

## Technical Notes &amp; Surgical Techniques

## Retroperitoneal schwannoma associated with neuropathic pain and electromyographic and nerve conduction abnormalities: A case report



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## ABSTRACT

**Background:** The retroperitoneal space is an uncommon location for schwannomas. A review of the existing literature revealed that only 0.3–3.2% of these tumors are found in this location. Presenting symptoms, if any, are usually vague and include abdominal discomfort. Neuropathic pain as a presenting symptom is rare. We present the first known report of retroperitoneal schwannoma associated with both neuropathic pain and positive EMG/NCS findings.

**Case description:** A 34-year-old woman presented initially with a lump underneath her previous cesarean section incision. She was taken to surgery for the suspected intra-abdominal mass. During surgery she was found to have a retroperitoneal mass associated with a large nerve root, so surgery was aborted and the neurosurgery team was then involved. Pre-operative evaluation revealed neuropathic pain in the distribution of the femoral nerve as well as EMG/NCS findings of femoral neuropathy and denervation. She was thus taken to surgery for resection. Pathology confirmed the diagnosis of retroperitoneal schwannoma.

**Conclusions:** Retroperitoneal schwannomas are usually incidental findings. If there are presenting symptoms they are usually vague abdominal complaints. While there is one report of neuropathic pain associated with retroperitoneal schwannoma, there are no previous reports of neuropathic pain with associated EMG/NCS findings.

## 1. Introduction

Schwannomas are benign tumors arising from the cells that form the peripheral nerve sheath. They are typically well encapsulated and grow slowly. They are usually found associated with spinal or cranial nerves, and only rarely are found in other sites such as the retroperitoneum; retroperitoneal or pelvic examples represent only 0.3–3.2% of these tumors [1,2]. Clinically, retroperitoneal schwannomas usually present with vague abdominal pain, constipation, or as an incidental finding on imaging studies [3–7]. Consequently they are frequently discovered incidentally by gynecologists, urologists, or general surgeons, and most reports of such tumors are in journals of urology, gynecology, general surgery, or radiology.

A review of the literature found two reports of retroperitoneal

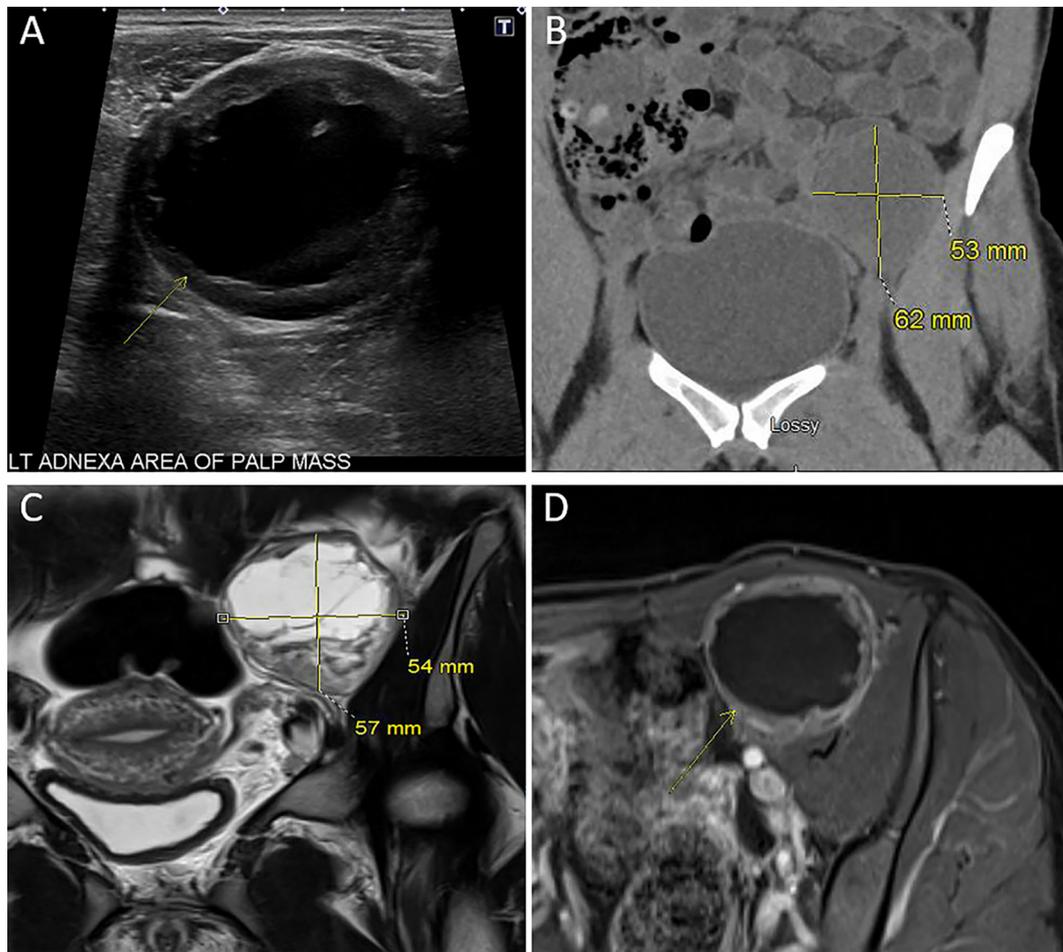
schwannomas associated with neuropathic pain or weakness, only one of which contained electromyography (EMG) and nerve conduction studies (NCS) which were negative. Here we describe a patient found to have a retroperitoneal schwannoma who presented with femoral neuropathic pain and in whom there were associated EMG/NCS findings.

