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CLINICAL CASE

Retroperitoneal malignant schwannoma and peritoneal malignant mesothelioma: a case report

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Summary

Malignant schwannoma and peritoneal malignant mesothelioma (MM) are very rare tumors. Schwannoma or neurilemmoma-benign or malignant-do not arise from the nerves, but from the supporting Schwann cells. Malignant peripheral nerve sheath tumors (MPNSTs) commonly are large in size. They are painful and may cause many different symptoms depending on their location and size. MM arises primarily from the surface serosal cells of the pleural, peritoneal, and pericardial cavities.

A malignant schwannoma was diagnosed in a 52-year-old woman, which was surgically treated. After a 17-month disease-free interval, local recurrence was diagnosed. The patient was operated on for second time and MM was diagnosed as second primary tumor, along with recurrence of the malignant schwannoma. The patient received postoperative adjuvant external beam radiotherapy and chemotherapy. Despite combined-modality treatment the disease progressed and the patient was operated on for third time 2.5 years after the first operation with partial tumor resection. She died 3 years after the first diagnosis.

Early diagnosis of these two types of tumors is very difficult because of unspecific clinical symptoms. Single-modality therapy of these tumors has shown poor results. Combined-modality approaches have shown some benefits, but further studies are required.

Key words: chemotherapy, malignant schwannoma, peritoneal malignant mesothelioma, radiotherapy, surgery

Introduction

Primary retroperitoneal tumors are very rare, comprising 0.1-0.3% of all tumors. MPNSTs originate from the nerve sheath rather than from the nerve itself. Ultrastructural studies, however, cannot always document Schwann cells in these tumors derived from nerve sheaths, and therefore the terms MPNST, neurogenic sarcoma, and neurofibrosarcoma are sometimes used [1]. Approximately 40-50% of all schwannomas are localised in the head and neck region and about 3% in the retroperitoneum [2]. Clinically, these tumors are aggressive, locally invasive, and highly metastatic. The diagnostic criteria of MPNSTs are controversial. Histologic findings include hypercellularity, pleomorphic dense nuclei, multiple and abnormal mitoses, and invasion of the surrounding structures. The MPNSTs are usually large (often larger than 5 cm in diameter), partially encapsulated, soft, gray, with hemorrhage and necrosis. Histologically, they are composed of spindle cells with comma-shaped, irregular nuclei. Neural and perineural invasion, mature cartilage, bone, striated muscle, squamous differentiation, and mucin-secreting glands may also be seen [3].

MMs are highly aggressive neoplasms that arise primarily from the surface serosal cells of the pleural, peritoneal, and pericardial cavities. Epidemiologic studies have established that exposure to asbestos

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fibers is the primary cause of MM, and more recent investigations have implicated simian virus 40 (SV40) in the etiology of some MMs [4-8]. MM is a very rare malignant neoplasm comprising 10-20% of all mesotheliomas. It was first described in 1908 [9]. The annual incidence of MM is not known with certainty. The neoplasm can be difficult to diagnose, even by expert pathologists [10].

The incidence of MM appears to be increasing. Peto et al. project that the risk of dying of MM in Western Europe will double over the next 20 years, with the highest risk of approximately 1 in 150 men in the 1945 to 1950 birth cohort [11].

No asbestos exposure can be documented in approximately 30 to 50% of MM cases [10].

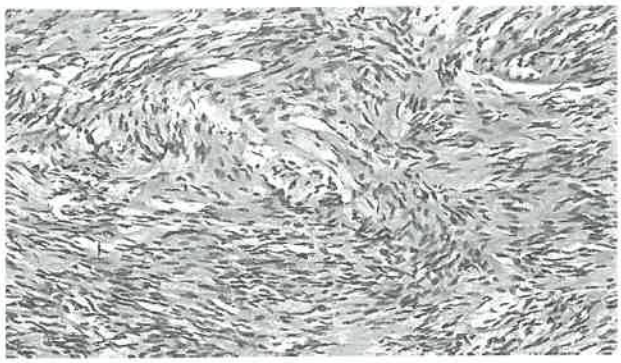


Figure 2. Highly specific histological appearance of malignant schwannoma, reminiscent of Antony type A (Veracay bodies) (H&E ×200).

