RETROPERITONEAL MALIGNANT SCHWANNOMA

Tevfik AKTOZ*, Mustafa KAPLAN*, Murat ÇALOĞLU**, İrfan Hüseyin ATAKAN*, Osman İNCİ*
** Department of Urology, Faculty of Medicine, Trakya University, EDİRNE, TURKEY*
*** Department of Radiation Oncology, Faculty of Medicine, Trakya University, EDİRNE, TURKEY*

ABSTRACT

Introduction: **Malignant schwannoma, recently renamed malignant peripheral nerve sheat tumors, represents 0.01 % of retroperitoneal tumors. Radical surgical excision is considered the best treatment for these neoplasms for poor responsiveness to chemotherapy and radiation therapy. We report a 66-year-old man who was operated for three times.**

Key words: **Malignant, Retroperitoneal, Schwannoma**

ÖZET

Son dönemlerde malin periferal sinir kılıfı tümörü olarak adlandırılan malin "schwannoma" retroperitoneal tümörlerin %0.01'ini oluşturmaktadır.

Altmışaltı yaşındaki erkek hasta iştahsızlık, halsizlik ve sırt ağrısı nedeniyle Nisan 2006 tarihinde kliniğimize başvurdu. Yapılan batın ultrasonografisi ve bilgisayarlı tomografi sonucu 12x11x10 cm boyutlarında, sol böbrek pelvikalisiel sistemi kompresyona uğratan, heterojen solid kitle saptandı. Kitlenin sol böbreğe çok yapışık olmasından dolayı kitlenin böbrekle birlikte çıkartılması gerekti. Patolojik inceleme sonucu tanı malin "schwannoma" olarak konuldu. Ameliyattan 11 ay sonra magnetik rezonans inceleme (MRI) sonucu 9x6,5 cm boyutlarında sağ retroperitoneal kitle saptandı. Nisan 2007 tarihinde bu kitle tamamen çıkartıldı ve patolojik olarak tekrar malin "schwannoma" olarak düşünüldü. Ağustos 2007 tarihinde tekrarlanan MRI sonucu sol retroperitoneal bölgede 15x10 cm ve 18x14 cm boyutlarında 2 adet kitle saptandı. Eylül 2007 tarihinde hasta 3. kez ameliyat edilerek kitleler çıkartıldı. Patolojik tanı bir kez daha malin "schwannoma" olarak düşünüldü. Ameliyattan 4 ay sonra yapılan MRI ile değerlendirmede L3 vertebrada metaztaz saptanması nedeniyle palyatif amaçlı radyoterapi uygulandı.

Malin periferal sinir kılıfı tümörleri ender olarak retroperitoneal bölgede oluşurlar. Genellikle asemptomatik seyrederek ileri boyutlara ulaşırlar ve komşu dokuları baskı altına alırlar. Özellikle bu olguda, lokal nükslerin sık olduğunu ve yavaş büyüyen tümörler oldukları için nükslerin geç saptanabileceğini vurgulamak istedik. Bu olgular uygun görüntüleme yöntemleri ile dikkatli takip edilmelidirler. Cerrahi olarak çıkartılamayan malin "schwannoma" olguları kemoterapi ve radyoterapiye iyi yanıt vermemekte ve seyirleri daha kötü olmaktadır. Bu nedenle bu tümörler mümkünse tamamen eksize edilmelidirler.

Anahtar kelimeler: Malin, Retroperitoneal, Schwannoma

INTRODUCTION

Schwannomas and neurofibromas are neural sheath tumors, with the former entity found in all organs and in the nerve trunk, although rarely in the retroperitoneum[1]. We report a case of retroperitoneal malignant schwannoma and discuss the clinical features briefly.