## 2. Case description

A 34-year-old woman with a history of cesarean section 3 years prior to presentation noted a painful “lump” at her left lower quadrant beneath the cesarean incision. Pain radiated to her left thigh when palpated. She was initially referred to several surgeons, and diagnoses of hernia or endometrioma were considered. However, abdominal ultrasound and a computed tomography (CT) scan of her abdomen

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**Fig. 1.** Pre-operative imaging.

A. Transcutaneous pelvic ultrasound of the left adnexal area demonstrates a  $5.8 \times 4.4 \times 5.0$  cm peripherally isoechoic and centrally hypoechoic mass abutting the anterior abdominal wall (arrow).

B. Coronal computed tomography (CT) scan soft tissue of the abdomen without contrast demonstrating a  $6.2 \times 5.3$  cm hypodense mass (right to left dimensions). The mass lies superficial to the psoas muscle and there is mass effect on the adjacent adnexa.

C. Coronal T2 weighted MRI of the pelvis demonstrates a  $5.4 \times 5.7$  cm mass in the left anterior aspect of the pelvis. There is central hyperintensity suggestive of cystic necrosis.

D. Axial T1 weighted MRI of the pelvis with gadolinium infusion demonstrates a peripherally enhancing mass located between the psoas muscle and the anterior abdominal wall (arrow).

(Fig. 1A–B) revealed a left adnexal mass for which the differential diagnosis was considered to be enteric duplication cyst, pseudocyst, lymphatic malformation, or a lymphocele. Thus she was referred to a gynecological surgeon who attempted a laparoscopic resection.

