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Review

# Surgical management of retroperitoneal pelvic schwannoma: experiences with four cases and review of literature

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**Summary**

Schwannomas generally occur in the head, neck, and extremities; however, its occurrence in the retroperitoneal pelvic space is rare. Here, we describe four successful surgical managements of retroperitoneal pelvic schwannoma which was identified in preoperative imaging studies. Computed tomography demonstrated heterogeneous masses in the retroperitoneum with well-defined margins. The retroperitoneal pelvic schwannomas were removed by complete excision or enucleation through either laparotomy or laparoscopy. The diagnoses of schwannoma were further confirmed via histopathology. Postoperatively, two patients had transient but minimal neurologic deficits in a lower extremity. No evidence of tumor recurrence was detected over a 12-month following-up of four patients. A meticulous review of three-dimensional imaging studies provided correct preoperative diagnoses of schwannomas. Postoperative neurologic deficits may be less serious than expected. Complete resection of the tumor is the treatment of choice, and recurrence is unusual.

*Key words*: Pelvic neoplasm; Retroperitoneum; Schwannoma; Neurologic Deficits.

## Introduction

Schwannomas are benign neurogenic tumors originating from Schwann cells of the nerve sheath. Schwannomas predominantly occur in the head, neck, and extremities; 3% of schwannomas rarely develop in the retroperitoneal pelvic space [1]. Retrope ritoneal pelvic schwannomas (RPSs) occur in a variety of nerves, such as obturator nerve, sacral plexus, and genitofemoral nerve. Thus, it is important to diagnose RPSs preoperatively and assess the risk of postoperative neurologic deficits. However, it is well-known that the preoperative diagnosis of RPS is difficult due to its slow growth, and nonspecific or subclinical symptoms [1]. Furthermore, the extensiveness of RPSs hampers clinical implication of imaging study in the differential diagnosis of retroperitoneal pelvic tumors. Here, we report the clinical potentiality and usefulness of imaging-based study in preoperative diagnosis of RPSs. Computed tomography (CT) of four patients showed a heterogeneous mass in the retroperitoneum with demarcated margins without adjacent tissue invasion or enlargement of pelvic lymph nodes. Schwannomas usually appear as a well-defined mass with heterogeneous low signal intensity on T1-weighted images and high signal intensity on T2-weighted images in magnetic resonance images (MRIs) [2]. All gynecological tumor markers, such as alpha-fetoprotein, cancer antigen 125, cancer antigen 19-9, human chorionic gonadotrophin, were within normal ranges in these cases. Complete laparoscopic tumor excision was carried out in three patients and enucleation was carried out in one patient. All these cases proved to be benign schwannoma in histopathology and none had evidence of recurrence during the minimum 12-month follow-up.

## Case Report

### *Case 1*

A 39-year-old woman was referred with a retroperitoneal pelvic tumor on a CT scan with right abdominal pain. CT showed a 45 × 40 × 33 mm well-defined cystic tumor along the right obturator nerve (Figure 1-A). After laparoscopic dissection of the ureter and iliac vessels, we found that the tumor originated from the obturator nerve (Figure 1-B). The tumor capsule was incised and tumor contents were enucleated until the tumor was completely resected. Afterward, we observed remnant nerve fascicles in the hollow of the tumor (Figure 1-C). Postoperatively, the patient had no obturator neuropathy. We observed the patient for 24 recurrence-free months.

### *Case 2*

A 49-year-old woman was referred with a left adnexal tumor on ultrasonography. CT scan showed a 45 × 50 × 34 mm well-encapsulated heterogeneous tumor in the left retroperitoneal pelvis with multiple internal low-attenuating areas, suggesting a mesenchymal tumor, such as neurogenic tumor or sarcoma (Figure 1-D). After laparotomic dissection of left retroperitoneal space, there was a 5 cm smooth-surfaced solid mass attached alongside the left obturator nerve. We performed complete excision of the tumor with some nerve fascicles (Figure 1-E, 1-F). Total hysterectomy with bilateral salpingo-oophorectomy, appendectomy, and multiple biopsies in peritoneum and omentum were simultaneously performed. Since the patient already had multiple uterine myomas with urinary frequency, the patient needed a hysterectomy. However, the

