Giant Pelvic Schwannoma: A Case Report

Dev Pelvik Schwannoma

Ahmet Hakan HALİLOĞLU, MD,[a]
Hatim Yahya USLU, MD,[b]
Nuket ÜZÜM, MD,[c]
Nuray HALİLOĞLU, MD,[d]
Orhan GÖĞÜŞ, MD,[a]

Departments of [a]Urology
[b]General Surgery,
Ufuk University Faculty of Medicine, Ankara
[c]Mikropat Pathology Laboratory, İstanbul
[d]Department of Radiology
Ankara University Faculty of Medicine, Ankara

Geliş Tarihi/*Received:* 13.06.2008
Kabul Tarihi/*Accepted:* 11.09.2008

Yazışma Adresi/*Correspondence:*
Ahmet Hakan HALİLOĞLU, MD,
Ufuk University Faculty of Medicine,
Department of Urology, Ankara,
TÜRKİYE/TURKEY
ahmethakan75@yahoo.com

ABSTRACT Schwannomas are benign peripheral nerve sheath tumors. Pelvic schwannomas which are usually seen in young adults are quite rare tumors and usually present as large masses causing vague symptoms. When asymptomatic huge pelvic masses are detected, it may not always be possible to differentiate benign and malignant lesions without histopathological examination. These tumors do not have specific clinical, laboratory, and radiological findings. Therefore preoperative diagnosis is usually not adequate and complete resection of the mass might be essential. Herein this report, we aimed to describe the clinical, radiological, and pathological findings of a giant pelvic schwannoma detected in a 53-year-old man.

Key Words: Neurilemmoma; nerve sheath neoplasms

ÖZET Schwannomalar iyi huylu periferal sinir kılıfı tümörleridir. Sıklıkla genç erişkinlerde görülen pelvik yerleşimli schwannomalar oldukça seyrek görülen tümörlerdir ve genellikle belirsiz semptomlara neden olan geniş boyutlu kitleler olarak karşımıza çıkarlar. Herhangi bir semptoma neden olmayan büyük boyutlu pelvik kitleler saptandığında benign ve malign kitle lezyonları arasında ayırıcı tanı yapmak her zaman mümkün olmayabilir. Bu tümörlere ait spesifik klinik, laboratuar ve radyolojik bulgu yoktur. Bu nedenle preoperatif tanı çoğu zaman yeterli değildir ve kitlenin tümüyle çıkarılması ve histopatolojik inceleme mutlak gerekli olabilir. Bu yazıda 53 yaşında bir erkek hastada saptanan dev bir pelvik schwannomanın klinik, radyolojik ve patolojik bulgularının tanımlanması amaçlanmıştır.

Anahtar Kelimeler: Nörilemmoma, sinir kılıfı tümörleri

Turkiye Klinikleri J Med Sci 2010;30(2):750-4

Schwannomas are infrequent peripheral nerve sheath tumors which account for 5% of benign soft tissue tumors. More than 50% of the lesions are seen in the head and neck region and extremities.[1] Pelvic schwannomas are quite rare tumors, compromising less than 1% of benign schwannomas.[2,3] When seen in the pelvic region they may present with symptoms like abdominal pain, hypertension, and frequent urination. Nevertheless the symptoms are usually vague and nonspesific hence there is frequently a delay in the detection of the tumor. It should be kept in mind that although rare, schwannomas can be malignant and as accurate preoperative diagnosis is difficult, and the mass has to be removed totally by dissecting from the neighbouring structures. We present here a histopathologically proven huge perivesical schwannoma with a brief review of the literature.

CASE REPORT

A 53-year old man was admitted to the urology department with left flank pain and gross hematuria with a history of 3-months. There was no significant finding in his personal or family medical history. A huge hard mass below the umblikus was detected on physical examination. Cystograpy revealed displacement of the bladder to the right side but there was no filling defect in the cavity and the bladder margins were smooth (Figure 1). There was no significant finding on neurological examination. Suprapubic ultrasonography (US) revealed a hypoechoic solid mass located posterolaterally to the left wall of the bladder with dimensions of 18 x 13 x 8 cm. There was no obvious relation between the soft tissue mass and the prostate gland on transrectal sonography. On computed tomography (CT), the well demarcated hypodense mass with minimal contrast enhancement was neighbouring the small intestine mesenteric fat superiorly and running down to the ischioanal fossa inferiorly. No calcifications were detected on CT images. The mass was displacing the bladder anterolateraly and rectum laterally. Magnetic resonance imaging (MRI) was performed in order to evaluate the exact nature of the lesion and to determine its relation to the neighbouring structures. MR images revealed a heterogenous mass with solid and cystic components located in the presacral region which was lying downwards to the left ischioanal fossa with dimensions of 24 x 14 x 11 cm. On contrast enhanced T1-weighted images the central part of the mass was enhancing predominantly and signal void areas representing vessels were present on T2-weighted images (Figures 2, 3). However MRI also failed to show the exact origin of the mass but it revealed that there was no invasion to the surrounding tissues.

