

# A case of Malignant Schwannoma of The Pelvic Sympathetic Plexus In A Girl With Neurofibromatosis Type 1

NÖROFİBROMATOZİS TIP VLI BİR KIZDA GÖRÜLEN PELVİK SEMPATETİK PLEKSUSUN MALİGN SCHWANNOMA VAKASI

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## SUMMARY

**Objective:** To present a case of retroperitoneal malignant schwannoma derived from pelvic sympathetic plexus.

**Institution:** Ankara Social Security Association Hospital, Department of Obstetrics and Gynecology.

**Materials and Methods:** This paper describes the occurrence of a retroperitoneal malignant schwannoma in a 19-year-old girl with neurofibromatosis type 1 (Von Recklinghausen's disease, NF 1) who underwent explorative laparotomy because of a pelvic mass.

**Findings and Results:** Neurofibromatosis is a disease that may effect every body organ. So, it should be kept in mind in the diferential diagnosis of pelvic masses.

**Key words:** Malignant schwannoma, retroperitoneal tumor, pelvis, neurofibromatosis

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Neural sheat tumors are a rare group of soft tissue neoplasms, most often encountered in the cutaneous nerves of extremities, head, neck and trunk (1). Retroperitoneal neural sheat tumors which include benign and malignant schwannomas and neurofibromas are extremely rare. In seperate studies, Stout and Das Gupta and Brasfield found 3% of malignant schwannomas to arise in the retroperitoneal space (1,2).

Neurofibromatosis is a genetic disorder transmitted by an autosomal dominant gene. It is characterized by pigmented cutaneous lesions and generalized tumors of neural -crest origin. It is classified into peripheral (NFI), central (NF2) and mixed forms (3).

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## ÖZET

**Amaç:** Pelvik sempatik pleksustan köken alan retroperitoneal yerleşimli bir malign schwannom olgusunun sunumu.

**Çalışmanın yapıldığı yer:** SSK Ankara Doğumevi ve Kadın Hastalıklardan Hastanesi

**Materyal ve Metod:** Bu makalede tip I nörofibromatosis tanısı ile izlenmekte olan ve pelvik kitle nedeniyle eksploratif laparotomiye alınan 19 yaşındaki bir genç kızda saptanan retroperitoneal yerleşimli bir malign schwannom olgusu sunulmaktadır.

**Bulgular ve Sonuç:** Nörofibromatosis vücudun tüm organlarını etkileyebilen bir hastalıktır. Bu yüzden pelvik kitlelerin ayıncı tanısında göz önünde bulundurulmalıdır.

**Anahtar kelimeler:** Malign schwannom, retroperitoneal tümör, pelvis, nörofibromatosis

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NFI is more common than NF2. A wide variety of neoplasms has been found in conjunction with neurofibromatosis. Many occur coincidentally but others particularly those arising in central nervous system have a much higher incidence in this disorder (4). It has been shown that 3 to 13% of the patients with neurofibromatosis will have malignant schwannoma, usually at deep locations, such as retroperitoneum (1).

Here, we want to present a case of retroperitoneal pelvic malignant schwannoma in a girl with NF1.

## CASE REPORT

A 19-year-old girl was referred to Ankara Social Security Association Hospital, Department of Obstetrics and Gynecology, in 1993 May the 6th, because of a big pelvic mass. She had been suffering from constipation and extreme difficulty in urination for the last 15 days. Rectal examination revealed the presence of a mass filling the pelvis.

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It was learned from the past history that she applied to Hacettepe University Department of Neurosurgery, in 1992, with the complaints of cafe-au-lait spots scattered through her body, being most prominent on her back and a swelling covered with hair and about 6x5 cm in diameter, located in the regions of the zygomatic and temporal bones since birth (Figure 1). Neurologic examination had been found to be normal. Cranial CT had showed an enlargement in right lateral ventricle. Cranial and spinal MRI examinations had revealed the presence of multiple contrast enhancing lesions in intracerebral, intraventricular, intraspinal-extradural spaces. On the basis of these findings and history, she had been diagnosed as neurofibromatosis type I (NF 1).

She had a positive family history of neurofibromatosis. Her father had been operated on because of neurofibromatosis and died on the second day of operation.

The routine laboratory examination results were in normal range. Ultrasonography showed a heterogeneous intrapelvic mass which was about 170x 140x 135 mm in size and contained cystic and solid components. IVP revealed that the bladder was elevated to the level of sacroiliac region, in barium enema, sigmoid colon was narrowed and haustration loss was observed. The results of tumour markers were unremarkable; CEA: 0,5 ng/ml, Ca 125 :14,13 U/ml, AFP: 5 IU/ml.

She underwent laparotomy in 1993, May the 7th because of the pelvic mass. On exploration, the uterus was normal in size and both adnexal structures were in normal appearance. However, the uterus was displaced to the right, because of a hard, irregular and highly vascularized mass which was about 20x15x15 cm in diameter, located in the retroperitoneal region just to the left of the uterus, behind the bladder, and extending alongside the urethra and vagina. The bladder was elevated and the sigmoid colon was compressed by the mass. After reaching the retroperitoneal space, the mass was completely excised. Histopathological examination of the surgical specimen showed it to be a malignant schwannoma derived from the pelvic sympathetic plexus.

Macroscopically, the tumour was a lobulated, capsulated and solid mass of 20x15x15 cm size. In the cross section of the mass, a diffusely solid, dirty-white coloured nodular structure was observed. There were also two other small, elastic and dirty white coloured nodular structures, one being 1,5 cm and the other 1 cm in diameter.