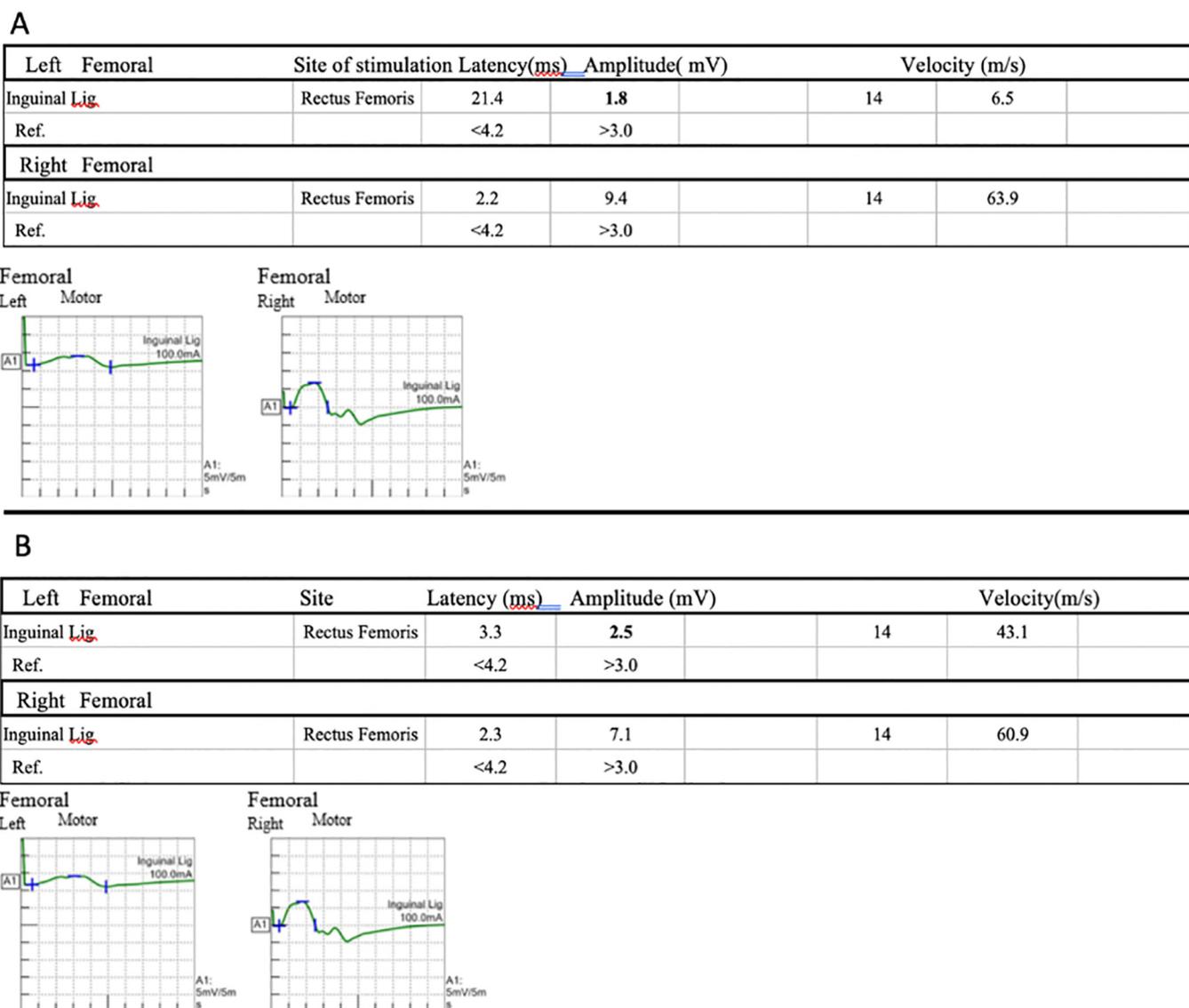
During the operation the uterus and adnexa were found to be normal; the mass was situated superficial to the left psoas muscle. Upon opening the retroperitoneum the mass was found to involve a large nerve, thus neurosurgery was consulted intra-operatively. A biopsy was deferred due to the perceived risk of spillage of cyst contents into the abdominal cavity, so the procedure was terminated and she was referred to the neurosurgery clinic for further management.

At her initial neurosurgery clinic evaluation she described 2 years of paresthesias in the left hip and anterior thigh with occasional electric shock-like pain. She was found to have decreased sensation of her left anterior thigh, 4+/5 weakness of knee extension, and an absent left patellar reflex (right patellar reflex 2+). The rest of her examination was symmetric and normal. A magnetic resonance image (MRI) (Fig. 1C–D) showed a  $4.5 \times 5.4 \times 5.6$  cm well-circumscribed centrally cystic left lower quadrant mass which had irregular peripheral enhancement after gadolinium administration. A PET scan showed no evidence of metastatic disease. An EMG/NCS examination showed

moderate to severe femoral neuropathy with evidence of ongoing denervation (Fig. 2A). A subsequent needle biopsy revealed a spindle cell neoplasm consistent with schwannoma. Due to her pain, weakness, and the results of the needle biopsy, surgical resection was recommended.

An open laparotomy for resection was performed utilizing intra-operative EMG monitoring (Fig. 3A–B). The mass was exposed in the retroperitoneum and the capsule was stimulated to find a safe entry zone. The tumor was dissected off of the nerve and removed in two large fragments. The last remaining portion of tumor was adherent to a nerve branch; stimulation of the nerve as well as other surrounding nerve branches demonstrated innervation of the vastus medialis and lateralis. The branch entering the tumor was sacrificed and the remaining tumor was removed. Intra-operative EMG remained stable throughout surgery.

