

A Rare Case of Obturator Nerve Schwannoma Resembling Ovarian Malignancy: Learning from our Lesson

Suwanit Therasakvichaya, M.D.*, Nida Jareemit, M.D.*, Suchanan Harnamornroongruang, M.D.** , Nalin Leelatian, M.D., Ph.D.***,****, *****

*Department of Obstetrics and Gynecology, **Department of Pathology, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok 10700, Thailand, ***Department of Cell and Developmental Biology, Vanderbilt University, Nashville, TN, 37232, USA, ****Vanderbilt-Ingram Cancer Center, Vanderbilt University, Nashville, TN, 37232, USA, *****Department of Pathology, Microbiology and Immunology, Vanderbilt University, Nashville, TN, 37232, USA.

ABSTRACT

Objective: Obturator nerve schwannoma is an uncommon pelvic tumor and, therefore, is usually overlooked as a probable diagnosis per initial, preoperative, radiologic evaluations. This leads to an unanticipated difficult intraoperative management in a narrow operative field, surrounded by vital organs. This report describes a patient with obturator nerve schwannoma who presented with a pelvic mass and underwent surgical evaluation for possible ovarian malignancy.

Case presentation: A 58-year-old woman, presenting with asymptomatic right solid-cystic pelvic mass. Exploratory laparotomy, which was performed due to suspicion of ovarian cancer, revealed a 9-cm retroperitoneal mass located within the right obturator fossa, originating from the obturator nerve. Hysterectomy, bilateral salpingo-oophorectomy and enucleation of retroperitoneal mass were performed. The right obturator nerve was entirely preserved. Histopathological examination revealed a schwannoma. Postoperatively, the patient reported paresthesia at the medial aspect of the right thigh without weakness. No recurrence was detected at 6-month follow up.

Conclusion: Although obturator nerve schwannoma is rare and often not diagnosed preoperatively, a thorough inspection of CT scan and MRI can potentially reveal a continuity with its anatomical origin. Subsequently, a needle biopsy can be performed to obtain a diagnosis. Laparoscopy is a safe therapeutic approach for schwannomas located within the obturator fossa.

Keywords: Obturator nerve; ovarian tumor; pelvic mass; schwannoma (Siriraj Med J 2018;70: 455-458)

INTRODUCTION

Schwannoma is a rare peripheral nerve sheath tumor, composed of well-differentiated Schwann cells.¹ Schwannomas most commonly arise from cranial or main peripheral nerves, especially within the head and neck region.¹ Pelvic retroperitoneal schwannoma is even rarer, comprising only 0.5-3% of all schwannomas.² Due to their infrequent incidence, pre-operative diagnosis of

pelvic retroperitoneal schwannomas is uncommon. In this circumstance, surgeon may face with intraoperative challenges in this narrow operative field which include adjacent major blood vessels and the preservation of nerve function. This study presents an unanticipated intraoperative event and management of obturator nerve schwannoma in a woman who underwent exploratory laparotomy for presumed ovarian cancer.

Correspondence to: Nida Jareemit

E-mail: nida.jareemit@gmail.com

Received 30 May 2018 Revised 21 September 2018 Accepted 24 September 2018

doi:10.14456/smj.2018.72

CASE PRESENTATION

A 58-year-old healthy postmenopausal woman, Para 2, presented with an asymptomatic pelvic mass, which was detected during an annual check-up visit. Her previous pelvic examination performed 6 years ago was normal. Pelvic examination revealed a normal uterine size and an 8-cm, firm and fixed mass located at the right pelvic region. No other abnormalities were detected. Whole abdominal computed tomography (CT) scan with contrast revealed an 8.2x9.0x8.5-cm mixed solid and cystic mass located at the right ovary, suspicious of ovarian cancer (Fig 1). No metastasis was shown. Chest X-ray was normal. CA125, CA19-9 and CEA value were 12.1 U/mL (normal <35 U/mL), <0.6 U/mL (normal <40 U/mL), and 1.6 ug/L (normal <4 ug/L), respectively. Concerned about having ovarian cancer, the patient underwent exploratory laparotomy for surgical staging. There was no evidence of pelvic organ abnormality except for a 9-cm retroperitoneal mass located at right obturator fossa and extended to presacral area. The mass was attached laterally to right obturator nerve. Hysterectomy and bilateral salpingo-oophorectomy were initially performed since gynecologic malignancy could not be excluded. The mass was completely enucleated. Only the capsule remained unresected.