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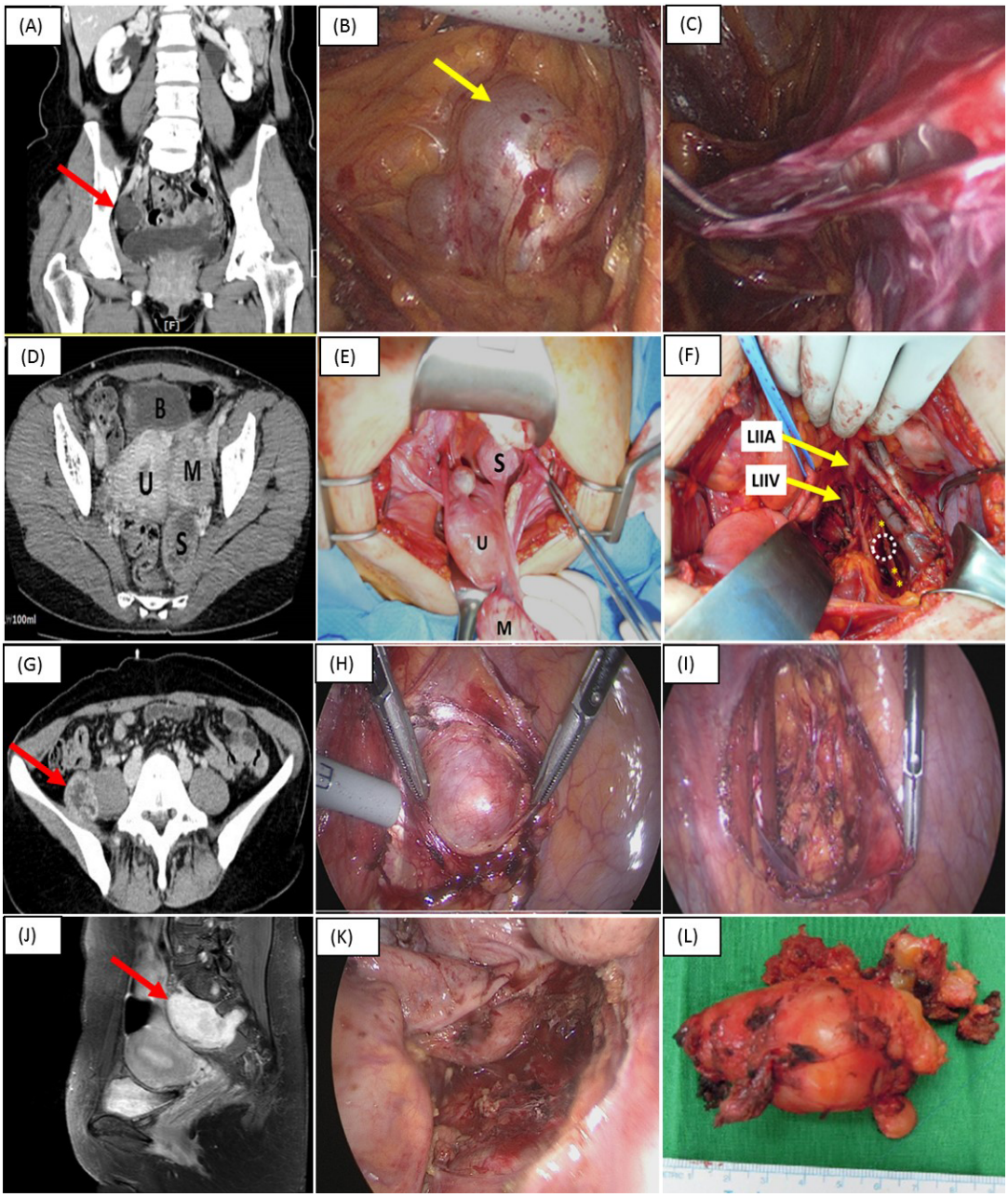


