Case Report/Olgu Sunumu

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Benign Retroperitoneal Schwannoma: A Case Report

Benign Retroperitoneal Schwannom: Olgu Sunumu

Elif Usturalı KESKİN[1], Ömer KORAŞ[2], Tufan YILMAZ[1]

[1] Mustafakemalpasa State Hospital, Pathology Department, Bursa, TURKEY

[2] Mustafakemalpasa State Hospital, Urology Department, Bursa, TURKEY

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ÖZET

Schwannom schwan hücrelerinden köken alan iyi huylu yavaş büyüyen bir tümördür. Retroperitoneal yerleşim oldukça nadir olup vakaların yaklaşık %0.5-5'inde izlenir. Biz preoperatif olarak diğer malign tümörlerden ayrılamayan retroperitoneal schwannom olgusunu sunduk.

55 yaşında kadın hasta künt karın ağrısı şikayeti ile kliniğe başvurdu. Fizik muayenede hafif batın hassasiyeti ve hipertansiyon mevcuttu. Ultrasonografide minimal serbest sıvı ve presakral bölgede kitle izlendi. MR T1 ağrlıklı görüntülemede hipointens, T2 ağırlıklı görüntülemede heterojen hiperintens 64x58x48mm boyutlarında lezyon görüldü. Makroskopik incelemede tümör 8x6x5cm boyutlarında iyi sınırlı ve kapsüllü kitle görünümündeydi. Histopatolojik ve immünohistokimyasal bulgularla benign schwannom tanısı verildi.

Retroperitoneal schwannom tedavisi tam cerrahi rezeksiyondur. Benign schwannomların prognozu iyidir ve en sık komplikasyon muhtemel tam olmayan rezeksiyona bağlı rekürrenstir.

Anahtar Kelimeler: Benign, retroperitoneal, schwannom

ABSTRACT

Schwannoma is a benign slow growing tumour arising from Schwann cells. The retroperitoneal location is very rare with approximately 0.5-5% of all cases. We report a case of retroperitoneal schwannoma which was unable to be preoperatively differantiated from malign tumours.

A 55-year-old woman presented with a complaint of dull abdominal pain. On physical examination there was mild abdominal tenderness and hypertension. Ultrasound abdomen showed minimal amount of free fluid with a mass at the presacral region. MR showed 64x58x48mm hypo intense lesion in T1 weighted images and heterogeneous hyper intense on T2 weighted images. On gross examination the tumour was 8x6x5 cm, with a well-defined encapsulated mass. The diagnosis of benign Schwannoma was made with histopathological and immunohistochemical findings.

The treatment of retroperitoneal schwannoma is complete surgical resection. The prognosis of benign schwannomas is good and the most frequent complication is recurrence possibly due to incomplete excision.

Key Words: Benign, retroperitoneal, schwannoma

INTRODUCTION

Schwannoma is a benign slow growing tumour arising from Schwann cells of peripheral nerves. Most of them occur in the cephalocervical region and limbs. The retroperitoneal location is very rare with approximately 0.5-5% of all cases [1].

Schwannomas are usually seen in adults in second and fifth decades of life [1,2]. They occur slightly more common in woman. It is very difficult to diagnose retroperitoneal tumours before the operation since specific clinical and radiological features are usually absent [3]. Though these tumours are mostly asymptomatic and may be found incidentally on examination or imaging [2].

Here, we present a case of retroperitoneal schwannoma which was unable to be preoperatively differentiated from malign tumours.

Case Report

A 55-year-old woman presented to urology department with a dull abdominal pain. On physical examination there was mild abdominal tenderness and hypertension. Physical examination was

***Sorumlu Yazar:** Elif Usturalı KESKİN. Mustafakemalpasa State Hospital, Pathology Department, Bursa, Turkey. E mail: drelifkeskin@gmail.com.

