

Retroperitoneal giant schwannoma eroding lumbal vertebra: a case report with a literature review

Case Report

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Abstract: A huge schwannoma, located in the retroperitoneal space, is found very rarely. The main purpose of this paper is to present the case of a giant retroperitoneal schwannoma which partly invaded L4 vertebral body. The secondary purpose is to summarize the case-report articles on retroperitoneal schwannomas. A circumscribed heterogenic tumour was revealed on transabdominal sonography. It extended into the right retroperitoneal space. CT and MRI revealed a paravertebral tumour in the size of 11 cm × 9 cm, which is causing a partial lysis of L4 vertebral body (15% of vertebral capacity), expanding intravertebral foramen and filling the right retroperitoneal space. A preoperative core needle biopsy was performed and a benign schwannoma was diagnosed. A complete surgical excision of the tumour was achieved by a two-step operation. During the first step, the neurosurgeons made L4 hemilaminectomy, facetectomy and a transverse process resection by posterior extended approach. The general surgeons excised the residual retroperitoneal part of the tumour by midline transabdominal approach 10 days later. The diagnosis of benign schwannoma was verified histochemically. There were no sign of tumour recurrence or spine destabilization at the six-month follow-up. In conclusion, although majority of giant retroperitoneal schwannomas can be completely removed performing one-step operation, a preoperative consideration about rationality of two-step operation should be mandatory when tumour destructs a part of vertebral body. Our case shows that the combined two stage complete surgical excision of a giant retroperitoneal schwannoma, eroding 15% of L4 vertebra's osseous capacity, is effective and does not have any negative influence on spinal stability.

Keywords: Schwannoma • Neurilemmoma • Retroperitoneal neoplasm • Surgery

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1. Introduction

Schwannoma is quite a common benign peripheral nerve sheath tumour. It arises from the Schwann cells, is circumscribed and appears encapsulated by epineurium, bulges eccentrically from the originating nerve [1-3]. Tumour size does not usually exceed a diameter of 5-6 cm. Large schwannomas are found very rarely, only 0.3-5% of all cases [1-7]. Most of them are found in the retroperitoneal space.

The goal of retroperitoneal schwannoma surgery is to achieve a complete removal of the tumour and preserve neurological function. Totally excised benign schwannoma shows no recurrence and no adjuvant therapy for this benign lesion is recommended [6,8-10]. Giant-sized retroperitoneal schwannoma is a challenge to the surgeons in terms of the best surgical approach and surgical radicality. Retroperitoneal schwannoma eroding lumbal vertebrae is even more specific tumour, because of its rarity, diverse and non-traditional surgical strategy, lack of specified literature and reviews.

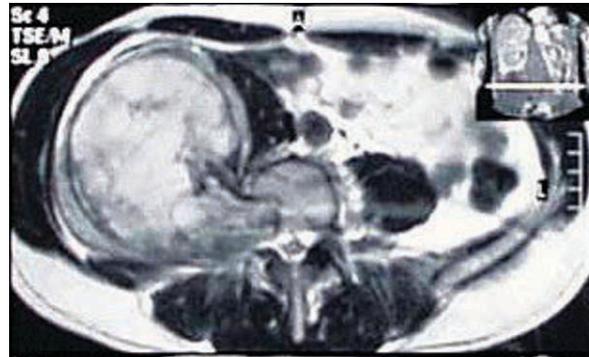
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The primary objective of this paper is to present the experience with a case of a giant retroperitoneal schwannoma, invasive into L4 vertebral body, which was treated by a two-stage combination surgical approach. The secondary objective is to summarize the case report articles on retroperitoneal schwannomas selected from Medline database.