Figure 1. — A to C (Case 1). A: Preoperative CT scan, retroperitoneal mass (red arrow) in right pelvis. B: Laparoscopic view, retroperitoneal pelvic schwannoma (RPS, yellow arrow). C: Postoperative view, remnant obturator nerve fascicles after enucleation of the cystic mass. D to F (Case 2). D: Preoperative CT scan, retroperitoneal mass in left pelvis. *B = Bladder, U = Uterus, M = Uterine myoma, S = Schwannoma*. E: Intraoperative view, left RPS. F: Postoperative view, resection margin of obturator nerve (dotted ring), whole left obturator nerve (*) after complete excision of RPS. *LIIA = Left internal iliac artery, LIIV = Left internal iliac vein*. G to I (Case 3). G: Preoperative CT scan, right peripsoas mass (red arrow). H: Laparoscopic view, peripsoas RPS. I: Postoperative view, dissection of genitofemoral nerve after complete removal of peripsoas RPS. J to L (Case 4). J: T2 weighted MRI, presacral cystic solid mass (red arrow). K: Laparoscopic view, right presacral area after complete resection of presacral RPS. L: Specimen of completely excised presacral RPS.

other surgeries can be thought that it was an overtreatment in benign schwannoma. Although the possibility of malignancy was low by CT, radiologists warned of a sarcoma. Thus, we took a more radical approach. The pathologic result showed a benign schwannoma with degenerative changes. The postoperative course was uneventful. The patient completed 22 months of follow-up with no postoperative complications.

### *Case 3*

A 35-year-old woman was referred for an incidental pelvic tumor found by pre- appendectomy CT. The pa-

tient had no chronic symptoms related to the tumor. CT scan showed a 33 × 27 × 30 mm well-circumscribed tumor with peripheral hyper-enhancing wall and central geographic cystic change in the right peripsoas space (Figure 1-G). With preoperative impression as a neurogenic tumor, the patient had a laparoscopic complete excision with dissecting of common iliac vessels and ureter (Figure 1-H, 1-I). The patient reported a decreased sensation of the right medial thigh, suggesting the presumptive neurologic deficit of the genitofemoral nerve. Neural deficit resolved within a month of surgery. The patient was well at 13 months of follow-up.

*Case 4*

A 41-year-old woman was referred for a presacral tumor with an 8-month history of progressive pelvic pain. CT scan and MRI showed a 73 × 61 × 30 mm presacral tumor with the widening of the second sacral foramen (Figure 1-J). With the impression of a neurogenic tumor, our surgical team included a neurosurgeon. The neurosurgeon performed partial sacral hemilaminectomy and ligation of sciatic nerve root through the posterior midsacral approach. Afterward, the gynecologist laparoscopically explored the perirectal space to dissect the presacral tumor from rectum and sacral space. We performed complete excision of the tumor while conserving the right hypogastric nerve (Figure 1-K). The tumor presented yellowish, firm, and ovoid shape with a tense capsule (Figure 1-L). The patient's pelvic pain resolved postoperatively. However, the patient reported a decreased sensation of the right medial thigh until 2 months postoperative. Her postoperative follow-up at 12 months was recurrence-free and revealed no sensory or motor weakness of a lower extremity.

## Discussion

When discovering a retroperitoneal pelvic tumor, the absence of related symptoms supports the possibility of schwannoma. RPSs are often asymptomatic and discovered incidentally. RPSs may become symptomatic by compressing adjacent structures according to their size and location. Also, the retroperitoneum is a compliant space that allows slow-growing lesions to reach a large size. The clinical aspects, when they exist, are varied and nonspecific: abdominopelvic pain, backache, renovascular hypertension, weight loss, unexplained fever, and venous thromboses [1].

The preoperative diagnosis of RPS is quite challenging because it lacks specific symptoms and characteristic radiological findings. A review of 82 retroperitoneal schwannomas revealed that only 15.9% were identified preoperatively by imaging studies [2]. Of primary retroperitoneal tumors, 70%-80% are malignant in nature [3]. Sarcomas make up a third of all the retroperitoneal tumors with predominantly liposarcomas (70%) and leiomyosarcomas (15%) [4]. Other malignant retroperitoneal tumors include lymphomas, malignant fibrous histiocytomas, desmoid tumors, extragonadal germ cell tumors [3].

