CASE STUDY

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Presacral schwannoma. Case description

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ABSTRACT

Schwannomas in the presacral region of vertebral column occur sporadically and are usually diagnosed incidentally during diagnostic procedures applied as a response to nonspecific complaints associated with vertebral column or abdominal cavity. This study focuses not only on the presentation of the case of the patient with giant schwannoma in the retrorectal area, but on the highlighting of the problems associated with diagnosis and treatment of tumours located in this anatomic region as well. The presented case involves a 23-year old woman. The diagnosis of the disease was made during gynecological examination accompanied by ultrasonography of pelvic organs. Neurological examination disclosed no deviations from the normal condition. MR imaging allowed to determine precise location of the tumour and its anatomic relations to pelvic visceral and vascular structures. The patient underwent a successful surgery using laparotomy. Histological examination revealed structures of schwannoma. Surgical radicality and the lack of relapse were confirmed by MR imaging taken five years after the surgery.

Keywords: schwannoma, presacral space, surgery.

Introduction

Tumours of the presacral regions occur at the frequency of one case per 40,000 persons admitted to hospital [1]. Schwannoma accounts for 3% to 3.2% tumours in this anatomical region. Abernanthey et al. (1986), working in Mayo Clinic in the period of 33 years collected a series of only 13 giant schwannomas of the presacral region [2].

The presacral region, also referred to as retrorectal region, is frontally confined by the rectum, dorsally by the sacral and coccygeal bones and on the inferior side by muscles of pelvic floor. Lateral limits are marked by ureters and iliac blood vessels [3].

Tumours of the spinal presacral region pose a complex problem, requiring collaboration of specialists in surgery, gynecology, urology and neurosurgery.

The objective of the study is the presentation of the case of a 23-year-old female with a giant schwannoma in the retrorectal region and the analysis of the problems resulting from diagnosis and surgical treatment of tumours located in this anatomic region.

Case description

23-year-old female patient during routine gynecological examination was diagnosed with a tumour of small pelvis. The tumour was confirmed by per rectum examination and transrectal ultrasonography (T-USG). Magnetic resonance imaging (MR) of abdominal cavity (**Figure 1a, b**) disclosed pathological solid mass (in size of 130 mm x 74 mm x 63 mm) with sharply outlined margins in the presacral area in small pelvis. In sagittal plane the tumour was located beginning at lower edge of L5 vertebral body down to interface of S2–S3 sacral segments, inducing an osteoblastic reaction in the sacral bone on its left side and bulging through widened sacral foramina at the level of S1 and S2 segments on the left side. In T2-weighted images the

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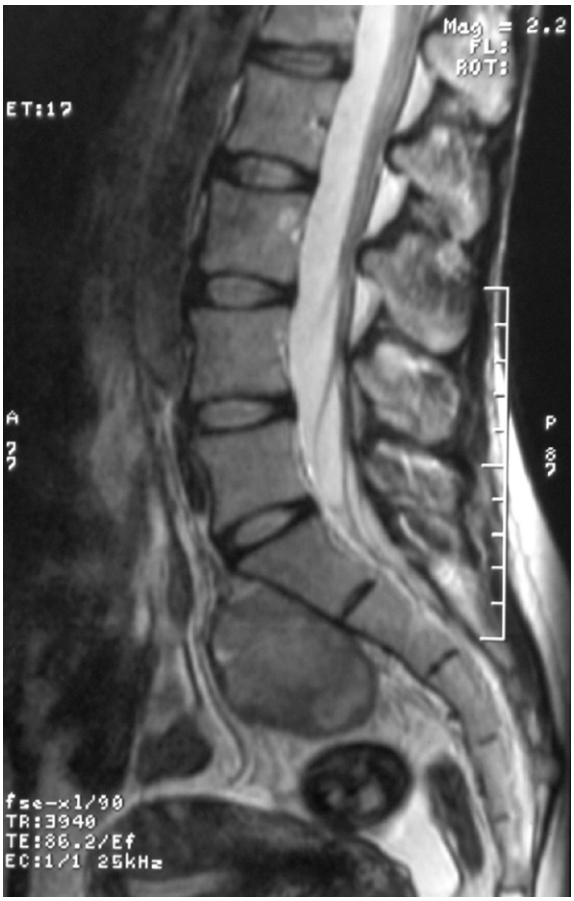