Case presentation

A 52-year-old woman was treated at the Clinic of Gynecology and Obstetrics, Clinical Center of Novi Sad, because of an extensive mass in the minor pelvis. Vaginal ultrasound examination showed a solid tumor behind the uterus measuring 100 mm in diameter, clearly separated from the gynecological organs. Computed tomography (CT) was also performed with the same results (Figure 1). The patient was operated on and the mass was totally removed along with the left adnexa. The internal iliac vein was injured and sutured during the operation. The postoperative period was uneventful. The final histological diagnosis was malignant schwannoma (Figure 2).

After 17 months, during follow-up by vaginal ultrasound and gynecological examination, a solid and cystic tumor measuring 80 mm in diameter was found behind the uterus in the pouch of Douglas. Also a solid tumor 70 mm in diameter was revealed in the left ischiorectal fossa. Both findings were also confirmed by magnetic resonance imaging (MRI). Grade III left hydronephrosis was also diagnosed by the ultrasound (Figure 3). The patient was operated on and total abdominal hysterectomy with right adnexectomy was performed. The tumor in the pouch of Douglas was totally removed along with 10 metastatic deposits at the parietal peritoneum (Figure 4), but because of the multiple metastatic spread at the parietal peritoneum and the impossibility for abdominal access into the left ischiorectal fossa, the operation ended at this point. The final histological diagnosis was MM, which confirmed a second malignant tumor, and recurrence of the malignant schwannoma in the ischiorectal fossa (Figure 5).

The patient received 4 courses of postoperative chemotherapy with cisplatin 60 mg/m^2 and doxorubicin 50 mg/m^2, every 3 weeks. After the 4th chemotherapy course external beam radiotherapy (2 Gy/day for 10 days) was delivered because of local progression with oedema of the left leg. Despite radiotherapy the disease progressed in the retroperitoneal space of the pelvis, pressing the rectosigmoid (Figure 6).



Figure 1. Computed tomographic image: presacral solid-cystic tumor 10 cm in diameter.

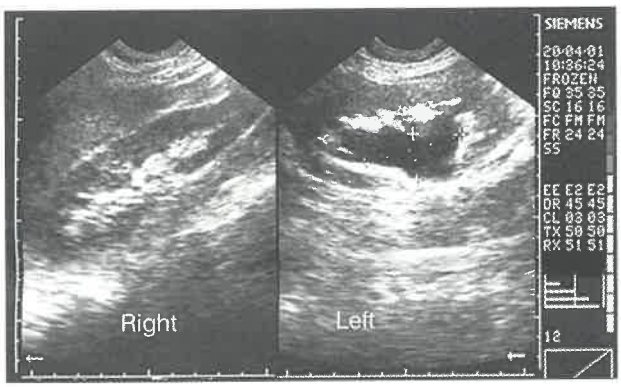


Figure 3. Ultrasound image: grade III left hydronephrosis.