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unremarkable. No specific dermatological and neurological findings associated with syndromes were detected. On laboratory tests biochemistry and routine blood examinations were in normal range. Ultrasound abdomen showed minimal amount of free fluid with a mass at the presacral region. MR showed 64x58x48mm hypo intense lesion in T1 weighted images and heterogeneous hyper intense on T2 weighted images (Figure1a,b). Operation was planned. Transperitoneal transverse section was applied. The mass was in the pelvis, near the left external iliac artery, vein and ureter, behind uterus and adherent to sacrum on posterior.

On gross examination the tumour was a well-defined encapsulated mass of 8x6x5cm. Cross sections of the mass showed solid yellow with gelatinous and white fibrotic areas in peripherally (Figure 2). Histopathologically, there were specific findings for the Shwannomas; hypercellular Antoni A and Antoni B areas with a loose matrix with hypocellular macrophage and verocay bodies. (Figure 3a). Mitosis was rare. Histopathology revealed features suggestive of benign schwannoma. Immunhistochemical stain was performed and S100 was diffuse positive in outer center laboratory (Figure 3b). So the diagnosis was verified.

Postoperative recovery was uneventful. There was no neurological deficit. The patient was discharged postoperative on third day. Follow-up was recommended and patient approval was receipt for this case report.

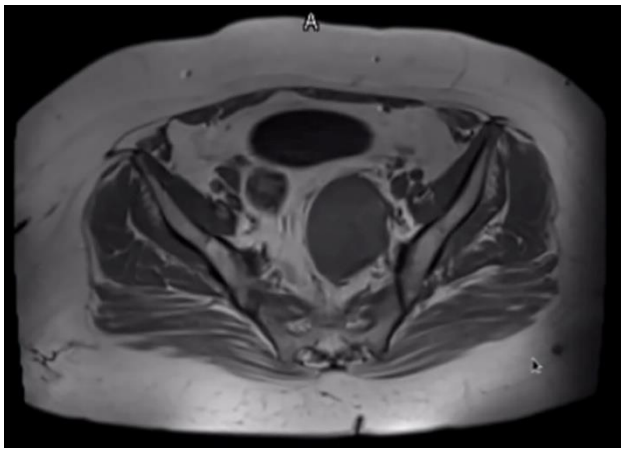

Figure 1: MR showed hypointense lesion in T1 weighted images

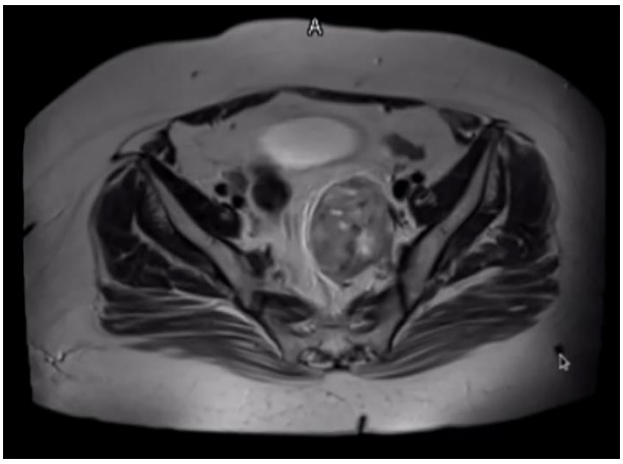

Figure 2: MR showed heterogen hyperintense lesion on T2 weighted images

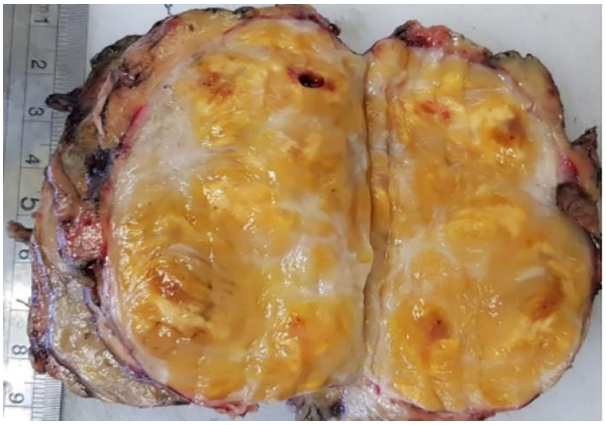

Figure 3: Lesion showed solid yellow with gelatinous and white fibrotic areas

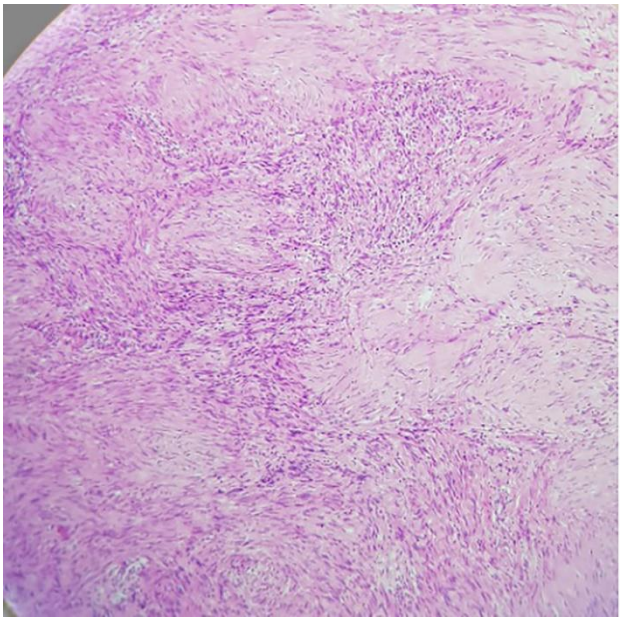

Figure 4: Histopathology showed hipocellular, loose areas (Antoni B), and hypercellular areas (Antoni A)

DISCUSSION

The retroperitoneum can host a wide spectrum of pathologies including rare benign tumours and malignant neoplasms that can be primary or metastatic [3]. Retroperitoneal schwannoma is uncommon comprising only 1-3% of all schwannoma and almost 1% of retroperitoneal neoplasms [3]. It occurs more commonly in adult females between the ages of 20-50 with a male to female ratio of 2:3 [2,4,5]. Our patient was a 55 years woman.

