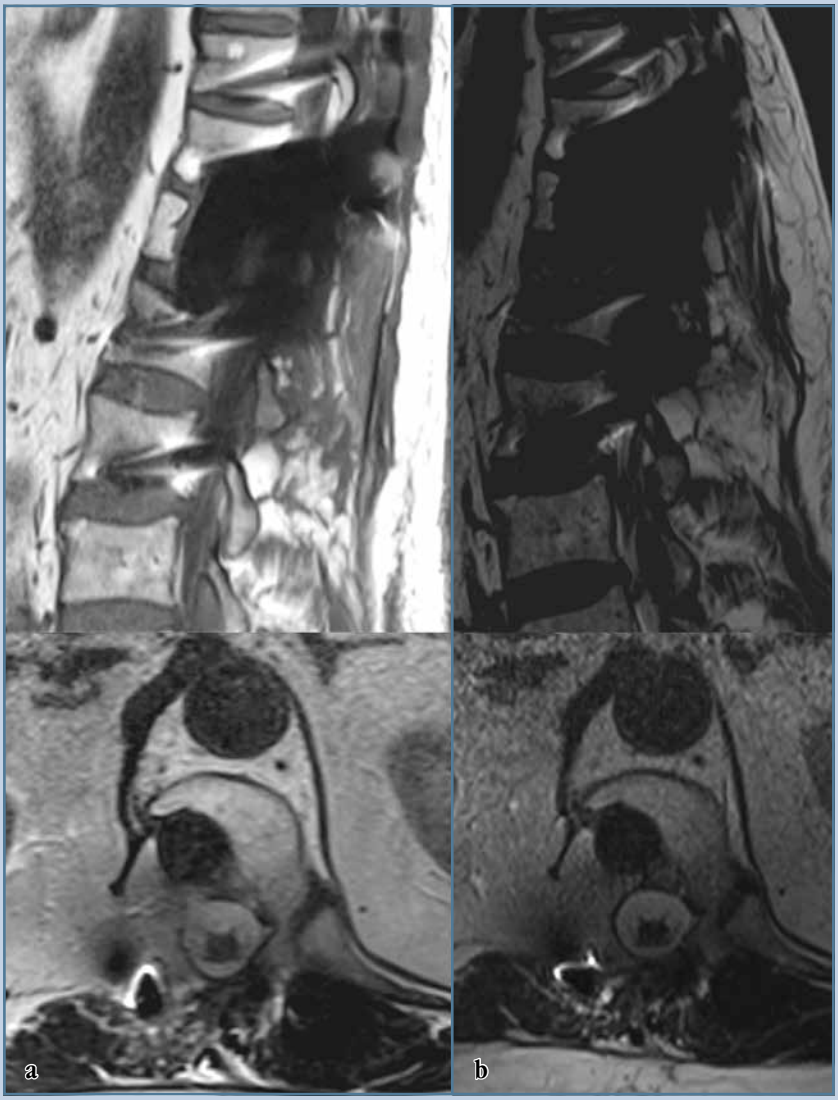

Fig. 6
Sagittal and axial MRI scans of the spine of the 66-year-old male patient P. at 6 **(a)** and 12 **(b)** months after surgery: there are no signs of continued tumor growth

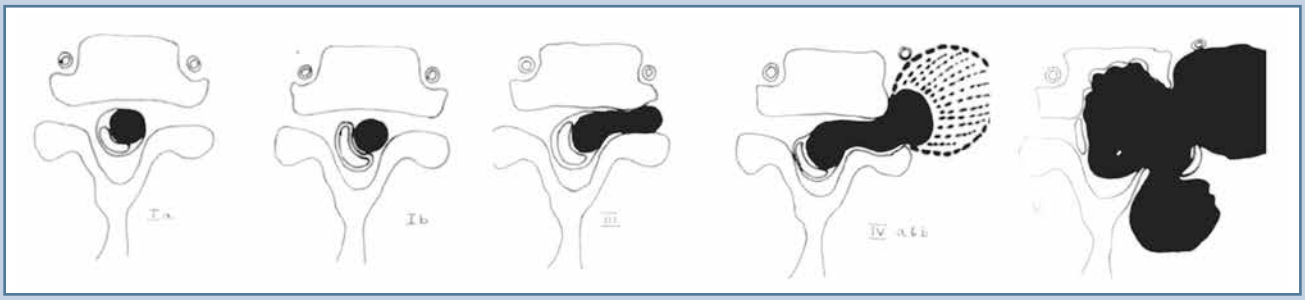

Fig. 7
Classification of spinal schwannomas according to Sridhar et al. [5]: type Ia – intradural spread over less than two spinal motion segments; type IB – intraspinal extradural spread over less than two spinal motion segments; type II – intraspinal spread over more than two spinal motion segments; type III – intraspinal extension into the nerve root foramen; type IV – extraspinal spread of the tumor with the paravertebral component of less than 2.5 cm (IVa) and more than 2.5 cm (IVb); type V – extraspinal spread over two or more spinal motion segments with destruction of spinal bone structures and involvement of the anterior, lateral, and posterior myofascial structures – giant invasive spinal schwannoma