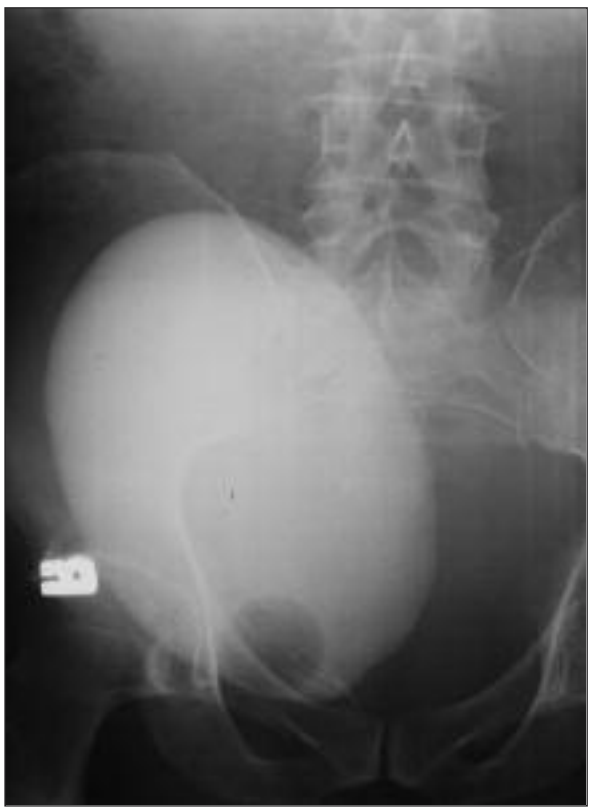


FIGURE 1: On cystographic examination the bladder seems to be normal except for lateral displacement reminding compression of a neighbouring mass.

As the preoperative diagnosis was not accurate, surgical exploration was considered to be essential. Previously, a cystoscopy was performed in order not to miss an occult intravesical lesion which may cause hematuria. The high-positioned bladder was hardly enrolled during the cystoscopic examination performed by 21 F rigid cystoscop. No intravesical lesions were detected and the ureteral orifices were normal.

During the exploration, the mass seemed to occupy the whole left perivesival region and extended towards to the bladder neck and vesicula seminalis but as it was not adhesive to the neighbouring structures and it could easily be removed with no significant bleeding at all. Since as the distal part of the left ureter was running so close to the mass, it was damaged during the exploration and ureterouretereostomy was performed. There was no need for blood transfusion during or after the surgery and no complications were noted in the postoperative period.

On the histopathological examination, macroscopically, the total specimen was well-circumscribed, encapsulated, with gelatinous appearance and gray in color, measured 30 cm by 23 cm and 20 cm. Microscopically, the tumor was exhibited a fascicular and from place to place whorled growth pattern with thin, loose and intact fibrous capsule. Hypercellular (Antony A component) and hypocellular (Antony B) components were seen in low-

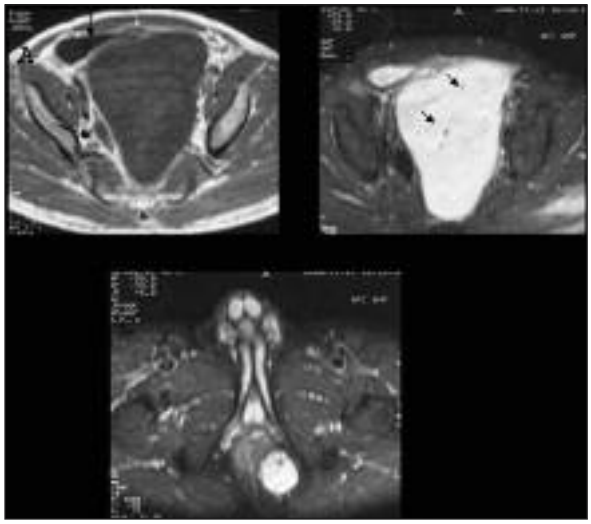


FIGURE 2: T1-weighted axial MR image (A) reveals a well-defined huge pelvic mass displacing the bladder anterolateraly (short arrow) and rectum laterally (long arrow). The central part of the mass is mildly hyperintense when compared to the muscle. On fat saturated T2-weighted axial image (B) there are signal void areas representing vasculary elements (arrows). The mass is continuing down to the ischioanal fossa (C).