CASE REPORT

A 66 year-old-man was admitted to our hospi-tal for further examination with complaints of anorexia, lassitude and lumbal pain in April 2006. The abdominal ultrasonography and computed tomography scan revealed a heterogeneously enhanced 12x11x10 cm retroperitoneal solid mass that compresses the left renal cavities (Figure 1). A tight adhesion of the tumor to the left kidney necessitated an en bloc resection of the tumor with the left kidney. The resected solid mass was 14x12x9 cm and weighed 2240 g. Histologically, hyperchromatism of nuclei of tumor cells and high cellularity indicated this mass to be a malignant schwannoma (Figure 2). However, 11 months after the operation magnetic resonance imaging (MRI) revealed a well-circumscribed (9x6.5 cm) and homogeneous right retroperitoneal tumor (Figure 3). Complete excision of the tumor was performed in April 2007 and histological diagnosis was malignant schwannoma. Later, however, he developed a multiple, local recurrence in left retroperitoneal region. MRI scan revealed a well-circumscribed 15x10 cm and 18x14 cm tumors (Figure 4). The patient was operated on for a third time in September 2007. Histological examination revealed a malignant schwan-

Date of First Application: 07.03.2008 **Accepted**: 18.09.2008

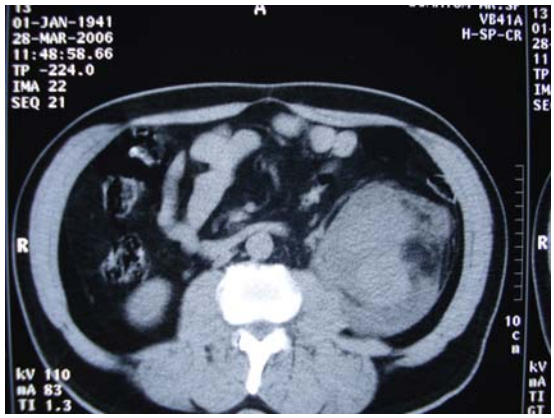

Figure 1. Computed tomography scan shows a mass arising in the left retroperitoneum before first operation

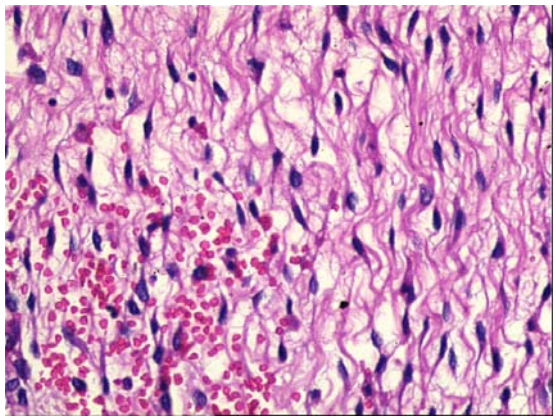

Figure 2. Hyperchromatism of nuclei of tumor cells and high cellularity (HE X 50)

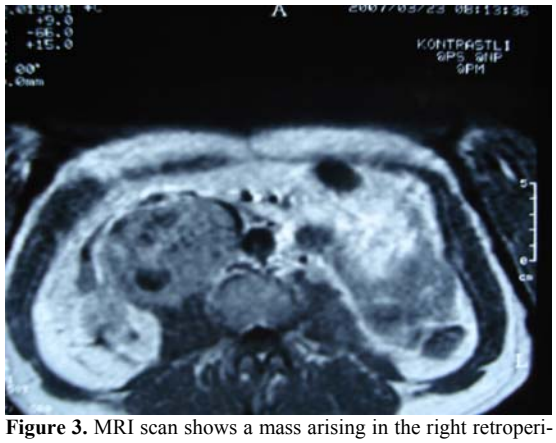

Figure 3. MRI scan shows a mass arising in the right retroperitoneum before second operation.

noma**.** Dissected lymph nodes had no metastatic involvement. MRI scan of the lomber vertebrae showed bone metastasis of L3 vertebra 4 months after the operation. Thus the patient received external beam radiotherapy with a total dose of 30 Gy in 10 fractions to the L3 vertebra for palliative purpose. Treatment was conducted on a linear accelerator of 18 MV photon beam.

