

Atypical presentations of retroperitoneal giant schwannomas

Sait Ozbir, Mehmet Cengiz Girgin, Cengiz Kara, Cetin Dincel

Department of 1st Urology, Izmir Ataturk Training and Research Hospital, Izmir, Turkey

Abstract

Schwannomas are usually benign rare tumors that originating from Schwann cells of peripheral nerve sheaths. Presentation is generally varied and changed in a non-specific range from abdominal mass, flank pain to incidental findings. Herein we report 2 cases of retroperitoneal giant schwannomas with different clinical presentations, of whom one presented with vague abdominal pain, palpable abdominal mass for 4 years, swelling and bilateral hydronephrosis that caused by giant abdominal mass; the other one presented with right flank pain, rectal hemorrhage and lower extremities edema. Two patients were treated by complete surgical excision of masses. The histological and immunohistochemical diagnosis was reported as benign schwannoma. Both of patients are doing well and had no recurrence in 9 years and 28 months follow-up, respectively.

Introduction

Schwannomas are rare and usually benign tumors originating from Schwann cells of peripheral nerve sheaths.¹⁻⁵ Most schwannomas arise from cranial or upper extremities nerves.¹⁻⁴ The schwannomas rarely occur in the retroperitoneum, about 0.5% to 5% of all schwannomas.²⁻⁶ Tumor size does not usually exceed a diameter of 5 cm. Involvement of the urinary tract is rare, but involvement of the penis, testes, spermatic cord, and tunica vaginalis has been previously reported.^{1,2,5} Most common presentation of the retroperitoneal schwannomas is vague abdominal or dull ache pain. The atypical presentations and giant retroperitoneal schwannomas are very rare.^{2,4} Herein we report 2 cases of benign retroperitoneal giant schwannomas with different clinical presentations and their subsequent management, of whom one presented with vague abdominal pain, palpable abdominal mass for 4 years, swelling and bilateral hydronephrosis that caused by giant abdominal mass; one presented with right flank pain, rectal hemorrhage and lower extremities edema.

Case Reports

Case #1

A 73-year-old male patient presented with swelling, vague abdominal pain and abdominal mass. He had been evaluated by another clinic with the same symptom 4 years before; he was admitted to our clinic. On physical examination, he had a palpable mass extending from epigastrium to symphysis pubis. His blood count and blood chemistry test results were normal. The patient's transrectal and abdominal ultrasonography (USG) showed a 18×13×11 cm enhancing solid mass locating between prostate and urinary bladder, extending to the umbilical level. Computed tomography (CT) scan of the abdomen showed a 20×18×13 cm enhancing capsulated solid mass with cystic and necrotic areas, arising from retroperitoneum (Figure 1A). He had also bilateral ureterohydronephrosis although renal function tests were normal (Figure 2). The urinary bladder displaced anterolaterally between the mass and anterior abdominal wall owing to compression of the mass. The patient was initially diagnosed as having a retroperitoneal tumor and underwent a mass excision with anterior laparotomy. A Retroperitoneal mass with capsulated distinct from the peripheral tissues without tissue invasion, was established. Not only both ureters were dilated but also the left one showed marked elongation. Ten centimeters of the left ureter were resected, anastomosed end-to-end; and double-J stents were placed in both ureters for 3 months. Macroscopic examination revealed a capsulated mass of 21×18×11 cm, with cystic and solid bright-yellow areas. Outer surface was smooth and no sign of invasion. On microscopic examination, cellular and hypocellular areas were prominent. Cellular area revealed spindle cells with oval, angulated nuclei and paled cytoplasm without pleomorphism. The cells were diffusely and strongly positive for S-100 protein. The tumor was diagnosed as schwannoma. At the 9th year follow-up, the patient remained asymptomatic and there was no evidence of recurrence. CT of the abdomen revealed bilateral hydronephrosis was decreased degree and also renal function tests results were normal.