2. Case report

A 56 year-old male complaining of the skin sensitivity disorder in the right lower extremity and a palpable bulging mass in the right iliac region for a period of one year was admitted to the department of surgery. Physical examination revealed a solid mass filling the right lower abdomen and anaesthesia by L4 radix of the right leg. A routine laboratory study showed no changes. A circumscribed heterogenic tumour with hypoechogenic insertions inside was revealed on transabdominal sonography. It extended into the right retroperitoneal space. CT and MRI revealed a paravertebral tumour in the size of 11 cm × 9 cm, which is causing a partial lysis of L4 vertebral body (15 % of vertebral capacity), expanding intravertebral foramen and filling the right retroperitoneal space (Figure 1 and 2). A preoperative core needle biopsy was performed and the diagnosis of a benign schwannoma was recorded. Due to the large size of the tumour, its retroperitoneal location, erosion of a vertebral body and high possibility of intraoperative complications, it was decided that a two-step operation should be performed. During the first step, the neurosurgeons made L4 hemilaminectomy, facetectomy and a transverse process resection by posterior extended approach. Considering the large size of the tumour, the transverse paraspinous muscles incision was made for better approach. The tumour derived from the L4 spinal nerve root extradurally, 1cm from

Figure 2. Magnetic resonance image showing a large mass in the retroperitoneal space (T2-WI).



dural sac and was located laterally within the vertebral foramen, extending extraspinally. The L4 nerve root was inseparable from the tumour, so it was sacrificed just medially from the spinal ganglion. After debulking and internal decompression, the tumour was removed from vertebral foramen and dissected from the vertebral body. The second step operation was performed after a period of 10 days. The general surgeons excised the residual retroperitoneal part of the tumour by the midline transabdominal approach.

After the second operation the patient complained of numbness, anaesthesia of the anterior region of the right thigh. Neurological examination confirmed the postoperative L2 – L5 radiculopathy with right leg proximal paresis (quadriceps muscle power - 0). There were no other postoperative complications.

Macroscopically a well capsulated, firm, gray color tumour was of 14×12×5.5 cm size. Dissection of the tumour showed a gray solid mass with some degenerative areas and cysts, brown and yellowish color zones (Figure 3). Microscopic examination revealed that tumour is composed of spindle cells with wavy appearing nuclei. Hypocellular, loosely textured tumour areas (Antoni B) alternate with areas of hypercellularity

Figure 1. CT scan shows tumour eroding L4 vertebral body.

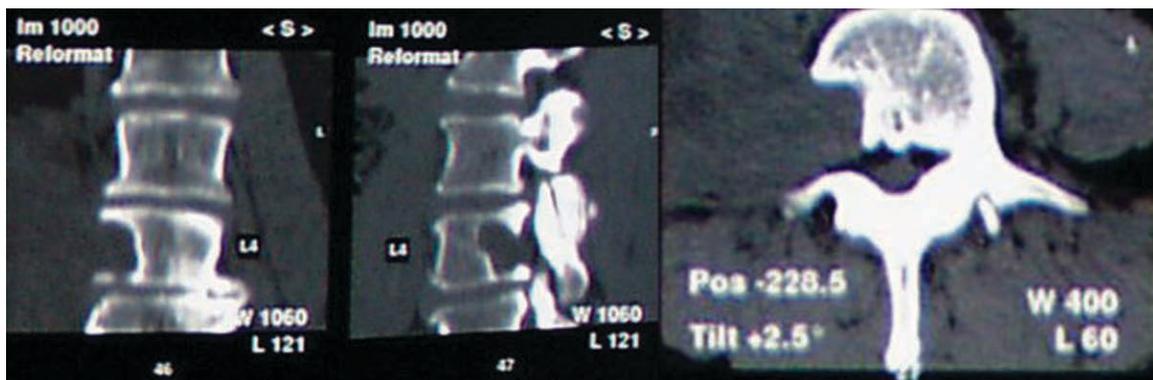
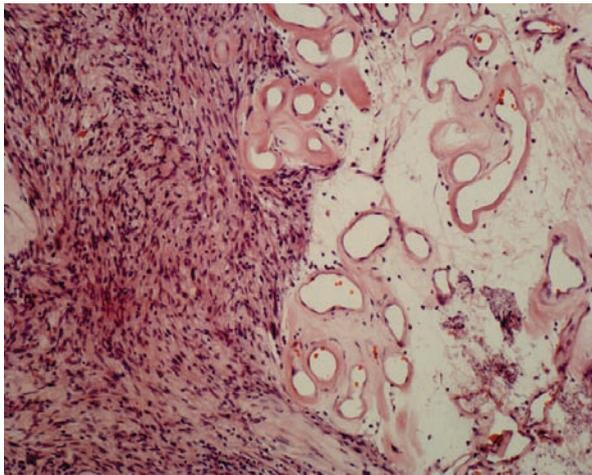


Figure 3. Dissection of the tumour shows a gray solid mass with degenerative areas, cysts and yellowish and brown color zones.