First, discrimination of RPSs from retroperitoneal malignant tumors is of importance to achieve a successful preoperative diagnosis. Recently, Zhu *et al*. studied the sensitivity and specificity of diagnostic CT indexes in the prediction of malignancy and proposed a combined score system while evaluating 194 cases of primary retroperitoneal tumors [5]. They showed that ill-defined margins, irregular tumor shape, large size, and solid texture were statistically significant indicators of malignancy. As these parameters were applied to our cases, the scores were below the threshold of malignancy (score 4), excluding the possibility of malignancies. The scores of cases 1 and 3 were 0. The score of case 2 was 1 due to solid and cystic texture. With solid-cystic proportion and large size, the score of case 4 was 2. Second, differentiation from other benign retroperitoneal tumors should be considered. The most common benign retroperitoneal tumors are schwannomas, neurofibromas, ganglioneuromas, paragangliomas, fibromatosis, and lipomas [6]. Although there are overlaps among benign retroperitoneal tumors, schwannomas are differentially diagnosed by characteristic imaging findings among benign tumors. CT scans typically show well-defined low or mixed attenuation with cystic necrotic central areas. Cystic changes occur more commonly in retroperitoneal schwannomas (up to 66%) than in other retroperitoneal tumors. Other degenerative changes, such as calcification, hemorrhage, and hyalinization, can also be present [7].

In MRIs, another imaging study to evaluate RPSs demonstrated that schwannomas appear as well-defined masses with heterogeneous low signal intensity on T1-weighted images and high signal intensity on T2-weighted images [1]. MRI may offer several advantages over CT especially in presacral tumors, as in our last case. MRI allows a better evaluation of the origin, extent, and internal composition of these lesions. Thus, MRI may help to characterize presacral schwannoma by narrowing the differential diagnosis of presacral tumors. MRI demonstrated details of intrasacral, intrapelvic, intra- or extradural, and nerve root compression, as well as displaying the relationship to neighboring structures and it helps the surgeon to determine a surgical plan [8].

Histologically, schwannomas consist of compact cellular lesions (Antoni type A) and loose, hypocellular myxoid lesions with microcystic spaces (Antoni type B). The hallmark of benign schwannoma is alternating Antoni A and B areas, with a diffuse positivity for S100 protein in the cytoplasm of the tumor cells [9].

Complete excision is the best treatment for RPSs. However, partial resection or enucleation can be performed when the mass is strongly adhered to core nerve fibers to prevent iatrogenic damage. Since the risk of recurrence is low and malignant transformation even lower, certain authors prefer enucleation with good results [2]. Sometimes, it is hard to excise RPSs with clear-cut margins. As in our case of presacral schwannoma, the tumors often erode into adjacent bony structures [10], making it difficult to dis-

Table 1. — *Literature review of studies on surgical resection of retroperitoneal pelvic schwannoma.*

| Case | Age/Sex | Presentation | Surgery | Presumptive nerve origin | Postoperative ND | Postoperative follow-up |
|---|---|---|---|---|---|---|
| Daneshmand, *Urology, 2003* [17] | 50/F | Numbness of leg | Laparoscopy CE | NA | NA | DF 38 months |
| Inoue, *J Obstet Gynaecol Res. 2004* [18] | 45/F | Hyper-menorrhea | Laparotomy almost CE | NA | Absent | DF 18 months |
| Goh BK, *Am J Surg. 2006* [10] | 50/F | Incidental palpable mass | Laparotomy CE | NA | NA | DF 12 months |
| Park, *J Laparoendosc Adv Surg Tech A. 2007* [15] | 44/F | Incidental | Laparoscopy CE | Obturator nerve | Absent | NA |
| Song, *J Obstet Gynaecol Res. 2007* [19] | 60/F | Abdominal discomfort | Laparotomy enucleation | Sacral plexus | NA | NA |
| Surendrababu, *J Clin Ultrasound. 2008* [20] | 65/F | Abdominal pain | Laparotomy CE | Lumbar nerve root | Transient | DF 6 weeks |
| Sinha, *J of Minim Invasive Gynecol. 2008* [21] | 42/F | Abdominal pain | Laparoscopy enucleation | NA | Absent | NA |
| Sinha, *J of Minim Invasive Gynecol. 2008* [21] | 50/F | Abdominal pain | Laparoscopy CE | NA | Absent | NA |
| Dawley, *J Minim Invasive Gynecol. 2008* [22] | 31/F | Chronic abdominal pain | Laparotomy CE | Femoral nerve | Transient | |
| Choudry, *World J Surg Oncol. 2009* [23] | 71/F | Back and leg pain | Laparotomy CE | NA | NA | |