The sections were stained with haematoxylin and eosin, van Gieson, Masson trichrome and silver stain for the light microscopic examination. Additional sections cut from the paraffin blocks were incubated with antibodies against S 100 protein for immunohistochemical investigation.

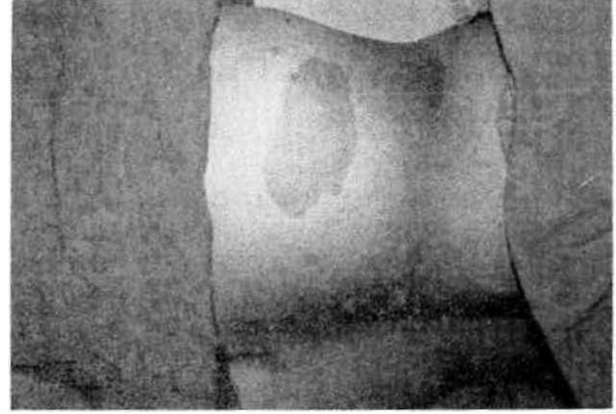


Figure 1. The appearance of cafe-au-lait spots in the back.  
Şekil 1, Cafe-au-lait beneklerinin sırttaki görünümü

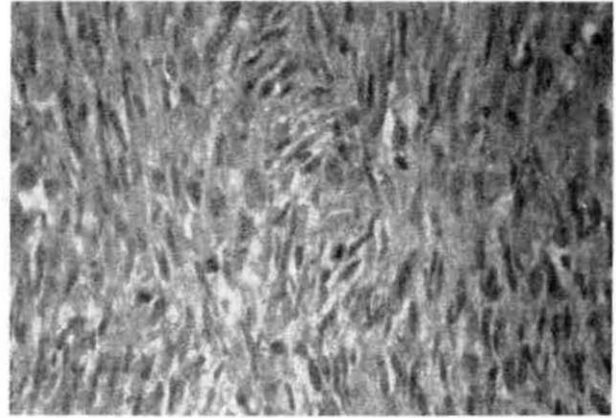


Figure 2. The presence of asymmetrical, oval-shaped, buckled cells or comma nuclei and increased mitotic activity is seen (H.Ex400).  
Şekil 2. Asimetrik, ovak şekilli, eğrilmiş hücreler veya virgül şeklinde nükleuslar ve artmış mitotik aktivite görülmektedir.

In microscopic examination, there were spindle cells forming widely and irregularly dispersed fasciculations. The tumoral tissue was rich in mitosis and some bizarre shaped cells were encountered (Figure 2). The two small nodular structures were neurofibroma nodules.

Postoperative course was uneventful. She was referred to Hacettepe University Department of Neurosurgery for further evaluation and treatment.

The patient was symptom free without any recurrence, when she was seen 4 months later.

## DISCUSSION

Malignant schwannoma is a rare malignant neoplasm, derived from the Schwann cells of the neural sheath (5). It occurs equally in men and women, and has been reported in patients aged from 1 to 95 years

(6). The most common clinical presentation is that of a painless mass. Large tumours are common with consequent pressure effects depending on the location. Our patient applied with the symptoms of urinary retention and constipation which were the result of the pressure of the big mass on the bladder and rectum.

Early investigators believed that, malignant schwannomas occurred only in the setting of neurofibromatosis. Now, however, it is accepted that they arise both in solitary form and in association with NF 1 with approximately equal frequency (7).

NF 1 is one of the most common inherited disorders in humans. The prevalence is estimated to be 1 in 1500 to 3300 (8). Barker et al (9) have localized the NF 1 gene in the proximal long arm of chromosome 17.

There appears to be a tendency for malignant schwannoma to occur in patients with NF 1. Our patient also had neurofibromatosis and a positive family history for this disease.

Retroperitoneal neural sheath tumours are difficult to diagnose early in their development, because they often do not produce symptoms until they become extremely large within the retroperitoneum (10). Investigators have shown that tumour size does not appear to indicate the degree of biological aggressiveness or malignant potential of the neoplasm (10). Mitotic activity is so rare in normal Schwann cells that the presence of one or more mitotic figures suggests malignancy (6). Pathological examination of our patient showed the tumoral tissue to be rich in mitosis. Practically, local invasion is taken as a stronger indicator of malignancy (11)

The schwannian origin of these tumours can be confirmed by their positive immunoreactivity for S 100 protein. The neoplastic cells show varying degree of immunoreactivity for S 100 protein and with the monoclonal antibody against the Leu 7 (HNK-1) antigen (12). We carried out immunohistochemical investigation besides haematoxylin and eosin, van Gieson, masson trichrome and silver stains in our patient. The results of staining procedures were in accordance with malignant schwannoma. Although we examined twice, immunoperoxidase stains were negative for S 100 protein.

In the case of malignant schwannoma, complete excision is the recommended treatment. It is especially important to obtain negative tissue margins to avoid the high incidence of local recurrence (1). Studies

show that, chemotherapy and radiotherapy following complete tumour excision are ineffective in the treatment of bulky and metastatic malignant schwannoma (7,10).

The malignant schwannoma seen with NF 1 is of a higher grade and these patients tend to have a worse prognosis with a 5-year survival rates of 15 to 30% (the retroperitoneal variety having a significantly worse survival) compared to 47 to 75% survival for that of the solitary form. They also have a significantly higher local recurrence rate (1).

It's important to remember that NF 1 has diverse manifestations that may affect every body organ and cause the patient to present to clinicians of any speciality. Finally, we hope to further increase the level of awareness of gynecologists to the existence of these rare tumours.

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