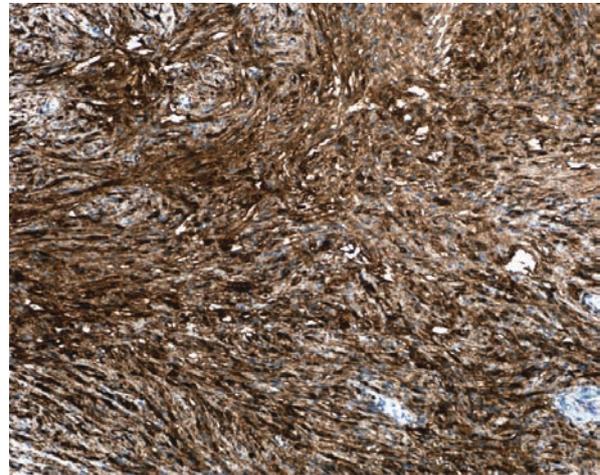
Figure 4. Schwann cells arranged in hypercellular (Antoni A) and hypocellular (Antoni B) areas.



(Antoni A), with occasional nuclear palisading (Figure 4). Mitotic activity is rare and there is a focal mild nuclear pleomorphism in the tumour. A variety of degenerative changes were found, including xanthoma cells, vessels with hyalinized walls. Tumour cells strongly and diffusely express S-100 protein (Figure 5), vimentin and focally express glial fibrillary acidic protein. Immunoreactivity of tumour cells is negative for CD117, CD34, desmin, CD57, alpha smooth muscle actin. These histopathological findings proved the diagnosis of a benign schwannoma.

The patient was seen and examined after 6 months and there were no signs of disease recurrence on CT scanning. The spinal column remained stable and did not necessitate additional fixation. Nerve function improved. A positive postoperative dynamic was observed on neurological examination: quadriceps muscle power increased from 0 to 3 points on manual muscle testing.

Figure 5. Strong immunoreactivity for S-100 protein in schwannoma.



3. Discussion

Schwannoma is also named neurolemoma, neurilemoma, neurinoma, perineural fibroblastoma, neurilemmoma, neurolemmoma. It is considered as a benign lesion. However, there is a lot of difficulty rejecting or confirming the malignancy. Discussing about the tumour's malignant or benign features, the attention must be paid to the size of the tumour, biological behavior, atypical histology, characteristic immunohistochemical staining and radiological findings.

When the tumour localizes retroperitoneally, it usually reaches a big size. A small well-encapsulated, non-adherent lesion within the substance of peripheral nerve, with axon that does not penetrate it, is most likely a benign schwannoma. But a large (>3 cm) lesion that is adherent to the surrounding tissues is likely a malignant schwannoma. In some literature the suspicion of the malignancy is based on the tumour size (more than 3 cm) [6].

The large sized tumour causing compression to the surrounding structures and eroding the bony structures simulates an invasion process. In addition, a rapid growth and invasion of adjacent structures are other features of a suspected malignancy. Malignant schwannomas act as a high grade sarcomas: give a local recurrence and distant metastasis. Von Recklinghausen's disease has a close association with malignant schwannomas [8,9,11].

The necessity of a preoperative biopsy is stressed by many authors, though it is not always successful or sometimes can not reject the possibility of malignancy. Ultrasonographically or endosonographically guided, CT guided fine needle aspiration or core needle biopsies

are performed to define the diagnosis [5,12-14].

Schwannomas have distinguishing histological features: spindle-shaped Schwann cells arranged in hypercellular (Antoni A) and hypocellular (Antoni B) areas, nuclei arranged in a palisading fashion. Focally, the nuclei may be enlarged and pleomorphic appearing. A mitotic activity is rare. The degenerative changes including hemorrhage, xanthoma cells, cyst formation, vessels with hyalinized walls may be found. The proliferative index Ki-67 is positive in less than 5% of tumour cells [11]. Routinely the schwannomas cells strongly and diffusely express S-100 protein. It helps in differentiation this type of tumour from the malignant peripheral nerve sheath tumours (MPNSTs). Usually only scattered peripheral nerve sheath tumour cells express S-100 protein and, additionally, it happens in 50%-70% of the malignant peripheral nerve sheath tumours (MPNSTs) [5,11,15-17].