Case #2

A 46-year-old female patient presented with 2-year history of flank pain, one episode of rectal hemorrhage and increasingly edema at lower extremities. On physical examination she had a palpable mass specially filling the right abdomen, extending from epigastrium to iliac crest. She had both moderate bilateral pitting edema on lower extremities without evidence of chronic venous stasis and hemorrhoids. The patient's blood count and blood

Correspondence: Sait Ozbir, Izmir Ataturk Egitim ve Arastirma Hastanesi, 1st Urology Clinic, 35000, Yesilyurt, Izmir, Turkey.
Tel: +90.5052841297 - Fax: + 90.2322444445.
E-mail: saitozbir@hotmail.com

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Figure 1. (A) Computed tomography scan showing a large pelvic mass compressing the bladder and ureters. (B) Computed tomography scan showing a large solid mass, compressing the vena cava inferior.



Figure 2. Computed tomography scan showing bilateral ureterohydronephrosis.

chemistry levels were in normal range. USG of the abdomen revealed a 17×14×10 cm enhancing heterogeneous solid mass filling the right hemi-abdomen, extending from right kidney lower pole to pelvis. CT scan of the abdomen showed a 17×15×11 cm enhancing solid mass with cystic and necrotic areas, just arising from lower level of porta hepatis on the right-side of abdomen, extending to iliac crest and compressing the right kidney from anterior and the vena cava inferior (Figure 1B). Some organs was pushed counter side of the abdomen. Below the abdomen, cleavage between the right psoas muscle and mass was not identified. Colonoscopy revealed no other remarkable pathology except from hemorrhoids. The patient was initially diagnosed as having a retroperitoneal tumor and underwent a mass excision with anterior laparotomy. The mass was dissected from retroperitoneum but was tightly adherent to the right ureter. The right kidney, right ureter and mass were excised separately. The postoperative period was uneventful. Macroscopic examination revealed a capsulated mass of 20×17×11 cm with cystic and solid bright-yellow areas and had a 7×0.5 cm structure with lumen on it. The kidney was unremarkable, and the left ureter was adhered but not invaded by the retroperitoneal mass. Microscopic examination revealed focal areas of Antoni types A and B tissue. The cells were immunoreactive for S-100 protein. The tumor was diagnosed as a schwannoma. The patient is doing well in the last 28 months of follow-up. There was no evidence of recurrence or deterioration of renal function.

Discussion

Schwannoma is nerve sheath tumor that usually affect the head, neck and the extremities. Localization of retroperitoneum comprises only 0.5% to 12% of all retroperitoneal tumors.⁵ Tumor may occur in all ages, but most commonly is seen between 40 and 60 years of age and dominantly in female (Male/female ratio 2:3).^{5,7} Large schwannomas are very rare, only 0.3-5% of all cases and most of them are located in the retroperitoneum.^{2,8,9} Although tumor size usually under the diameter of 5-6 cm, some cases was reported which diameter was reached 26 cm.¹⁰ Diagnosis of the retroperitoneal schwannomas is usually difficult, and a mass is usually presented before without any symptoms. Generally the symptoms are vague and nonspecific, such as vague abdominal or dull ache pain. Atypical presentations of retroperitoneal schwannomas are very rare.^{3,5} These atypical presentations may be variable, such as headache, flank pain, hematuria, sec-

ondary hypertension, recurrent renal colic pain and deep venous thrombosis.^{4,5,11} In our cases also were presented atypical symptoms and giant retroperitoneal masses with largest tumor diameter were 21 and 20 cm, respectively. Macroscopically, most of schwannomas are solitary, capsulated, well circumscribed, firm, smooth-surfaced tumor which is gray in color. Sometimes secondary changes may associate with this tumor including hemorrhage, cysts and calcification.^{2,5} Histopathologically, elongated bipolar spindle cells arranged in hypercellular (Antoni A) and hypocellular (Antoni B) areas, nuclei arranged in a palisading pattern. A mitotic activity is uncommon. The degenerative changes may be found; such as hemorrhage, cyst formation, hyalinized walls of vessels.^{5,7,8} Immunohistologically; strongly and diffusely staining of S-100 protein in schwannomas cells is required for the exact diagnosis.^{2,10}