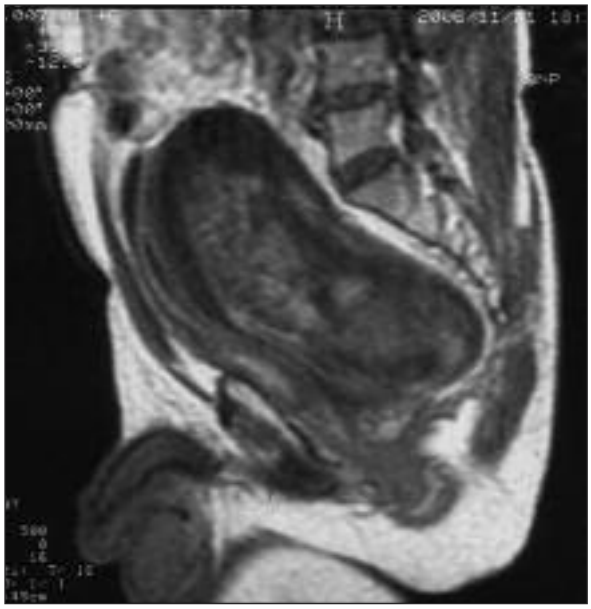


FIGURE 3: On contrast enhanced T1-weighted sagittal MR image the central part of the presacral mass enhances predominantly whereas the upper parts seems to be cystic.

power field. The cells were similar with elongated cell shape and wavy nuclei, in both areas. There were nuclear palisading areas in tumor. Prominent thick hyaline vessel walls and widespread myxoid areas were present within the tumor. Mitoses were extremely scanty (Figure 4). There were no cytological atypia or necrosis. The immunohistochemical study revealed a diffuse positive staining in both cytoplasms and nuclei of the cells with S-100 (Figure 5).

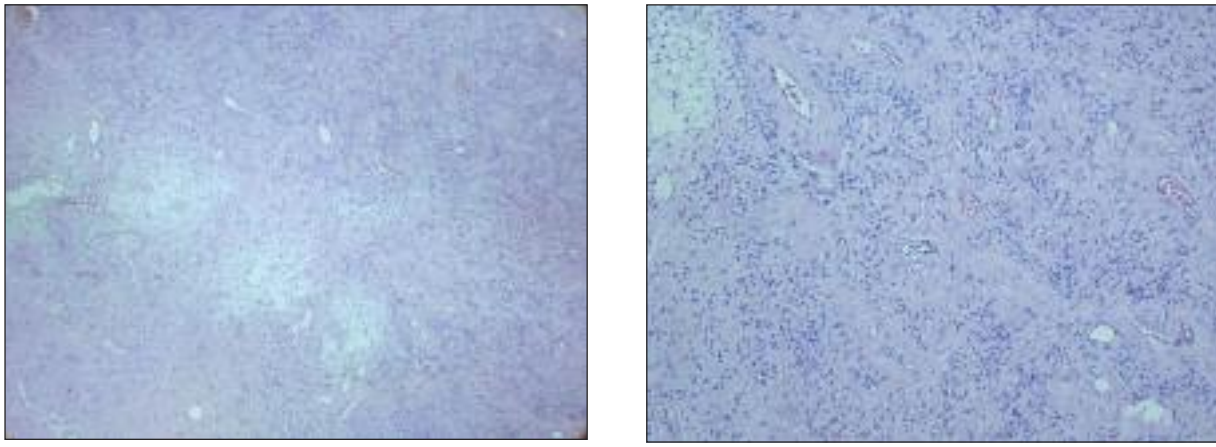
The patient was discharged on the 5th day of the surgery. Pelvic CT was performed in 6 months time and there were no pathological findings. He is still under follow-up.