*ND, neurologic defect; CE, complete excision; NA, not available; DF, disease free*

sect the tumors completely. To maximize tumor resection, nerve root transection with laminectomy is possibly needed. Thus, preoperative neurologic evaluation and active neurosurgical involvement are recommended.

Various methods of perioperative neurological monitoring have been developed, including spontaneous electromyography (spEMG) and somatosensory evoked potential (SSEP), in addition to motor-evoked potential (MEP). When intraoperative EMG confirmed that an entrapped fascicle was nonfunctional, the fascicle could be resected with the tumor. In contrast, small pieces of tumor that were tightly adhered to a functional fascicle could be left unresected. The technique provides benefits throughout the procedure by facilitating the identification of the nerve, minimizing trauma to the nerve during dissection, and allowing final confirmation of neural integrity at the end of surgery [11]. Sasaki *et al*. demonstrated that even if a nerve is not transected or injured, traction or compression of a peripheral nerve may induce ischemia, which can be monitored using MEP [12]. Although MEP alone was not able to predict postoperative transient sensory or motor deficits, the combination of MEP with other methods of neurological monitoring may improve accuracy and should be investigated in future studies [12].

Surgical extirpation of retroperitoneal tumors may be accompanied by operative difficulties due to limited access and poor visualization in a narrow pelvis. Some authors suggest that laparoscopic surgery is safe and feasible for removing benign pelvic tumors [13]. However, we caution against a laparoscopic approach for a pelvic tumor when malignancy is suspected. We could not rule out malignancy with low possibility such as retroperitoneal sarcoma, as in our case 2, an open approach is safer and more appropriate for removing mass and suspicious lesions. Also, if there is a risk of tumor rupture during laparoscopy, it is mandatory to convert to laparotomy to avoid tumor cell dissemination and partial resection [14]. Partial contraindications to laparoscopy include large tumor size and previous surgery. In our cases, the relatively small size of the tumors, their radiological appearances (well-circumscribed mass with smooth, regular margins, and central cystic degeneration that displaced rather than invaded local surrounding structures with no regional lymphadenopathy) and no previous abdominal surgeries oriented us to the laparoscopic approach.

Recurrence or malignant transformation after complete excision is very rare for RPSs [15]. However, a few cases reported metastases that occurred after resection of a histologically benign schwannoma [7]. Therefore, it is suggested that careful monitoring is necessary after the removal of RPSs.

The incidence of neurological deficits after radical operation of schwannoma is low by literature reviews (Table 1). Most schwannomas are uninodular, well encapsulated, eccentric masses, dislocating the nerve fibers that can be visualized in the periphery, flattened along with the capsule, but not penetrating the substance of the tumors [16]. Thus,

it is possible to remove RPSs while maximally preserving nerve fibers. Accordingly, most postoperative neurologic deficits are transient or minimal.