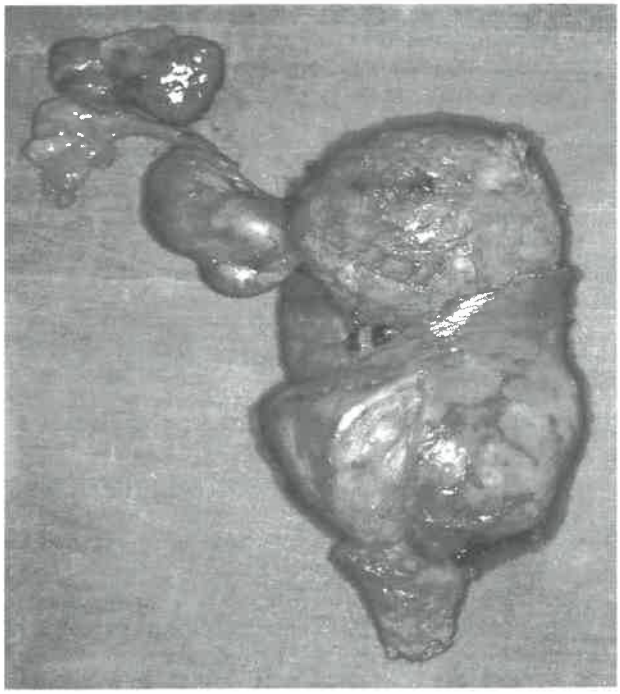


Figure 4. Removed uterus with right adnexa and tumor mass.



Figure 5. Polymorphous histological appearance of malignant mesothelioma (H&E ×100).



Figure 6. Coronal T1WI (A) and transverse T1WI (B) spin echo demonstrate massive and extensive recurrent tumor.

The patient was then operated on for a third time and recurrence of the malignant schwannoma was confirmed, which infiltrated the abdominal wall and large vessels. A partial tumor resection was performed.

Four months later the patient was hospitalized because of obvious disease progression and died 3 years after the first diagnosis.

Discussion

The clinical diagnosis of retroperitoneal malignant schwannoma is very difficult. The localisation of the tumor allows a growth without clinical symptoms at the beginning, and usually symptoms appear because of pressure on local organs. Pain is a usual symptom which is constant or acute if necrosis or tumor bleeding occur. Diagnostic methods are clinical examination, ultrasound, CT, MRI, and sometimes urography and angiography. Surgery is the treatment of choice. Despite complete removal, recurrences are relatively frequent, leading to a 75% 5-year survival rate. Patients with neurofibromatosis and a MPNST have a 15 to 30% 5-year survival rate. Radiation therapy can offer significant palliation and prolongation of life, but no cures have been observed. Other forms of therapy (chemotherapy, immunotherapy, etc.) have little to offer [12,13].

The clinical symptoms of peritoneal MM are nonspecific. Patients usually present with symptoms and signs of advanced disease including pain, ascites, weight loss, or an abdominal mass [14, 15]. Preoperative diagnosis is very difficult. No satisfactory staging system has been proposed for peritoneal MM, which is usually confined to the abdomen at diagnosis. Classic findings on CT scan include mesenteric thickening, peritoneal studding, hemorrhage within the tumor mass, and ascites; however, patients may have advanced disease with relatively normal CTs [16]. In 90% of the patients ascites occurs. Massive ascites may result in confusion between mesothelioma and severe cirrhosis. Cytology establishes the diagnosis in only 5 to 10% of the cases [17]. Peritoneal MM can be confused with adenocarcinomas arising from any abdominal organ such as adenocarcinoma of the ovary or carcinoma arising from müllerian duct remnants in the peritoneum.

Ultimately, definitive diagnosis requires adequate tissue sampling, preferably from peritoneoscopy or an open biopsy.

Complete surgical resection is rarely, if ever, feasible, and has not been shown to offer survival

benefit in the absence of additional therapy. Surgical intervention can provide palliation for small bowel obstruction and relief of massive ascites by peritonovenous shunting or paracentesis via Tenckhoff's catheter [18]. The role of radiation therapy in the treatment of MM remains unclear. There are some reports about the effectiveness of external beam radiotherapy and intraperitoneal instillation of radiocolloids [19-21].

Because the results of intravenous chemotherapy in patients with MM have been disappointing, interest has focused on the use of intraperitoneal chemotherapy [22]. Advantages include greatly enhanced drug concentrations in the peritoneal cavity and decreased systemic toxicity. Intraperitoneal cisplatin in 19 patients (combined with mitomycin in 18) resulted in 2 (10%) disease-free status for more than 5 years [23]. Results obtained in these reports were from small groups of patients, so further investigations are required.

Today, combined-modality approaches are carried out in the treatment of MM, because single-modality therapies were unsuccessful [15,24,25].

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