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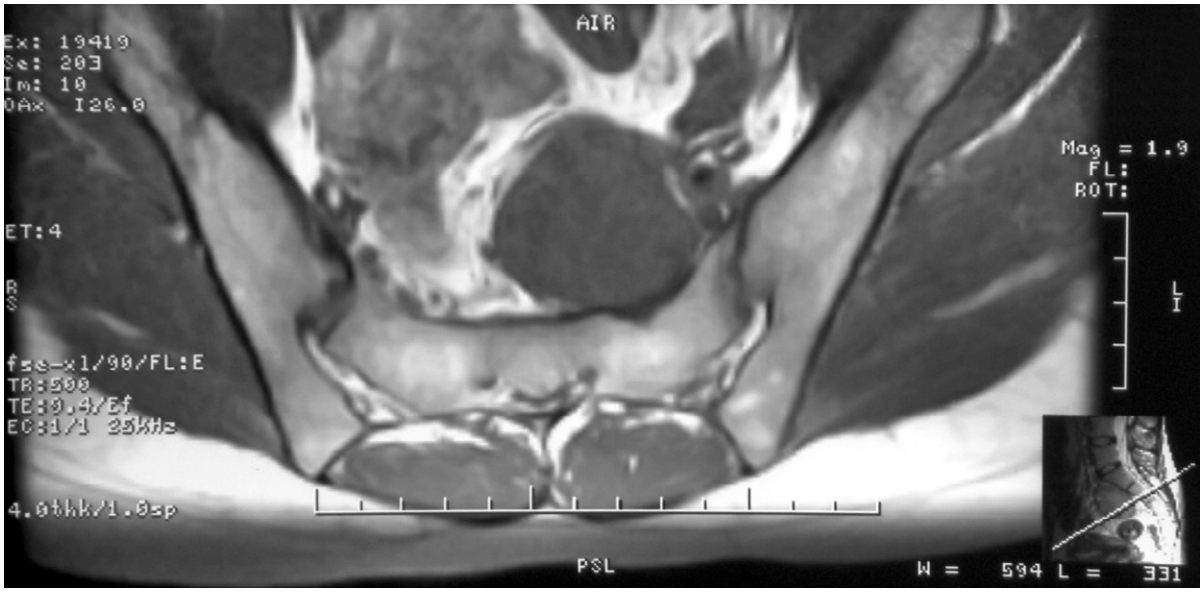

Figure 1. MRI in (a) sagittal and (b) axial projections presents location of the tumour in the presacral region

tumour manifested a non-uniform signal intensity and the examination disclosed fine foci of destruction within the tumour. The tumour, out of necrotic areas, demonstrated evident amplification following intravenous administration of paramagnetic contrast agent (gadolinium). The tumour compressed and modelled uterine corpus. The left ovary manifested normal structure, was compressed and frontally and medially displaced. Left parametrium with urinary bladder were also compressed. The radiologist put forward the diagnosis of schwannoma.

The patient was admitted to the Department of Neurosurgery in a good general condition, with no complaints reported. Neurological examination

revealed no pathology. Preoperative supplementary tests were made including urography, which demonstrated unitemporal, normal urinary excretion of contrast agent by both kidneys and a normal calyces/pelvic system in both kidneys. The left ureter was normal in its upper segment while at the level of L5 vertebra and S1 segment of sacral bone it was widened to 9 mm, with no peristaltic movements and from the level of S2 segment to its ostium in the urinary bladder it was narrow, with irregular wall surface. The right ureter in its upper fragment manifested normal course with preserved peristalsis but in its lower portion, beginning at the upper limit of the sacral bone it did not show up. Its bladder ostium was visible but narrow, modelled on the tumour mass. The tumour compression on the urinary bladder resulted in its asymmetric filling with urine. The tests ordered by the consulting surgeon: rectoscopy, sigmoidoscopy and barium enema, demonstrated no lesions within anus and colon.