The patient awoke with 2/5 weakness in knee extension, hypoesthesia in the anterior thigh, and continued areflexia of the left patella. She was discharged 3 days later with outpatient physical therapy. At last follow-up 18 months post-operatively she was walking with a knee brace, and her left anterior thigh pain had resolved apart from some paresthesias controlled with gabapentin. Her weakness had improved to 4/5 in knee extension, and her sensation was progressively



**Fig. 2.** Pre and post operative nerve conduction studies.  
 A. Pre-operative bilateral femoral motor nerve conduction studies with recording from rectus femoris. The amplitude of the left femoral nerve is 1.8 mV which is significantly diminished compared to the right side.  
 B. Post-operative bilateral femoral motor nerve conduction studies with recording from the rectus femoris. The amplitude of the left femoral nerve has improved to 2.5 mV which is evidence of reinnervation.

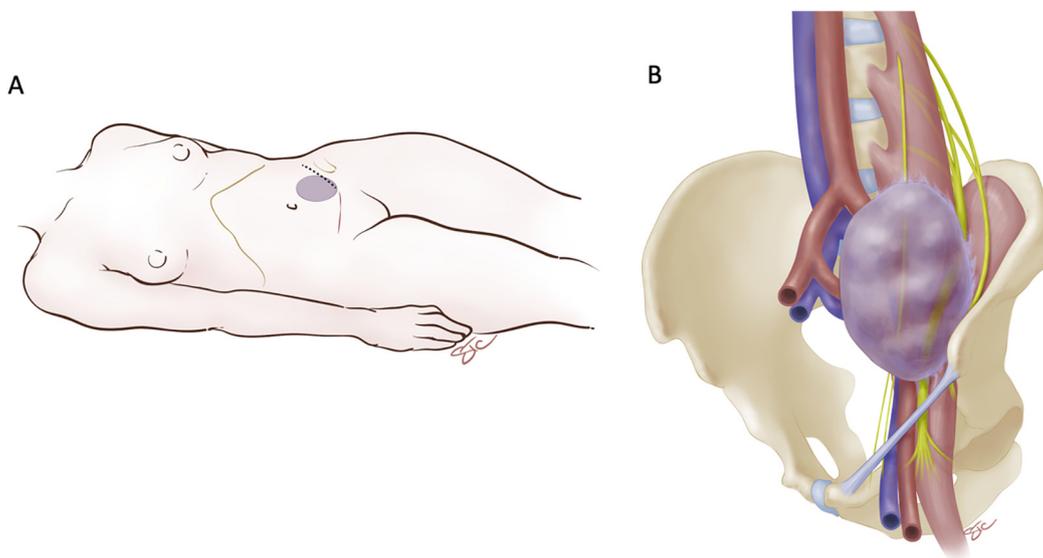
improving. An EMG/NCS at this follow-up showed improved amplitude of the femoral nerve action potential but with continued chronic denervation (Fig. 2B). The follow up MRI showed no evidence of residual or recurrent tumor (Fig. 4A–B).

The excised tumor was histopathologically confirmed to be a schwannoma (Fig. 5). It was submitted as two specimens, one with two pieces, 5.5 × 4.5 × 2.5 cm and 3.0 × 2.0 × 1.5 cm, respectively, and the other 6.5 × 3.0 × 2.5 cm, and was quite large. There was indeed a central cyst with a dark brown lining (Fig. 5A). The tumor was focally highly cellular but otherwise was histologically an ordinary example of a Schwannoma (Fig. 5B–C); a Ki67 immunostain revealed a maximal proliferation index of 5.2%.

### 3. Discussion

The tumor found in our patient was large, and indeed from the literature the non-specific symptoms which are most often what brought the patients to attention and the ultimate diagnosis of a retroperitoneal schwannoma likely contribute to the large size of these