The overwhelming majority of schwannomas arose in nerves with a sensory component and were associated with sensory ganglia of the nerves. Very few schwannomas arose from pure motor nerves [24]. Generally, the injury of a sensory nerve can be accustomed or unrecognized as time passed. However, the damage of a motor nerve sometimes leaves a serious physical disability. Fortunately, these motor nerves usually had additional innervations in the pelvis. For the above reasons, even if the involved nerve is completely resected during the surgery, the neurologic complications would not be catastrophic. For example, injury of the obturator nerve, which is responsible for the sensory innervation of the medial thigh and the motor innervation of the adductor muscles of the lower limb, results in the paraesthesia and motor weakness. Patients become accustomed or gradually unaware of paraesthesia in the medial thigh within a few months. Also, since the thigh adductor muscles are primarily innervated by the femoral nerve [25], motor weakness is minor and can be rehabilitated with physiotherapy.

Injury of the sacral plexus has similar features as the obturator nerve. Postoperative lower leg numbness or pain reportedly disappears quietly. Since the sacral plexus is a network of multiple nerves in the pelvis, even if complete nerve resection occurs, postoperative motor weakness is minor.

In conclusion, RPS is rare and not easy to diagnose preoperatively. CT and MRI can narrow the wide spectrum of differential diagnoses of retroperitoneal pelvic tumors. Treatment goals should be complete excision with negative margins whenever feasible. Tumor enucleation or partial resection of schwannoma is a good countermeasure to avoid postoperative neural complications, as supported by distinctive schwannoma characteristics such as low recurrence rate, rare malignant transformation, and a slow-growing. Laparoscopic resection is a safe and feasible method for treating RPSs. However, if a tumor appears to be malignant preoperatively or intraoperatively, wide resection through the open laparotomy is recommended. To remove the tumor safely, a multidisciplinary team approach is needed. At the time of this report, patients appear in good health with no evidence of recurrence in the short term. Longer follow-up may be needed to assess the true recurrence of these tumors.

## Ethics approval and consent to participate

All subjects gave their informed consent for inclusion before they participated in the study. The study was conducted in accordance with the Declaration of Helsinki, and the protocol was approved by the Ethics Committee of Ulsan University Hospital (approval number: UUH 2019-10-045).

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## Conflict of Interest

The authors declare no conflict of interest.

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## References

[1] Ningshu L., Min Y., Xieqiao Y., Yuanqing Y., Xiaoqiang M., Rubing L.: "Laparoscopic management of obturator nerve schwannomas: experiences with 6 cases and review of the literature". *Surg. Laparosc. Endosc. Percutan. Tech.,* 2012, *22*, 143.

[2] Cury J., Coelho R.F., Srougi M.: "Retroperitoneal schwannoma: case series and literature review". *Clinics (Sao Paulo),* 2007, *62*, 359.

[3] Hoarau N., Slim K., Da Ines D.: "CT and MR imaging of retroperitoneal schwannoma". *Diagn. Interv. Imaging.,* 2013, *94*, 1133.

[4] Li Q., Gao C., Juzi J.T., Hao X.: "Analysis of 82 cases of retroperitoneal schwannoma". *ANZ. J. Surg.,* 2007, *77*, 237.

[5] Osman S., Lehnert B.E., Elojeimy S., Cruite I., Mannelli L., Bhargava P*., et al.*: "A comprehensive review of the retroperitoneal anatomy, neoplasms, and pattern of disease spread". *Curr. Probl. Diagn. Radiol.,* 2013, *42*, 191.

[6] Clark M.A., Fisher C., Judson I., Thomas J.M.: "Soft-tissue sarcomas in adults". *N. Engl. J. Med.,* 2005, *353*, 701.

[7] Zhu Z., Zhao X., Zhao Y., Yang L., Zhao J., Dai J*., et al.*: "Evaluation of CT findings for the differentiation of benign from malignant primary retroperitoneal tumors". *Chin. Med. J. (Engl.),* 2014, *127*, 114.

[8] Van Roggen J.F., Hogendoorn P.C.: "Soft tissue tumours of the retroperitoneum". *Sarcoma,* 2000, *4*, 17.

[9] Chandhanayingyong C., Asavamongkolkul A., Lektrakul N., Muangsomboon S.: "The management of sacral schwannoma: report of four cases and review of literature". *Sarcoma.,* 2008, *2008*, 845132.