The right obturator nerve was entirely preserved. During the procedure, the adjacent venous plexus was injured, leading to a massive bleeding (8,500 mL). Packed red blood cells 9 units, fresh frozen plasma 2,000 mL, platelet concentration 12 units and cryoprecipitate 12 units were transfused intraoperatively.

The gross examination of retroperitoneal mass revealed a 9x8x6-cm soft, lobulated, heterogeneous light brown and yellow tissue. There was an area of cystic degeneration measured 3x3 cm, with hemorrhagic tissue lining (Fig 2). Microscopically, there was a combination of two distinct growth patterns, Antoni A and Antoni B (Fig 3). Additionally, vascular hyalinization was observed, representing a degenerative change. The tumor exhibited immunoreactivity with S100 antibody, but not marked with CD34, SMA, desmin or CD117. Ki67 labeling index was less than 5%, indicating low proliferation (Fig 3). The histopathological findings and the anatomical location of the tumor supported the diagnosis of obturator nerve schwannoma. The patient was discharged on day 4. Besides having paresthesia at the medial aspect of the right thigh without evidence of weakness, she had an unremarkable recovery. At 6-month follow up, paresthesia persisted despite receiving rehabilitation, but no tumor recurrence was detected.

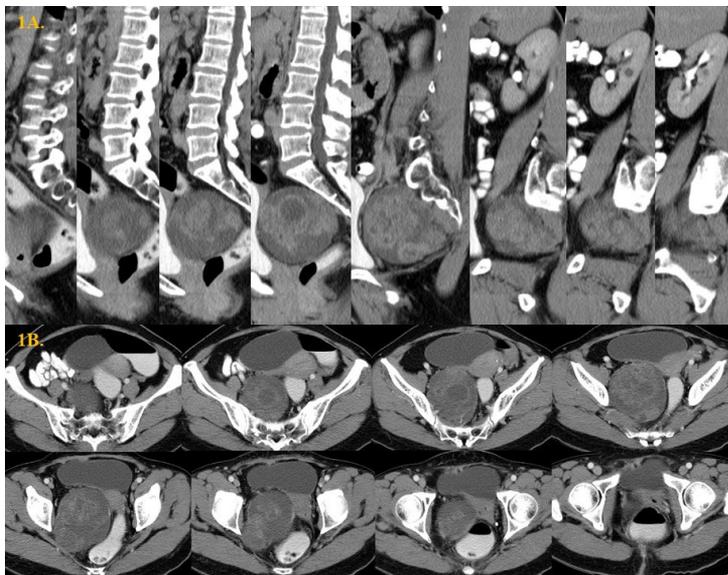


Fig 1. CT whole abdomen with contrast revealed a well-circumscribed heterogeneous solid-cystic mass at right pelvis (1A) sagittal view (1B) axial view