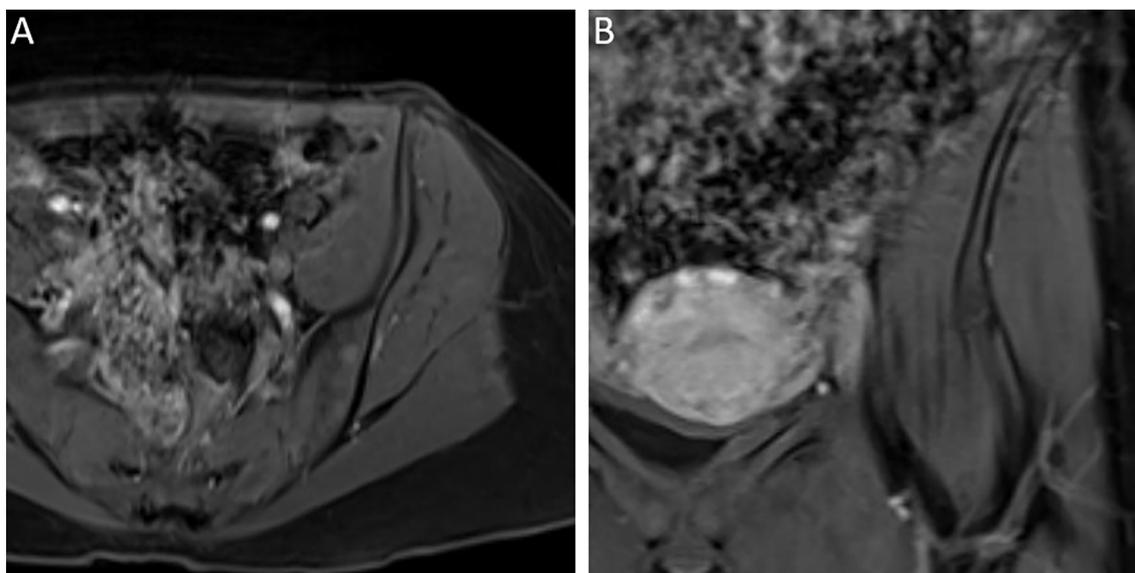
tumors as they go undiagnosed for significant periods of time [9]. It is not surprising that retroperitoneal schwannomas usually present without neuropathic pain because there are no rigid anatomic constraints in the retroperitoneum against which a nerve can be compressed. That situation is in contrast to similar tumors on spinal nerve roots which grow among bony structures. As stated previously only two reports were found of retroperitoneal schwannoma with associated neuropathic pain, and only one reported EMG/NCS test results which did not reveal any abnormality. This makes the patient we report here all the more unusual because of her neuropathic symptoms and the associated EMG/NCS findings which matched them. It is unclear why our patient presented with these symptoms as opposed to the usual symptoms presented above. We speculate that our patient presented with these symptoms possibly due to her BMI of 18, thus she did not have much physical space for such a large mass. The tumor could have been wedged between the iliopectineal arch and inguinal ligament thus compressing the nerve. It is also possible that scar tissue from her previous cesarean delivery could have caused the tumor to adhere to surrounding structures.



**Fig. 3.** Surgical approach.

A. Illustration showing the surgical positioning of the patient. The dotted lines represent the incision used, and the shaded area is the approximate location of the tumor relative to the incision.

B. Illustration showing the position of the tumor located superficial to the psoas muscle within the pelvic cavity. Also seen are the iliac vessels which were adherent medially and had to be dissected free. The position of the femoral nerve relative to the tumor can also be seen.



**Fig. 4.** Post-operative imaging.

A/B. Axial (A) and Coronal (B) T1 weighted MRI of the pelvis with gadolinium infusion demonstrates complete resection of the mass with no evidence of recurrent tumor.

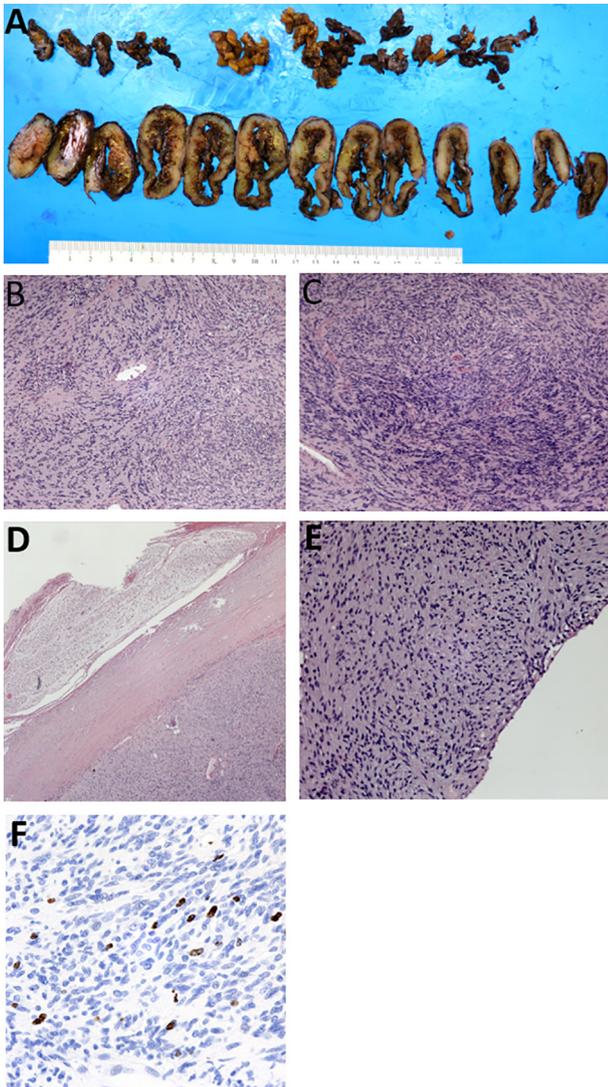
Although our patient had symptoms of neuropathic pain along with an abdominal mass a diagnosis of nerve sheath tumor initially was not entertained. Had that been considered she may not have had to undergo two operations along with the associated risks in order to achieve definitive treatment. It is likely that the rarity of retroperitoneal schwannoma lead to the lack of suspicion for this diagnosis. Furthermore, the neuropathic pain in this case may have been considered a separate issue entirely because retroperitoneal schwannomas rarely present with those symptoms. Hopefully this case will raise awareness of the possibility of retroperitoneal schwannoma with similar cases in the future.