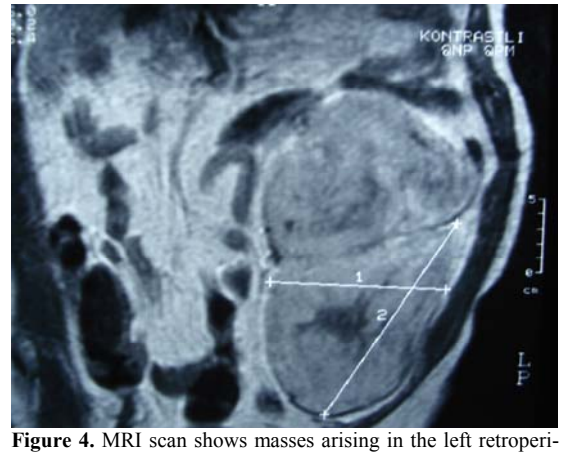

Figure 4. MRI scan shows masses arising in the left retroperitoneum before third operation

DISCUSSION

Malignant peripheral nerve sheath tumors rarely occur in the retroperitoneum. They are usually asymptomatic but as they enlarge they may compress adjacent structures, which leads to a wide spectrum of non-specific symptoms, including lumbar pain, headache, secondary hypertension, abdominal pain and renal colicky pain[2].

In this case report we pointed out that local recurrence is not infrequent and because these tumors are slow growing recurrence may be late. Careful follow-up, including approprite imaging studies, is required. If the tumor recurs, repeat excision should be attempted. In our case, informed consent included the possibility of nephrectomy to avoid any medicolegal complications before first operation because a tight adhesion of the tumor to the left kidney necessitated an en bloc resection.

The retroperitoneal space is quite enough for the development of large tumor masses. The present case combines most characteristics of retroperitoneal neoplasms: Large or very large size, quasi-absent symptomatology, difficulty in preoperative diagnosis, surgical tactics and techniques-quite often, the total extirpation of tumor mass led to the sacrifice of other organs within the limits of a jus-

tified risk-and unforeseeable evolution with relapses having the same characteristics[3].

Radiologic findings are usually nondiagnostic. On MRI, primary retroperitoneal schwannomas show different signal intensity characteristics, including cystic degeneration[4].

Malignant schwannomas are insensitive to chemotherapy and radiation, resulting in poor prognosis[5]. The prognosis of patients with a malignant schwannoma that do not undergo radical surgery is extremely poor and the treatment of choice is complete surgical excision if possible[6,7].

KAYNAKLAR

1- **Inokuchi T, Takiuchi H, Moriwaki Y, et al:** Retroperitoneal ancient schwannoma presenting as an adrenal incidentaloma: CT and MR findings. Magn Reson Imaging, 24: 1389-1393, 2006.

2- **Fass G, Hossey D, Nyst M, et al:** Benign retroperitoneal schwannoma presenting as colitis: A case report. World J Gastroenterol 13: 5521-5524, 2007.

3- **Munteanu M, Pîrşcoveanu M, Gugilă I, et al:** Rare case of retroperitoneal malignant Schwannoma. Chirurgia 99: 345-350, 2004.

4- **Hayasaka K, Tanaka Y, Soeda S, et al:** MR findings in primary retroperitoneal schwannoma. Acta Radiol 40: 78-82, 1999.

5- **Li Q, Gao C, Juzi JT, et al:** Analysis of 82 cases of retroperitoneal schwannoma. ANZ J Surg 77: 237-240, 2007.

6- **Kojima S, Satake I, Tsujii T, et al:** Multiple malignant schwannoma treated with a multidisciplinary therapy-a case report. Gan No Rinsho 36: 939-944, 1996.

7- **Tortorelli AP, Rosa F, Papa V, et al:** Retroperitoneal schwannomas: Diagnostic and therapeutic implications. Tumori 93: 312-315, 2007.