Fig 2. Gross findings of retroperitoneal mass

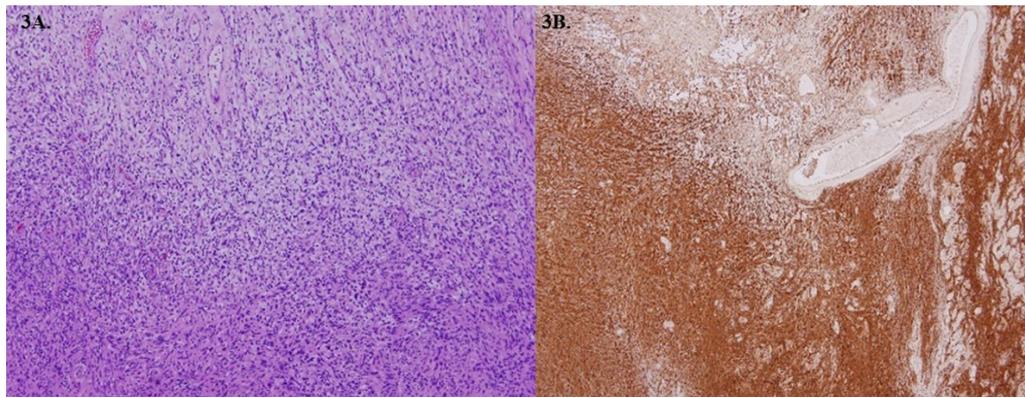


Fig 3. Microscopic appearance (3A) mixture of two growth pattern; Antoni A and Antoni B with area of vascular hyalinization (3B) diffuse reactivity for S-100 protein

DISCUSSION

Schwannomas are generally benign nerve sheath tumors. Patients diagnosed with schwannomas usually present with solitary and slow-growing masses that are otherwise asymptomatic. Compressive symptoms can occur due to tumor size. Neurological symptoms including pain and paresthesia have been reported in up to one third of the cases, particularly in large or deep-seated tumors.³

Preoperative diagnosis of schwannomas is extremely challenging due to their non-specific imaging characteristics. A previous study showed that only 15.9% (13/82) of retroperitoneal schwannomas were accurately diagnosed preoperatively using either ultrasonography, CT scan, or MRI⁴. Specifically for obturator nerve schwannomas, successful preoperative diagnosis is extremely rare, and to our knowledge, only one case has been reported to date.⁵ Some studies proposed that presence of certain imaging features could raise the suspicion of nerve sheath tumors.⁶ These findings included round and sharply demarcated solid tumors, which might exhibit cystic regions as signs of degenerative changes or, rarely, malignant transformation. MRI provides a better tissue analysis than CT and might be the superior imaging modality for the preoperative evaluation of schwannomas.⁷ Schwannomas can appear as low signal intensity on T1-weighted images and high signal intensity on T2-weighted images.⁶ Either central (target-like) or peripheral enhancement can be found.⁶ However, the most reliable finding was presence of connecting between tumor and the originating nerve, in this case, the obturator nerve.⁵

Prior studies evidenced an advantage of needle biopsy. Strauss et al., showed that histopathological diagnosis of schwannoma were successfully obtained in 19 out of 28 patients who underwent needle biopsy.⁸ This was further confirmed by the study of Ferretti et al.,

who showed that needle biopsy or fine needle aspiration can differentiate between benign and malignant tumor.⁹

For the presented case, since the CT scan suggested that the pelvic mass originated from the ovary, MRI was not performed for further evaluation. Because preoperative diagnosis is critical for the selection of surgical approaches and presurgical mapping, in situations where the diagnosis of a pelvic mass is uncertain, a combination of multiple imaging modalities with a needle biopsy might be necessary.

Surgical management of obturator nerve schwannoma presents a great challenge. These difficulties include a limited operative field in the obturator fossa, presence of adjacent structures such as the obturator nerve, the iliac vessels, and the venous plexus. Damages to obturator nerve can result in a transient or permanent weakness of hip adduction and outward rotation, as well as a sensory loss over the medial thigh. There are two major types of surgical management of obturator nerve schwannoma: enucleation or tumor resection.⁵ Enucleation is the preferable method since it minimizes nerve damage while sufficiently removes the tumor.¹⁰ In contrast, tumor resection is usually performed in rare cases that are suspicious for malignancy, such as patients with neurofibromatosis or Recklinghausen's disease, as well as tumors containing cystic changes.⁵ Critically, if malignancy is suspected, tumor margins should be obtained and evaluated to prevent local recurrence.^{5,11} Moreover, enucleation is usually performed on large schwannomas (>5 cm), whereas tumor resection should be performed on small tumors attached to one side of nerve.^{5,6,10} Laparoscopic surgery is the recommended procedure since 1) its magnifying effect enhances the visualization of the structures surrounding the tumor, and 2) it is advantageous to conventional methods especially in small areas with limited access.^{5,10} A recent study also showed that the use of robotic EndoWrist® and