#### 4. Conclusions

Schwannomas located in the retroperitoneum are uncommon. Neuropathic pain as a presenting symptom was only found to be reported in two cases, and there were no reports of positive EMG/NCS findings. We have described an unusual case in which this rare location for a schwannoma is coupled to a unique presentation: neuropathic pain along with abnormal findings in the involved nerves by EMG/NCS. An abdominal mass along with neuropathic pain should lead to a higher index of suspicion for nerve sheath tumor.

#### Declaration of Competing Interest

The authors declare that they have no known competing financial



**Fig. 5.** Pathological specimen.

A. Gross specimen of tumor demonstrating the central cyst.

B. Histological appearance of the tumor. The bulk of the tumor had the Antoni A pattern with solid tissue with an eosinophilic background populated by spindle cells arranged in fascicles with complex intersecting patterns, and with parallel nuclear palisades (Verocay Bodies). No mitotic figures were apparent. H&E, original magnification 100 $\times$ .

C. Highest cell density of the tumor. Focally there was greater cellularity, still without apparent mitotic figures. H&E, original magnification 100 $\times$ .

D. At one edge of the tumor there was a fascicle of atrophic uninjured peripheral nerve, adherent to the tumor's capsule. H&E, original magnification 40 $\times$ .

E. The lining of the cyst wall in the tumor had no epithelioid differentiation. H&E, original magnification 200 $\times$ .

F. A Ki67 immunostain labels 5.2% of the tumor nuclei in this most labeled focus (labeling index determined by computer-assisted morphometry). Original magnification 400 $\times$ .

interests or personal relationships that could have appeared to

influence the work reported in this paper.

## References

- [1] R. Tong, N. Collier, A. Kaye, Chronic sciatica secondary to retroperitoneal pelvic schwannoma, *J. Clin. Neurosci.* 10 (1) (2003) 108–111.
- [2] G. Sharifi, Persistent L5 lumbosacral radiculopathy caused by lumbosacral trunk schwannoma, *Asian J. Neurosurg.* 12 (1) (2017) 51–54.
- [3] N. Machairiotis, P. Zarogoulidis, A. Stylianaki, et al., Pelvic schwannoma in the right parametrium, *Int. J. Gen. Med.* 6 (2013) 123–126.
- [4] L. Fenoglio, S. Severini, P. Cena, et al., Common bile duct schwannoma: a case report and review of literature, *World J. Gastroenterol.* 13 (2007) 1275–1278.
- [5] M. Mehta, W.A. Thurston, N. Merchant, K.J. Murphy, Obturator nerve schwannoma presenting as an adnexal mass: case report, *Can. Assoc. Radiol. J.* 50 (1) (1999) 20–22.
- [6] B. Dawley, A retroperitoneal femoral nerve schwannoma as a cause of chronic pelvic pain, *J. Minim. Invasive Gynecol.* 15 (4) (2008) 491–493.
- [7] B. Duran, T. Guvenal, E. Yildiz, M. Cetin, O. Erden, N. Demirkoprulu, An unusual cause of adnexal mass: fallopian tube schwannoma, *Gynecol. Oncol.* 92 (1) (2004) 343–346.
- [9] K. Rathod, V. Kate, B. Badhe, Cystic retroperitoneal swelling occupying the whole abdomen, *Gastroenterology* 141 (4) (2011) e7–e8.