The surgery by means of laparotomy was made in general endotracheal anesthesia. The vertical incision within abdominal skin was made in the midline, bypassing the umbilliculus. The incision of peritoneum and the separation of intestinal loops enabled the access to prevertebral space, uncovering the division of aorta into common iliac arteries, the outflow of common iliac veins into inferior caval vein, L4, L5 vertebral bodies and a segment of S1 vertebra. The tumour was positioned in front of the vertebral column, between lower margin of L5 vertebral body and S3 segment of the sacral bone. The tumour, (120 mm x 90 mm x 80 mm in size), on its left side was strictly connected to the sacral bone. The ureters passed in the tumour capsule on its both sides. At the next stage of the surgery, by means of microsurgical technique, the ureters were dissected free from the tumour capsule. In the back, on the right side, the inferior caval vein adhered to the tumour capsule. After dissecting the vein from the tumour capsule, the accreting tumour was separated from the sacral bone: using a surgical microscope the pathological mass was separated from roots of S1 and S2 sacral nerves on the left hand side and from the front surface of sacral bone it was fused with. Continuity of the nerve roots was preserved. The tumour was completely dissected, with blood loss of 700 ml. After assuring hemostasis in the site, the wound was closed in layers, with a drain remaining in the retroperitoneal space. To alleviate the pain a drain was introduced to extrameningeal space of the vertebral canal, which provided the potential for the administration of 0.5% Bupivicaine.

The macroscopic dimensions of the dissected tumour were 120 mm x 80 mm x 60 mm (**Figure 2**). The cross-section of the tumour was macroscopically uniform and grey-white in colour. In macro- and microscopic evaluation the tumour had an evident capsule of connective tissue, its texture was solid and contained moderately high number of cells. The elongated, spin-

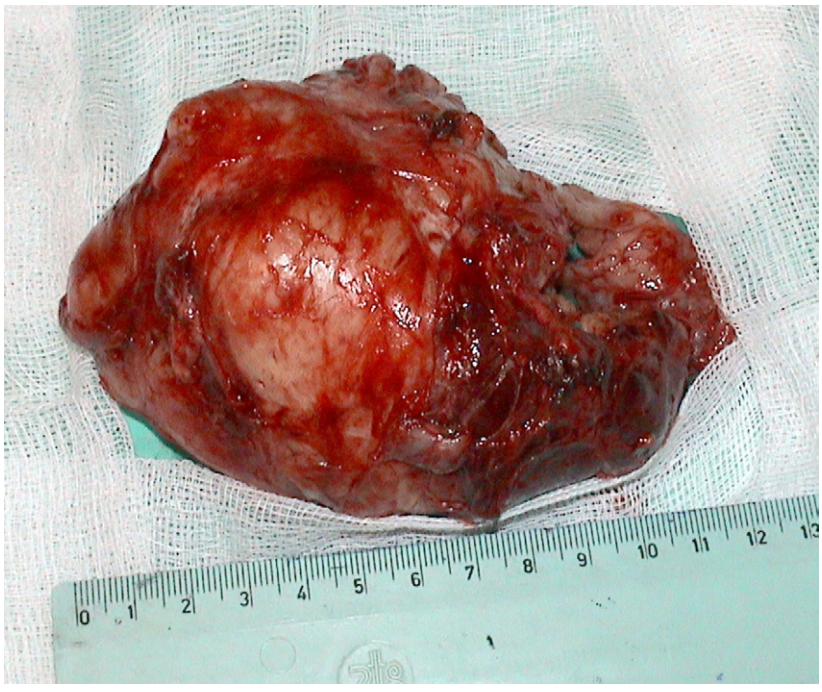

Figure 2. Macroscopic outlook of the tumour

dle-shaped cells manifested a slightly blurred margins of cytoplasm. The cells formed parallel or interwoven bands. The nuclei of cells were elongated, rod-shaped and positioned in the long axis of a cell. Nuclear chromatin had uniform character, with nucleolus noted only occasionally. Palisade set-ups of cell nuclei were not encountered in the examined case. Occasionally, the tumour texture contained fibrous acellular regions and foci of xanthomatous cells. The vascular supply was relatively rich. The blood vessels frequently manifested thickened walls, with the presence of homogenous hyaline masses. Routine histological examination of the surgical material allowed for the diagnosis of schwannoma, manifesting, according to WHO, I degree of biological malignancy (**Figure 3**).

The post-operational course was free of complications. After healing of the post-operational wound, on the ninth day following the operation, the patient showed a normal condition in neurological examination, complained of no pain and was released home.

The patient continued her studies, then took up professional work, staying under control in the outpatient clinic. Currently the patient reports no complaints and consecutive neurological examinations demonstrate no deviations from a normal condition. MR control examination of lumbal/sacral vertebral column made 5 years after the operation revealed a condition following a complete removal of the tumour (**Figure 4**).

Discussion

Schwannomas in the retrorectal region are slowly growing tumours leading to transplacement of surrounding anatomic structures. They are observed more frequently among females [4, 5].