3-dimensional viewing can successfully remove pelvic schwannomas while preserving neurological functions.¹

In this case, enucleation was performed because of large tumor size. Massive bleeding caused by venous plexus injury was subsequently stopped by ligation. A number of strategies have been proposed to solve this life-threatening condition are packing, tacking, topical hemostatic agents, electrocoagulation and suture.¹² To prevent pelvic hemorrhage, preoperative embolization, internal iliac artery ligation or harmonic scalpel use are suggested.^{1,10}

Recurrence is uncommon in benign schwannoma following a complete resection. In contrast, local recurrence was found in 20% and 72% of malignant schwannomas that underwent complete and incomplete resections, respectively, which emphasizes the importance of total excision in malignant schwannomas.¹

CONCLUSION

Surgical management of obturator nerve schwannoma would be a great challenge avoiding injury to adjacent vital organs. Obtaining preoperative diagnosis using imaging modalities combined with needle biopsy can aid the selection of operative procedure and surgical planning. Although rare, schwannomas should always be considered a plausible diagnosis in patients who present with pelvic mass of uncertain origin since these tumors can be removed using a safe laparoscopic approach.

Conflict of interest declaration

All authors declare no personal or professional conflicts of interest, and no financial support from the companies that produce and/or distribute the drugs, devices, or materials described in this report.

Funding disclosure

This was an unfunded study.

REFERENCES

1. Perrin H, Brunner P, Ortega JC, Mercier B, Clement N, Robino C, et al. Robotic resection of an obturator schwannoma with preservation of normal nerve fascicles and function. *J Robot Surg.* 2017;11:479-83.
2. Borghese M, Corigliano N, Gabriele R, Antoniozzi A, Izzo L, Barbaro M, et al. Benign schwannoma of the pelvic retroperitoneum. Report of a case and review of the literature. *G Chir.* 2000;21:232-8.
3. Rodriguez-Peralto JL, Riveiro-Falkenbach E, Carrillo R. Benign cutaneous neural tumors. *Semin Diagn Pathol.* 2013;30:45-57.
4. Li Q, Gao C, Juzi JT, Hao X. Analysis of 82 cases of retroperitoneal schwannoma. *ANZ J Surg.* 2007;77:237-40.
5. Takahashi H, Hara M, Tsuboi K, Sagawa H, Ishiguro H, Matsuo Y, et al. Laparoscopically resected obturator nerve schwannoma: A case report. *Asian J Endosc Surg.* 2016;9:307-10.
6. Gleason T, Le BH, Parthasarathy K, Robinson-Bennett B. Obturator Nerve Schwannoma as a Mimic of Ovarian Malignancy. *Case Rep Obstet Gynecol.* 2017;2017:9724827.
7. Liu HL, Yu SY, Li GK, Wei WI. Extracranial head and neck Schwannomas: a study of the nerve of origin. *Eur Arch Otorhinolaryngol.* 2011;268:1343-7.
8. Strauss DC, Qureshi YA, Hayes AJ, Thomas JM. Management of benign retroperitoneal schwannomas: a single-center experience. *Am J Surg.* 2011;202:194-8.
9. Ferretti M, Gusella PM, Mancini AM, Mancini L, Vecchi A. Progressive approach to the cytologic diagnosis of retroperitoneal spindle cell tumors. *Acta Cytol.* 1997;41:450-60.
10. 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;22:143-7.
11. Park NY, Chong GO, Lee YS. Laparoscopic resection of schwannoma in the anomaly of obturator nerve. *J Laparoendosc Adv Surg Tech A.* 2007;17:769-73.
12. Celentano V, Ausobsky JR, Vowden P. Surgical management of presacral bleeding. *Ann R Coll Surg Engl.* 2014;96:261-5.