Among retroperitoneal schwannomas around 0.3% are benign and 3.2% are malignant [5]. Most retroperitoneal tumours are asymptomatic and are diagnosed incidentally while evaluating the patient for some other disease [4]. Our patient presented with a vague abdominal pain.

Schwannomas are seen as masses with hypointensity on T1 weighted images and hyperintensity on T2 weighted images on MR [4,6,7]. These findings are characteristic but not specific and have been reported in only 57% of the cases in previous studies [2,4,6]. Schwannomas typically appear as solitary well encapsulated masses. Our patient's MRI images showed hypo intense lesion in T1 weighted images and heterogeneous hyper intense on T2 weighted images. After operation the gross mass was firm, round with a smooth surface encapsulated. Most of schwannomas reported in literature have a diameter of 5 to 15cm [6]. In our case it was 8x6x5cm.

Since the retroperitoneal space is large and flexible the symptoms of benign retroperitoneal schwannomas are nonspecific and are usually associated with compression of adjacent structures [3,6]. It changes according to the location and size of the lesion [1] The most common symptom is abdominal distension with vague abdominal pain [6]. Other symptoms include secondary hypertension, hematuria, and renal colic [1]. In our case the mass was adherent to sacrum near left external iliac artery, vein and ureter.

It is difficult to determine a diagnosis preoperatively. A definitive diagnosis is based on pathological, histological and immunohistochemical findings [4]. In our case both malignant and benign tumours were considered. Pathologically it was a well encapsulated lesion with fibrous capsule and demonstrating specific Antoni A/B areas accompanied with degenerative changes and nuclear palisation (Verocay bodies) is often seen around fibrillar processes in cellular areas. Positive expression of S100, and negative expressions of SMA was valuable for diagnosis. Additionally calretinin, CD56, SOX10, Podoplanin, GFAP may be positive immunohistochemically [8]. Ki-67 proliferation index was low.