Table
Literature data on surgical treatment of giant invasive spinal schwannomas

Authors	Patients, n	Localization of vertebral destruction (level : quantity)	Long-term outcome
Sridhar et al. [5]	10	C/T/L/LS : 1/2/5/2	Liquorrhea (1), recurrence (1), 3 year post-op
Bunc et al. [11]	1	TL	Recurrence (1), 12 year post-op
Chiang et al. [7]	1	L	Recovery, 1 year post-op
Ozdemir et al. [12]	6	C/T/L/S : 3/1/1/1	Recurrence (1), 2 year post-op, death (1)
Pongsthorn et al. [13]	6	S	Recurrence (1), 7 year post-op
Alfieri et al. [14]	1	S	Recovery, 10 year post-op
Yu et al. [8]	14	C/CT/T/TL/LS/S : 1/1/1/2/4/5	Recurrence (1), 1 year post-op
Kataria et al. [15]	1	LS	No data
Iizuka et al. [16]	1	L	Recovery, 2 year post-op
Valle-Giler et al. [17]	1	T	Recovery, 1 year post-op
Togral et al. [18]	1	LS	Recovery, 1 year post-op
Tonomura et al. [6]	1	L	Recovery, 8 year post-op
Khan et al. [19]	1	S	No data

References

1. Pathology and Genetics of Tumors of Soft Tissue and Bone, ed. by Fletcher CD, Unni KK, Mertens F. Lyon: IARC Press, 2002:331.
2. **Kim NR, Suh YL, Shin HJ.** Thoracic pediatric intramedullary schwannoma: report of a case. Pediatr Neurosurg. 2009;45:396–401. DOI: 10.1159/000260911.
3. **Antinheimo J, Sankila R, Carpen O, Pukkala E, Sainio M, Jaaskelainen J.** Population-based analysis of sporadic and type 2 neurofibromatosis-associated meningiomas and schwannomas. Neurology. 2000;54:71–76. DOI: 10.1212/WNL.54.1.7.1.
4. **Woodruff JM, Selig AM, Crowley K, Allen PW.** Schwannoma (neurilemoma) with malignant transformation. A rare, distinctive peripheral nerve tumor. Am J Surg Pathol. 1994;18:882–895. DOI: 10.1097/00000478-199409000-00003.
5. **Sridhar K, Ramamurthi R, Vasudevan MC, Ramamurthi B.** Giant invasive spinal schwannomas: definition and surgical management. J Neurosurg. 2001;94(2 Suppl):210–215. DOI: 10.3171/spi.2001.94.2.0210.
6. **Tonomura H, Hatta Y, Nagae M, Takatori R, Kubo T.** Posterior resection of fifth lumbar giant schwannoma combined with a recapping transiliac approach: case report and technical note. Eur J Orthop Surg Traumatol. 2018;28:1209–1214. DOI: 10.1007/s00590-018-2178-y.
7. **Chiang ER, Chang MC, Chen TH.** Giant retroperitoneal schwannoma from the fifth lumbar nerve root with vertebral body osteolysis: a case report and literature review. Arch Orthop Trauma Surg. 2009;129:495–499. DOI: 10.1007/s00402-008-0635-5.
8. **Yu NH, Lee SE, Jahng TA, Chung CK.** Giant invasive spinal schwannoma: its clinical features and surgical management. Neurosurgery. 2012;71:58–66. DOI: 10.1227/NEU.0b013e31824f4f96.
9. **Boriani S, Weinstein JN, Biagini R.** Primary bone tumors of the spine. Terminology and surgical staging. Spine. 1997;22:1036–1044. DOI: 10.1097/00007632-199705010-00020.
10. **Fehlings MG, Nater A, Zamorano JJ, Tetreault LA, Varga PP, Gokaslan ZL, Boriani S, Fisher CG, Rhines L, Bettegowda C, Kawahara N, Chou D.** Risk factors for recurrence of surgically treated conventional spinal schwannomas: analysis of 169 patients from a multicenter international database. Spine. 2016;41:390–398. DOI: 10.1097/BRS.0000000000001232.
11. **Bunc G, Kramberger S, Kovacic S, Seruga T, Vorsic M.** Recurrent giant invasive thoracolumbar schwannoma. Wien Klin Wochenschr. 2004;116 Suppl 2:93–96.
12. **Ozdemir N, Bezircioglu H, Akar O.** Giant erosive spinal schwannomas: surgical management. Br J Neurosurg. 2010;24:526–531. DOI: 10.3109/02688697.2010.487129.
13. **Pongsthorn C, Ozawa H, Aizawa T, Kusakabe T, Nakamura T, Itoi E.** Giant sacral schwannoma: A report of six cases. Ups J Med Sci. 2010;115:146–152. DOI: 10.3109/03009730903359674.
14. **Alfieri A, Campello M, Broger M, Vitale M, Schwarz A.** Low-back pain as the presenting sign in a patient with a giant, sacral cellular schwannoma: 10-year follow-up. J Neurosurg Spine. 2011;14:167–171. DOI: 10.3171/2010.10.SPINE1015.
15. **Kataria R, Kurmi DJ, Mehta J, Sinha VD.** Giant lumbosacral schwannoma in a child. J Pediatr Neurosci. 2012;7:126–128. DOI: 10.4103/1817-1745.102576.
16. **Iizuka Y, Iizuka H, Kobayashi R, Mieda T, Takagishi K.** Giant schwannoma with extensive scalloping of the lumbar vertebral body treated with one-stage posterior surgery: a case report. J Med Case Rep. 2014;8:421. DOI: 10.1186/1752-1947-8-421.
17. **Valle-Giler EP, Garces J, Smith RD, Sulaiman AR.** One-stage resection of giant invasive thoracic schwannoma: case report and review of literature. Ochsner J. 2014;14:135–140.
18. **Togral G, Arikan M, Hasturk AE, Gungor S.** Incidentally diagnosed giant invasive sacral schwannoma. Its clinical features and surgical management without stability. Neurosciences (Riyadh). 2014;19:224–228.
19. **Khan UA, Ismayl G, Malik I.** Giant sacral schwannoma treated with a 360 approach: a rare case and systematic review of the literature. World Neurosurg. 2018;115:65–72. DOI: 10.1016/j.wneu.2018.03.203.

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Received 25.09.2018
Review completed 26.11.2018
Passed for printing 30.11.2018

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