The tumour evokes ailments when it reaches a large size. The clinical symptoms result from compression from the tumour on the neighbouring anatomic structures: nervous, vascular and visceral. Most frequently the patient complains of a discomfort in abdominal

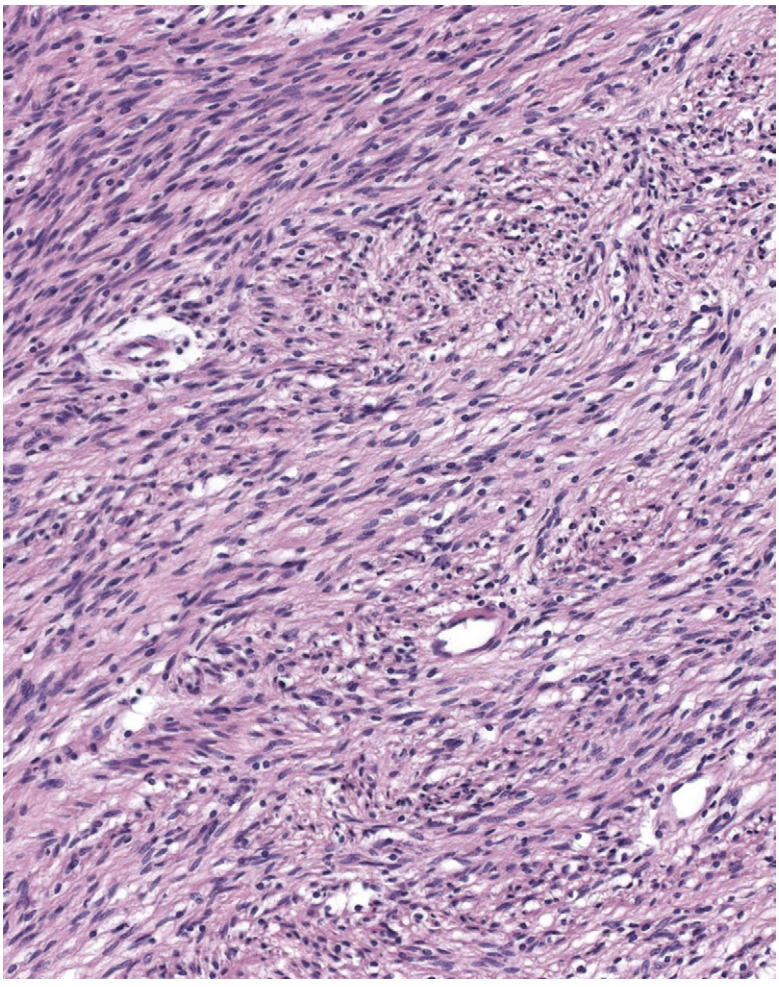

Figure 3. Histological preparation of the tumour. H&E staining, magnification 150 x