No specific radiological features concerning schwannomas are found. But some signs can be suggestive: well circumscribed rounded mass with heterogeneous contrast enhancement due to cystic and hemorrhagic changes, calcification and cystic changes in big tumours. 40% of schwannomas demonstrate cystic components [3,5,18,19].

MRI seems to offer a better resolution and more specificity than CT and can better delineate the tumour from the surrounding structures. CT images failed to adequately reproduce a stroma heterogeneity, the main characteristic sign for the ancient Schwannomas. Schwannomas possess low signal intensity in T1-weighted images similar to muscle and a high signal on T2 weighted images similar to fat. There are recommendations beside standard T1 and T2 weighted images to make a fat suppression sequence (STIR) on which the schwannoma will maintain its high signal allowing a delineation from pure lipomatous tumours [18]. Schwannoma typically enhances dramatically with gadolinium contrast on MRI imaging. The malignancy sign can be asymmetry, irregular infiltrative borders, big tumour size, infiltration, heterogenic pattern and mixed intensity [3,20-23].

Giant invasive retroperitoneal schwannomas are defined as lesions that erode spine vertebrae and extend posterior and laterally into myofascial planes. They are found very rarely and it is too little literature how to treat such lesions [24-28].

Giant invasive schwannomas growing in all directions is a challenge to the surgeon. The usual objects of discussion are the surgical approaches and techniques. Even a minimal invasive surgery such as endoscope-assisted minilaparotomy or a laparoscopic surgery of retroperitoneal tumours has been mentioned

in the literature as the choice of the operative technique [29-34]. Another question is whether to perform only a local excision of the tumour or tumour resection together with surrounding tissues. In both ways, the nerve roots must be preserved if it is possible. However, big-size tumours usually extend over the several vertebral levels, go anterolaterally into the extraspinal space via foramina, which they erode and widen. Also they erode vertebral bodies to the varying extent. The resectability of the tumour and the stability of the spine are other topics under consideration. The amount of bony lesion suggests if the patient requires additional spine stabilization procedure and a bed regime for the several months after the operation [32].

Twenty-five suitable cases of retroperitoneal schwannomas based on Medline database were found [1-5,12-14,18,20,29-31,33,35-40]. The main characteristics of these report cases are summarised in the Table 1.

Specifically, retroperitoneal schwannoma predominated in women. This tumour was confirmed in 16 women (64%) and in 9 men, within all age groups (min – 25, max – 73). The smallest tumour was 2 cm in size (patient no. 6), the biggest one up to 21 cm (patient no. 18).

Vague abdominal pain, hypertension, slight local pain radiating or not to the legs (anterolateral thigh area), abdominal discomfort, slowly growing lower abdomen or unilateral inguinal swelling, dilated superficial suprapubic and scrotal veins, palpable mass transabdominally, rectally or vaginally, and secondary specific urological symptoms like urinary incontinence, colicky pain, hematuria were the most frequent symptoms of retroperitoneal schwannomas. This was observed in 17 symptomatic patients. In general, clinical symptoms are not specific. The hypoaesthesia or mild nerve deficit is rather characteristic. The time from onset of symptoms to diagnosis takes up to several years. Giant schwannomas can cause compressions to the surrounding organs and mimic different pathology such as spine disc herniation, pancreatic cyst, adrenal lesion, psoas muscle abscess, hepatic tumor, bowel or urinary bladder dysfunction and others [1,2,4,13,37].

Clinically asymptomatic retroperitoneal schwannomas were found in 8 patients (32%). The tumours were diagnosed accidentally mostly by performing ultrasound investigation. CT and MRI were the next diagnostic modalities for further more exact description of the tumours in terms of anatomical topography, invasiveness and radiological features. CT or US-guided tumour tissue FNA or core needle biopsy was performed in 13 patients (52%). The smallest biopsied tumour was of 3 cm size. In this case, a FNA

was inconclusive. In most other cases only the huge tumours were biopsied. In two patients, core needle biopsy was made transrectally or transvaginally. Only the first one was rightly conclusive. In total, preoperative verification of schwannoma diagnosis was obtained only in 7 out of 25 patients. Other preoperative diagnoses were the following: adrenal gland; renal; presacral; ovarian; hepatic caudate lobe (HCV positive) tumours; extra-adrenal pheochromocytoma; extra-adrenal ganglioneuroma; retroperitoneal lymphoma; and, a cold psoas abscess.