DISCUSSION

Schwannomas originate from the Schwann cells of the neural sheath, and especially when multiple lesions are present, they can be associated with neurofibromatosis type I.[4,5] The tumor may occur in any nerve trunk except for the cranial nerves I and II which lack schwann cells. Pelvic schwannomas mostly arise from the lumbar and sacral nerves.[6,7] In the present case, the presacral tumor was multilobulated and it was running down to the ischioanal fossa, but there was no pedicle of the tumor attaching it to the lumbar or sacral nerve roots suggesting a neurogenic origin.

Schwannomas slow-growing tumors but as they do not cause symptoms unless they reach a large size enough to compress adjacent structures, and as retroperitoneum is a flexible location allowing the tumor grow-up easily, pelvic schwannomas may present as huge masses with cystic degeneration at the initial diagnosis.[4,8]

It has been estimated that pelvic schwannomas are freqently located in the lower retroperitoneum, and they are usually ovoid or spherical in shape with smooth, well-defined borders. As the size of the mass increases, the amount of cystic degeneration remains.[4] Although there is no imaging characteristic finding unique to this tumor, presence the cystic degeneration in a semisolid tumor on US may suggest the diagnosis, as retroperitoneal tumors are usually not cystic. On CT scans, schwannomas appear as heterogenous masses, and if present punctate or mottled calcifications can easily be detected by CT.[9] MRI is the most accurate imaging method in the diagnosis of soft-tissue tumors with its high contrast resolution and multiplanar capability. Schwannomas are usually hypointense or isointense with muscle on T1- weighted images, and hyperin-



a b

FIGURE 4: Antoni type A (cellular areas) and antoni type B (hypocellular areas) tissue. (HE x 40) (A). The myxoid matrix and hyaline vessel walls in tumor (HE x 100) (B).

tense on T2-weighted images. Nevertheless Heterogenity due to cystic and hemorrhagic components can also be detected on MR images. Solid components would enhance after intravenous contrast medium administration.[4] MRI is also capable of showing the relationship of the tumor with adjacent anatomic structures and delineating the vascular nature of the tumor.[10]

CT-guided biopsy is usually not recommended since cellular pleomorphism may lead to misinterpretion as malignancy. On the other hand, if the mass is malignant, tumor seeding and the other well known risks of percutaneous biopsy like hemorrhage or infection may occur.[7,9]

The ideal choice of treatment is complete excision with preservation of the associated nerve. As malignant schwannomas are aggressive neoplasms acting as high-grade sarcomas and the risk of malignancy can not be excluded preoperatively or by frozen section analysis, neighbouring structures can be sacrificied in order to attain negative tumor margins. In the case we presented here, the left ureter was running so close to the mass that it was damaged during exploration. Oppositely, one can consider partial resection or simple enucleation relying on the common benign nature of the tumor and also considering the age, general health, and symptoms of the patient.[9,10]

On histopathological examination two patterns exist for schwannomas, Antoni A and B types representing high and low cellularity, respectively.

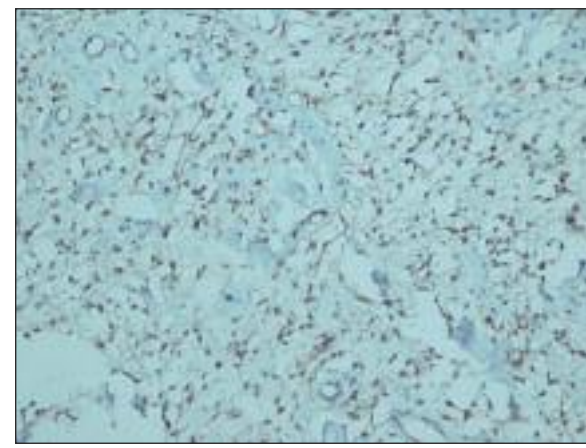


FIGURE 5: S-100 immunoreactivity in schwann cells (Streptavidin-biotin peroxidase x 200)

Antoni A areas are composed of spindle-shaped cells and Antoni B areas are composed of edematous and degenerated tissues with fewer cells. Diffuse staining of S-100 protein is helpfull to confirm the diagnosis of schwannoma.[6,8] The present tumor consisted of both Antoni A and Antoni B cells and immunohistochemical study revealed a diffuse positive staining of the cells with S-100 protein. There was no sign of malignant transformation.

In conclusion; there is no spesific imaging finding for benign pelvic schwannomas, and although rare, malignant transformation may occur. Therefore, complete surgical resection should be the choice of treatment depending on the patient's general status, and the relationship of the tumor with the adjacent vital structures.

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