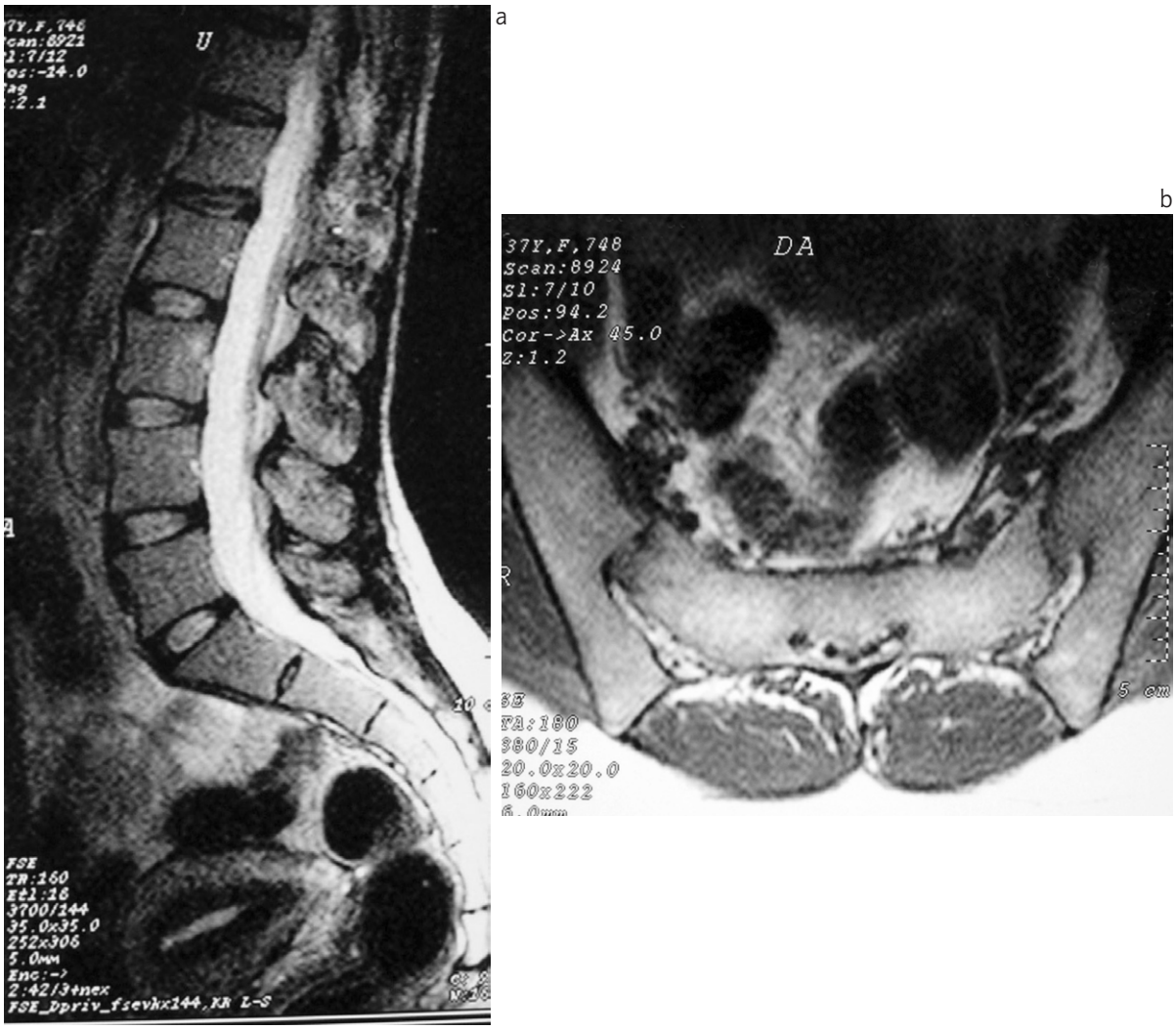

Figure 4. Control MRI in sagittal projection (a) and axial projection (b) presents the condition following complete excision of the tumour

cavity, lumbosacral pain, pain in the lumbar/sacral region, hypogastric pain radiating to groins, frequent urination or difficulties in passing urine. Some patients complain of unpleasant sensation of a filled anus and constipation. Sporadically, neurological defects are also observed [6, 7].

The symptoms are usually nonspecific and in over 26% the tumours are completely asymptomatic [3]. In such cases presacral schwannomas are recognised during physical examination of the abdominal cavity, gynecological examination "per rectum" or during radiological examination made due to nonspecific complaints [6]. This confirms our case, in which suspicion of a tumour in the presacral region was put forward by a gynecologist during a routine gynecological examination. The presence of a tumour in the retrorectal space may hamper a delivery of a newborn [6, 7].

Tumours located in this anatomic region may manifest an inborn character (meningeal hernias, dysontogenetic tumours), be of primary origin (chordoma, osseous giant cell tumour, chondrosarcoma, immature neuroma), of metastatic or inflammatory character [3, 6].

Apart from the above mentioned tumours, schwannoma has to be distinguished from ovarian tumours, ureteral tumours, retroperitoneal sarcoma or abscess in iliopsoas muscle [8–11].

Nevertheless, despite such broad range of differential diagnosis, similarly to the discussed case, the classical variant of schwannoma presents no diagnostic difficulties in the postoperative material already on the level of routine morphological techniques. In doubtful cases, immunohistochemical techniques and occasionally electron-microscopic techniques turned out to be useful.

Schwannoma stems from ventral roots of sacral nerves. In plain radiography and computer-assisted tomography (CT) lesions of erosion type and irregular destruction of frontal pelvic foramens of sacral bone may be evident [12]. In CT examination and in magnetic resonance imaging (MRI) schwannoma is very well confined, manifests smooth edges and transplaces the neighbouring anatomic structures. The tumour is frequently featured by a heterogenous pattern, with centrally located cysts and it manifests marginal augmentation following administration of a contrast agent [4]. Lesions of cystic type are present much more frequently (in 60%) in schwannomas than in other tumours [4, 13]. MRI patterns of schannomas present hemorrhagic, necrotic and calcified foci. The lesion is hypointense on T1 weighted images and hyperintense on T2 weighted images [14].