Different surgical approaches were mentioned: hand-assisted transperitoneal laparoscopic excision, laparoscopic resection, mini-invasive anterior retroperitoneal approach, parasagittal incision, anterior transabdominal approach, laparotomy through a median incision, suprapubic incision, transverse suprapubic incision, external incision over the swelling, posterior approach (sacral laminectomy), oblique skin incision in the left axillary line. The laparoscopic technologies were used in 6 patients (no.1, 6, 14-16, 23). A maximum size of the tumour removed laparoscopically was 8 cm in size. However, three of them were up to 3 cm in size. In general, the type of incision depended on the tumour location. All patients but one underwent one stage surgical procedure. The patient numbered 17 underwent a two stage surgical procedure – sacral laminectomy and one month later a transabdominal presacral tumour portion excision.

Immunohistochemistry revealed strong expression of S-100 in all patients. On the contrary, reactions for actin, desmin, CD34 and CD117 were negative. Vimentin was positive in 2 cases. In addition, immunohistochemistry for neuron specific enolase was done in one case and it appeared to be positive.

It is difficult to define the origin of retroperitoneal schwannoma. However, in some cases, the origin of schwannoma was emphasized: S2, L3, L5-S1 nerve roots. By the way, S2 nerve root appeared to be the most common mentioned – 3 cases out of 6.

There were no postoperative mortalities amongst the 25 patients. Nevertheless, operative complications and postoperative morbidity in some of them were very obvious. There was a clear observation concerning the significant blood loss in patients numbered 12, 21, 24 and 25. The highest amount of blood loss was up to 5 litres, specifically occurred in patients numbered 24 and 25. Postoperative morbidity mostly was related with an inevitable damage to the nerve roots during the operation. The hypoesthesia of a corresponding damaged nerve root was mentioned in three patients (patients no. 3, 21, 22). The following concomitant illness associated with benign schwannoma was found

in 5 patients (patients numbered 4, 10, 11, 16 and 25): adenocarcinoma of sigmoid colon; HCV positive chronic hepatitis; obstructive nephropathy and renal stones; acoustic nerve schwannoma; and, breast cancer. There are not enough data about long-term survival after surgery due to retroperitoneal schwannoma. However, one patient control was performed after 8 years.

This review of the case-report articles confirmed that the retroperitoneal schwannoma eroding adjacent osseous structures appeared to be very rare. The involvement of the adjacent osseous structures of two patients were stated. Vertebrae of the sacrum were damaged in both patients (patients no. 12 and 17). A two-step operation was reasonably selected as the best option for one of them.

In our case, we have described a giant invasive retroperitoneal schwannoma which eroded L4 vertebral body. A preoperative biopsy confirmed the tumour being the benign schwannoma. Later, postoperative immunohistochemistry verified that. Despite a huge size of the removed tumour there were no histological signs of malignant peripheral nerve sheath tumours.

A combined surgery approach was considered for this tumour excision: two-step operation and two different surgical approaches. The vertebral osseous tissue was damaged partially, 15% of its capacity. In our opinion, such lesion does not affect the spine stability. After six months, a follow-up CT scanning showed no sign of the tumour recurrence or the spine destabilization.

In conclusion, the findings of review still confirm that it is rather difficult to make a preoperative diagnosis of any size retroperitoneal schwannoma due to lack of specific clinical symptoms or laboratory data. The computer tomography or MRI may suggest the diagnosis, but histopathological verification is needed. However, even preoperative tumour biopsy might be inconclusive. The choice of the operative strategy mostly depends on surgeon's skills, tumour location and its size. Despite the fact that majority of giant retroperitoneal schwannoma can be completely removed performing one-step operation, a preoperative consideration about rationality of two-step operation should be mandatory when tumour destructs a part of vertebral body. Our case shows that the combined two stage complete surgical excision of a giant retroperitoneal schwannoma, eroding 15% of L4 vertebra's osseous capacity, is effective and does not have any negative influence on spinal stability.

Table 1. Twenty-five suitable cases of retroperitoneal schwannomas.