[10] Goh B.K., Tan Y.M., Chung Y.F., Chow P.K., Ooi L.L., Wong W.K.: "Retroperitoneal schwannoma". *Am. J. Surg.,* 2006, *192*, 14.

[11] Khatib R.A., Khalil A.M., Saba M.I., Aswad N.K., Mroueh A.M.: "A pelvic retroperitoneal Schwannoma presenting as an adnexal mass". *Gynecol. Oncol.,* 1994, *53*, 242.

[12] Lee D.Y., Chi J.Y., Seok J., Han S., Lee M.H., Jeong W.J*., et al.*: "Feasibility of brachial plexus schwannoma enucleation with intraoperative neuromonitoring". *Clin. Exp. Otorhinolaryngol.,* 2020, *13*, 203.

[13] Sasaki H., Nagano S., Yokouchi M., Setoguchi T., Shimada H., Yamamoto T*., et al.*: "Utility of intraoperative monitoring with motor-evoked potential during the surgical enucleation of peripheral nerve schwannoma". *Oncol. Lett.,* 2018, *15*, 9327.

[14] Di Furia M., Salvatorelli A., Della Penna A., Vicentini V., Sista F., Chiominto A*., et al.*: "Advantage of laparoscopic resection for pelvic Schwannoma: Case report and review of the literature". *Int. J. Surg. Case. Rep.,* 2018, *45*, 38.

[15] Park N.Y., Chong G.O., Lee Y.S.: "Laparoscopic resection of schwannoma in the anomaly of obturator nerve". *J. Laparoendosc. Adv. Surg. Tech. A.,* 2007, *17*, 769.

[16] Hamada Y., Iwaki T., Fukui M., Tateishi J.: "A comparative study of embedded nerve tissue in six NF2-associated schwannomas and 17 nonassociated NF2 schwannomas". *Surg. Neurol.,* 1997, *48*, 395.

[17] Daneshmand S., Youssefzadeh D., Chamie K., Boswell W., Wu N., Stein J.P., Boyd S., Skinner D.G.: "Benign retroperitoneal schwannoma: A case series and review of the literature". *Urology,* 2003, *62*, 993-997

[18] Inoue T., Kato H., Yoshikawa K., Adachi T., Etoh K., Wake N.: "Retroperitoneal schwannoma bearing at the right vaginal wall". *J. Obstet. Gynaecol. Res.,* 2004, *30*, 454-457.

[19] Song J.Y., Kim S.Y., Park E.G., Kim C.J., Kim D.G., Lee H.K., Park I.Y. "Schwannoma in the retroperitoneum". *J. Obstet. Gynaecol. Res.,* 2007, *33*, 371-375.
[20] Surendrababu N.R., Cherian S.R., Janakiraman R., Walter N.: "Large retroperitoneal schwannoma mimicking a cystic ovarian mass in a patient with hansen's disease". *J. Clin. Ultrasound,* 2008, *36*, 318-320.
[21] Sinha R., Sundaram M., Hegde A., Mahajan C.: "Pelvic schwannoma masquerading as broad ligament myoma". *J. Minim. Invasive. Gynecol.,* 2008, *15*, 217-219.
[22] Dawley B.: "A retroperitoneal femoral nerve schwannoma as a cause of chronic pelvic pain". *J. Minim. Invasive. Gynecol.,* 2008, *15*, 491-493.
[23] Choudry H.A., Nikfarjam M., Liang J.J., Kimchi E.T., Conter R., Gusani N.J., Staveley-O'Carroll K.F.: "Diagnosis and management of retroperitoneal ancient schwannomas". *World J. Surg. Oncol.,* 2009, *7*, 12.
[24] Tryggvason G., Barnett A., Kim J., Soken H., Maley J., Hansen M.R.: "Radiographic association of schwannomas with sensory ganglia". *Otol. Neurotol.,* 2012, *33*, 1276.
[25] Ramage J.L., Varacallo M.: "Anatomy, Bony Pelvis and Lower Limb, Medial Thigh Muscles". Treasure Island: StatPearls Publishing; 2018.

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