Needle biopsy of tumours in the retrorectal region is a safe approach to make the diagnosis strict. Histological evaluation of the obtained material in 95% cases allows to conclude whether the lesion is of a benign or a malignant character. The tumour type can be precisely defined in 81% cases [15]. Indications for needle biopsy in cases of schwannomas are controversial due to their sufficiently distinct image in MRI examination. Proponents of needle biopsy are of the opinion that such a biopsy should be made in patients with the tumour evoking no clinical symptoms and the patient does not decide to undergo surgery [7].

The size and location of the tumour have to be very precisely determined prior to surgery. A particular attention should be paid to position of arterial and venous blood vessels in the anatomic region, to the determination of tumour relationships to ureters, urinary bladder, posterior wall of rectum and other closely positioned anatomic structures. The risk of intraoperative injuries has to be evaluated before taking the decision about the surgery. In our case, this led to the extension of diagnostic procedures (apart from MRI) by rectoscopy, sigmoidoscopy, barium enema to the large intestine.

Depending of tumour location, size, relationships to other anatomic structures and type of tumour in the presacral region various surgical approaches are applied, including frontal access (through the peritoneal cavity), posterior access (through the sacral region), frontal and posterior in parallel, transrectal and transvaginal one [3, 6, 16]. Schwannomas are approached through the frontal access. In the case of small dimension tumours the endoscopic technique is used with transrectal or transvaginal operative approach [17].

Surgical treatment of schwannoma aims at its complete removal. Problems with reaching the aim stem from the size of the tumour, its rich blood supply and vicinity to important anatomic structures. The tumours in this area are usually supplied with blood through sacral arteries, medial and lateral arteries, lumbar arteries and internal iliac arteries.

During the surgery, the surgeon should focus on preserving continuity of sacral nerve roots (S1, S2, S3) which exit from sacral bone through pelvic foramina, fused with the capsule of the tumour. Damage to the nerves results in dysfunction of vesical and anal sphincter muscles, disturbs sexual functions and leads to paresis of greatest gluteal muscle [1]. In order to preserve the continuity of nerve roots S1 to S3 a microsurgical technique has been applied during our operation.

Complete excision of a tumour in the presacral region is possible in cases of benign tumours and in three quarters of cases of malignant tumours. A partial tumour resection is made when there are no chances for a complete removal of the pathological lesion [7]. In such cases relapses if schwannoma are described, usually within a time distance of a few years [18].

Complications occur quite frequently, in 1/3 of the cases, but they are usually of reversible character [3]. They most frequently include the injury to venous or arterial blood vessels, a disturbed function of vesical and anal sphincters and in men disturbances in erection. Deficits in sensation in perineal region and frontal surface of thighs were also described [2, 3, 18].

In patients with asymptomatic schwannoma the possibility of conservative treatment with periodic control of MR examination should be taken into account. Strauss et al. treated 28 patients with schwannoma in retroperitoneal space, out of which 8 had the tumour accidentally detected due to their nonspecific complaints and signs in MR imaging [7]. The diagnosis of schwannoma was established by needle biopsy and the patients were subject to further observation. After 32 months no progression of the tumour was revealed by MRI control, the patients reported no pains and neurological examination detected no abnormalities. However, other authors reported very good results of surgical treatment applied to asymptomatic presacral schwannomas [19, 20].

Application of radiotherapy in cases of incomplete excision of schwannoma is controversial and rather not recommended as it may induce the transformation of schwannoma cells and the development of malignant neuroma [21, 22]. Stereotactic radiosurgery is recommended in cases of small schwannomas,

with the diameter of less than 30 mm, positioned in the sacral bone [23].

Conclusions

Giant schwannomas of sacral bone occur rarely and pose diagnostic and therapeutic difficulties. They manifest a slow local growth and induce nonspecific symptoms.

Their preoperative procedures require magnetic resonance imaging of lumbosacral vertebral column and multispecialistic evaluation of pelvic visceral structures.

Complete tumour excision is equal to cure.

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Conflict of interest statement
The authors declare that there is no conflict of interest in the authorship or publication of contribution.

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