Schwannomas are usually benign, slow growing, encapsulated tumours. Differential diagnosis should be made histopathologically from neurofibroma, paraganglioma, phaeochromocytoma, liposarcoma,solitary circumscribed neuroma [9]. Differential diagnosis is made from these tumours with both histomorphological and immunohistochemical findings. However there can be considerable morphologic overlap between schwannoma and neurofibroma. Positivity for both calretinin and CD56 in the S100 positive cases is highly suggestive of schwannomas but CD34 appears more sensitive for neurofibromas [10]. Rarely schwannomas undergo malign transformation and invades adjacent organs. Malignancy is characterized by mitotic figures, pleomorphism, and infiltration of blood vessels. Malignancy couldn't be excluded pre operatively and so complete excision with negative margins is recommended [5]. Neither the size of the lesion nor the depth of invasion is associated with the possibility of malignancy [1]. Malignant schwannomas are frequently associated with von Reclinghausen syndrome or other types of neurofibromatosis [3]. Although there is a connection between nerve sheath tumours and neurofibromatoses this could not be confirmed in our patient.

The treatment of retroperitoneal schwannoma is complete surgical resection [3,7]. Schwannomas are not sensitive to radiotherapy and chemotherapy [3]. If the patient is not suitable for a surgical procedure because of existing comorbidities, radio-frequency ablation could be an alternative method of therapy [11]. In patients who underwent partial resection, recurrence rates have been up to 10-20% even in benign circumstances [7].

The prognosis of benign schwannomas is good and the most frequent complication is recurrence may be because of incomplete excision [3]. Even in pathologically benign and benign borders, recurrence is probable therefore careful follow-up is required.

Conflict of Interest

We have no conflict of interest.

KAYNAKLAR

Cayirli H, Tanriverdi HI, Ozguven AA, Gunsar C, Ersoy B, Kandiloglu AR. Schwannoma Localized Retroperitoneally in a 14-Year-Old Boy. Case Rep Pediatr. 2016;2016:1210874.

Kuriakose S, Vikram S, Salih S, Balasubramanian S, Mangalasseri Pareekutty N, Nayanar S. Unique surgical issues in the management of a giant retroperitoneal schwannoma and brief review of literature. Case Rep Med. 2014;2014:781347.

Fu H, Lu B. Giant retroperitoneal schwannoma: a case report. Int J Clin Exp Med. 2015 Jul 15;8(7):11598-601.

Maruyama T, Sugamoto Y, Miyagishima D, Fukunaga T, Tasaki K, Takeshita N, Tamachi T, Asai Y, Hosokawa T, Ninomiya E, Kimura M. Laparoscopic resection of a retroperitoneal schwannoma located in the hepatic hilus. Surg Case Rep. 2015 Dec;1(1):18.

Vijayan SK, Shetty S, Bhat SR, Shetty S, Khadilkar UN. Retroperitoneal schwannoma: an atypical presentation. J Clin Diagn Res. 2014 Oct;8(10):ND22-3.

Singh M, Kumar L, Chejara R, Prasad OP, Kolhe Y, Saxena A. Diagnostic dilemma of a rare, giant retroperitoneal schwannoma: a case report and review of literature. Case Rep Oncol Med. 2014;2014:628538.

Mozafar M, Lotfollahzadeh S, Atqiaee K, Adhamy F. Rare presentation of retroperitoneal schwannoma: a case report. Iran J Cancer Prev. 2014 Spring;7(2):107-10.

Correa J, Cardoso R. Immunohistochemical Markers for Schwannomas, Neurofibromas and Malignant Peripheral Nerve Sheath tumours-What Can the Recent Literature Tell Us? Arq Bras Neurocir 2018;37:105–112.

Goh BK, Tan YM, Chung YF, Chow P, Wong W. Retroperitoneal schwannoma. Am J Surg. 2006;192:14–18.

Park J, Park H, Park N, Park J, Sung H, Lee S. Use of Calretinin, CD56, and CD34 for Differential Diagnosis of Schwannoma and Neurofibroma. The Korean Journal of Pathology 2011; 45: 30-35.

Herden J, Drebber U, Ural Y, Zimmer S, Wille S, Engelmann UH. Retroperitoneal schwannomas of renal and pararenal origin: presentation of two case reports. Rare tumours. 2015 Feb 19;7(1):5616.