No.	Sex, age	Symptoms	CT	MRI	US	Size	Biopsy	Diagnosis before operation	Operation	Origin, nerve root	Immunohisto-chemistry	Morbidity	Control, recurrence
1.	F, 49	Vague abdominal pain, headache, hypertension.	Contrast enhancing mass arising in the Lt adrenal	Gd enhancement, T1 – hypointensive, T2 - hyperintensive	n/a	7 cm	n/a	Lt adrenal lesion	Hand-assisted retroperitoneal laparoscopic excision	n/a	S-100 (+), Sma (-)	n/a	n/a
2.	F, 26	n/a	Well-circumscribed mass anterior to the sacrum	Gd enhancement, T1 - homogeneous, T2 - heterogeneous	n/a	4 cm	CT guided biopsy (+) for Schwannoma	Presacral tumour, schwannoma	Mini-invasive anterior retroperitoneal approach, parasagittal incision	Lt sacral plexus	S-100 (+)	n/a	n/a
3.	F, 19	Accidentally found in minor pelvis by US	Round mass within the sacrum expanding into the retrorectal space, expanded S2 foramen	Intrasacral and retroperitoneal mass	retro-uterine, rounded mass	8 cm+ 5x4 cm intrasacral	n/a	Presacral tumour	Anterior, transabdominal approach, suprapubic incision	S2	(+) for schwannoma	Hypa-esthesia on dorsal aspect of the Rt thigh	MRI (-)
4.	F, 67	Accidentally found by US, 6 y. slight pain in the right leg. Also polypoid adenocarcinoma in sigmoid colon	Cystic smooth mass in contact with the Rt psoas muscle	Contained fluid and lobulated mass	Retroperitoneal hypoechoic mass	9 cm	US guided FNA (-), but (+) pain in the leg	A cold psoas abscess	Median Laparotomy	L3	S-100 (+)	n/a	5 m. (-)
5.	F, 41	2 y. slowly growing right inguinal swelling, a firm mass felt in Rt adnexal region	n/a	n/a	Extra peritoneal tumour, extended into the inguinal region, ovarian mass	10x8+ 4x3 cm	n/a	Ovarian tumour	Transverse suprapubic incision and externally incision over the inguinal swelling	n/a	Sma (-), Vimentin (+)	n/a	6m. (-)

continued **Table 1.** Twenty-five suitable cases of retroperitoneal schwannomas.

	M,43	M,29	F,66	F,40	F,62
12.	Dilated superficial veins in suprapubic and scrotal areas	Mild epigastric pain, radiating to the back	Accidentally found mass in retroperitoneum, increased NA, VMA	Accidentally found by US in retroperitoneum	Accidentally found by US. Also 20 y. before acoustic nerve Schwannoma
	Well-defined heterogeneous enhancing mass with calcifications, eroding vertebra, anterior to the sacrum	Contrast enhancing hypodense mass	Solid mass with clear margins in left side of SMA	Well-defined round mass with cystic change, contrast enhancement	Well-defined round, solid mass between Rt kidney and the IVC
	Gd enhancement, T1-hypointensive, T2-intermediate intensity	n/a	T1-low intensity, T2-slightly high intensity	T1 – low intensity T2 – increased intensity	n/a
	Homogeneous mass, displaced the iliac vessels	Slightly hypoechoic mass between Lt hepatic lobe and pancreas	n/a	Mass around the kidney	Retro-peritoneal mass close to the IVC
	16x11x10 cm	3,5x2,8 cm	6x4,5x3,5 cm	n/a	8x5x3,5 cm
	(+) for Schwannoma	Endosonographically guided FNA (+)	n/a	n/a	Laparoscopic intraoperative biopsy (+)
	Schwannoma	Schwannoma	Extra-adrenal pheochromocytoma	Neural origin of the tumour	Lymphoma or neurogenic tumour
	Elective resection	Surgical resection	Elective, hand-assisted laparoscopic surgery	Endoscopic minilaparotomy	Laparoscopic resection
	L5-S1	n/a	n/a	n/a	n/a
	(+) for schwannoma with degenerative changes (ancient Schwannoma)	S-100 (+), Sma (-), Desmin (-)	S-100 (+), neuron-specific enolase (+), CD 34 (-), Sma (-), Desmin (-), with degenerative changes	(+) for schwannoma	S-100 (+)
	Blood loss from presacral vascular plexus	n/a	n/a	n/a	n/a
	1 y (-)	n/a	3y. and 2m.	n/a	n/a

continued **Table 1.** Twenty-five suitable cases of retroperitoneal schwannomas.