Imaging studies are necessary in order to diagnostic evaluation. USG can discriminate solid tumors from cystic masses without exact diagnosis. CT scans typically show well-defined low or mixed attenuation with cystic necrotic central areas. Cystic changes are more common in retroperitoneal schwannomas than in other retroperitoneal tumors. Takatera et al. reported that 63% of the benign and 75% of the malignant schwannomas were found to be cystic.¹² Also other degenerative changes may be present, such as hemorrhage, calcification and hyalinization. But, these changes are not specific and not conclusive enough to define the origin of schwannoma.⁵ Likewise, CTs of our patients revealed cystic and necrotic areas and one of them had bilateral hydronephrosis without deterioration of renal function.

It is recommended that the management of retroperitoneal schwannomas is complete surgical excision with negative soft tissue margins. In order to obtain this aim; may be sacrificed adjacent tissue and viscera, if necessary. But some authors suggested that simple enucleation or partial excision is sufficient because of the benign nature of the masses and the morbidity associated with resection of adjacent tissue may be more important in prognosis. In first case, the mass isolated from adjacent structures, and resected completely with negative tissue margins. In the second case, the mass was tightly adherent to the right ureter, and we performed the resection of right kidney, right ureter and mass because of the great mass diameter and could not exclude the malignancy. Prognosis of retroperitoneal benign schwannomas is extremely good and recurrence is very rare. In case of recurrences surgical resection is recommended. Adjuvant treatment is not recommended. Both of patients are doing well and had no recurrence

or abnormal renal function test results in 9 years and 28 months follow-up, respectively. Although the malignant transformations have been reported, they are usually associated with von Recklinghausen's disease.^{2,5} We believe that schwannomas should be kept in mind in giant retroperitoneal masses. Although these tumors are so large, our cases indicates that the prognosis is favorable.

References

1. Micali S, Virgili G, Vespasiani G, et al. Benign schwannoma surrounding and obstructing the ureteropelvic junction. *Eur Urol* 1997;32:121-3.
2. Girgin C, Ozkan U, Sezer A, Tugyan N. Large pelvic schwannoma causing bilateral hydronephrosis. *Int J Urol* 2003;10:616-8.
3. Daneshmand S, Youssefzadeh D, Chamie K, et al. Benign retroperitoneal schwannoma: a case series and review of literature. *Urology* 2003;62:993-7.
4. Singh V, Kapoor R. Atypical presentation of benign retroperitoneal schwannoma: report of three cases with review of literature. *Int Urol Nephrol* 2005;37:547-9.
5. Cury J, Coelho RF, Srougi M. Retroperitoneal schwannoma: case series and literature review. *Clinics (Sao Paulo)* 2007;62:359-62.
6. Borghese M, Corigliano N, Gabriele R, et al. [Benign schwannoma of the pelvic retroperitoneum. Report of a case and review of the literature]. *G Chir* 2000;21:232-8.
7. Ohigashi T, Nonaka S, Nakanoma T, et al. Laparoscopic treatment of retroperitoneal benign schwannoma. *Int J Urol* 1999;6:100-3.
8. Sakalauskaite M, Stanaitis J, Cepkus S, et al. Retroperitoneal giant schwannoma eroding lumbar vertebra: a case report with a literature review. *Cent Eur J Med* 2008;3:233-44.
9. Li Q, Gao C, Juzi JT, Hao X. Analysis of 82 cases of retroperitoneal schwannoma. *ANZ J Surg* 2007;77:237-40.
10. Kuznetsov D, Peabody T, Posner MC, et al. The giant of the giant schwannomas. *J Urol* 1999;162:791-2.
11. Alshehri FM, Hussain M, Aljuaid E, et al. Retroperitoneal schwannoma presenting as deep venous thrombosis. *Eur J Radiol Extra* 2011;77:e59-e62.
12. Takatera H, Takiuchi H, Namiki M, et al. Retroperitoneal schwannoma. *Urology* 1986;28:529-31.