	F,25	M,73	F,35	F,42	F,40
17.	Mild low-back pain radiating to the quadriceps, episodes of urinary incontinence	4 y palpable mass in the abdomen	Lt parailiac mass, urinary incontinence, Lt leg pain	2 weeks Rt pelvic pain, abdominal discomfort	6 y lower back pain, urinary incontinence, painless swelling in lower abdomen
	Heterogeneous calcified mass from the sacrum with osseous changes, enlarged sacral foramina	Capsulated solid mass with cysts, necrosis, bilateral hydronephrosis	Moderately hyperdense in centre, heterogenic character mass from Lt parailiac region	n/a	Well-defined, encapsulated, central area with heterogenic change
	Intra-and presacral tumour	n/a	n/a	n/a	Gd enhancement
	n/a	9x6x6 cm	5x6 cm	7.1x5.4 cm	35x25cm
	n/a	n/a	n/a	FNA (-)	CT guided needle biopsy - ancient Schwannoma
	n/a	A solid mass between urinary bladder extending to the umbilicus	n/a	Mass from Rt adnexial area	n/a
	Posterior approach – sacral laminectomy, 1 month later transabdominal approach presacral tumour portion excision(used nerve stimulator and EMG)	Retroperitoneal tumour, uretrohydro-nephrosis	Laparotomy	Laparotomy	Transabdominal-transperitoneal approach
	S2	n/a	n/a	n/a	n/a
	(+) for schwannoma	S-100 (+)	(+) for schwannoma	(+) for schwannoma	(+) for schwannoma with degenerative changes
	n/a	n/a	n/a	n/a	Blood loss from vaginal sinus, hypoesthesia S1-2, urge/stress incontinence
	MRI (-) 1 y	6m. (-)	n/a	n/a	n/a

continued Table 1. Twenty-five suitable cases of retroperitoneal schwannomas.

	22.	M,37	18 y Lt side back pain, lateral abdominal pain, pain in anterolateral thigh	Gd enhancement	Encapsulated mass in retroperitoneum	Encapsulated mass with fluid areas	20x12cm	Soft tissue needle biopsy (+) for schwannoma	Schwannoma	Oblique skin incision in the left axillary line	n/a	(+) for schwannoma	Para-esthesia L3, lower quadriceps power	6m. (-)
	M,28	Asymptomatic, accidentally found by US in retroperitoneum	Mass with central low density area, attached to psoas major muscle	T1-low intensity, T2-increased intensity	Well-circumscribed mass between Rt kidney and IVC	2.9x2.7x2.5 cm	n/a	Extra-adrenal ganglioneuroma in the retroperitoneum	Laparoscopic resection	n/a	S-100 (+) with hemorrhage areas	n/a	6m-CT	
	M,41	6y. low back pain, some urinary urgency, large palpable pelvic mass	Solid heterogeneous pelvic mass with areas of calcification with osseus change	Well-defined heterogeneous presacral tumour	n/a	14x11x11 cm	CT biopsy (-), transrectal core needle biopsy (+)	Cellular Schwannoma	Midline laparotomy	S2 nerve root	(+) for schwannoma	Blood loss	1y. (-)	
	F,70	Several y., abdominal pain, palpable mass, large mass on vaginal and rectal examination, Also breast cancer	Well-defined retrorectal mass with low density centre, calcification in the wall, bilateral hydronephrosis secondary to a presacral mass	n/a	Mass of the ovarian origin	20x14x8 cm	Core needle biopsy through posterior fornix – cellular Schwannoma	Cellular Schwannoma	Laparotomy	n/a	S-100 (+)	Blood loss, thrombosis, pulmonary embolism	2y (-)	

Lt – left; Rt – right; Gd enhancement – tumour enhanced after administration of Gadolinium; n/a – not available; y. – years; m. – months; US – ultrasonography; Sma – smooth muscle actin; NA – noradrenalin; VMA – vanillylmandelic acid; SMA – superior mesenteric artery; IVC – inferior vena cava; EMG – electromyography

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