Retroperitoneal Fibrosis Secondary to Ovarian Malignancy: Case Report

Over Malignensisine Sekonder Retroperitoneal Fibrozis

ABSTRACT A 48-year-old woman was admitted with the complaint of lower abdominal pain for two years and left leg edema for one month. Physical examination revealed bilateral pelvic mass and imaging techniques including ultrasound and computed tomography revealed a normal uterus and solid-cystic mass invading bilateral ovaries. Intravenous pyelogram showed a delay of the contrast material and mild hydronephrosis on the left side. The patient underwent total abdominal hysterectomy, bilateral salpingo-oopherectomy, ureterolysis, omentectomy, pelvic and paraaortic lymph node dissection. On the left side, retroperitoneal space was extremely fibrotic and the left ureter was rigid like a lead tube. Histopathologic examination revealed bilateral malignant serous tumor of the ovary and retroperitoneal fibrosis on the left side. Lower limb edema and renal function improved rapidly. Although retroperitoneal fibrosis is a rare disease and is usually idiopathic, 8% of cases are associated with malignancy. Ovarian malignancy related to retroperitoneal fibrosis has not been reported before in the literature.

Key Words: Ovarian neoplasms; retroperitoneal fibrosis


Anahtar Kelimeler: Over tümörleri; retroperitoneal fibrozis


Retroperitoneal fibrosis is characterized by the development of extensive fibrosis throughout the retroperitoneum. This fibrosis leads to entrapment and obstruction of the retroperitoneal structures, including ureters and retroperitoneal lymphatics.1 The incidence of retroperitoneal fibrosis estimates vary from 1:200 000 to 1:500 000 per year. The peak incidence is at 40-60 years of age.2
There are idiopathic and secondary forms of retroperitoneal fibrosis. The pathogenesis of idiopathic disease is unclear. Retroperitoneal fibrosis can also be secondary to drugs, malignancies, infectious disease, trauma or abdominal surgery. Secondary forms of retroperitoneal fibrosis account for over 30% of the cases.3

In this report, we presented a patient with retroperitoneal fibrosis secondary to ovarian malignancy.

CASE REPORT
A 48-year-old (gravida-2 para-2) woman with complaints of lower abdominal pain for two years and edema in the left leg for one month admitted to our clinic. She had no past surgical history. She had been treated with antibiotics and non-steroidal anti-inflammatory drugs for several courses but did not show any improvement. Physical examination revealed edema in the left lower limb. Gynecologic examination revealed a normal uterus and bilateral adnexal masses. Ultrasound demonstrated that these adnexal masses were related to ovaries and renal pelvis was dilated on the left side. Computed tomography scan was performed, there were bilateral adnexal masses with solid and cystic areas (Figure 1). There was no clear boundary between uterus and mass in the left site. Delay of contrast material was observed in the left kidney. Intravenous pyelogram pointed out that the left kidney filtrated urine later than the right kidney, the ureter and the renal pelvis were mildly dilated (Figure 2). In laboratory investigations, Ca-125 level was 255.5 U/mL and serum creatinine level was 1.48 mg/dL. Erythrocyte sedimentation rate, C-reactive protein, white blood cell count, and alkaline phosphatase were normal. The patient underwent exploratory laparotomy, total abdominal hysterectomy, bilateral salpingo-oopherectomy, omentectomy, and pelvic and paraaortic lymph node dissection. Retroperitoneal space was extremely fibrotic and the ureter was like a lead tube in the left side. Ureterolysis and resection of ureteral stricture were performed. Histopathologic examination revealed serous carcinoma of the ovary in the both sides. Although the capsule integrity of the tumor was preserved, there was a tumor metastasis on the left tuba and the abdominal cytology result was malignant. She was diagnosed with Stage IIC disease. The biopsy specimen from the paraureteral-retroperitoneal space revealed numerous small blood vessels with perivas-
cular lymphocytic infiltrate (Figure 3). The lower limb edema advanced in the early postoperative period. Venous Doppler of the lower limb revealed a venous compression, but not thrombosis. The lower limb edema and renal function improved rapidly. Six courses of cytotoxic chemotherapy including carboplatin AUC6 d 1500 mg and Paclitaxel 175 mg/m² 300 mg were administered on a monthly basis. The patient was followed up for 16 months, and there was no pelvic or retroperitoneal disease.

DISCUSSION

Retroperitoneal fibrosis presents insidiously, often making the diagnosis difficult. Early symptoms are often non specific systemic complaints. The most common presenting symptom is pain in the lower back, flank or abdomen.¹,³ There are chronic inflammation, fibroblast proliferation, and excessive extracellular matrix deposition. Acute phase reactants elevate in varying degrees.³ The elevation is related to the inflammatory nature of the disease. Encasement of vessels and ureters by the retroperitoneal mass may cause obstructive complications. Clinical manifestation and course are similar in both idiopathic and secondary form of the disease. The late stage of all retroperitoneal fibrosis forms is characterized by progressive ureteral obstruction.³

Approximately 8% is associated with malignancy, mostly metastatic disease from lung, breast, prostate or digestive tract, and malignant lymphoma or sarcoma.¹,⁴ Ovarian malignancy associated with retroperitoneal fibrosis has not been reported in the literature before.

The presented case had been consulted by various specialists for two years but no significant pathology was found. During her last admission, abdominal ultrasound revealed a dilated renal pelvis and bilateral adnexal masses. Exploratory laparotomy was performed and it was noticed that the ureter was fixed and like a lead tube at the retroperitoneum. In patients with retroperitoneal fibrosis associated with malignancy, metastatic tumor cells within the retroperitoneum cause an exuberant desmoplastic response.¹ Although there was no metastatic tumor cells, severe desmoplastic response was demonstrated in the retroperitoneal space.

In the presented case, the onset of symptoms associated with retroperitoneal fibrosis started earlier than the adnexal mass history. Although ovarian masses were bilateral, the fibrotic lesion of retroperitoneum was unilateral. Additionally, tumor capsule was intact at the ipsilateral ovarian tumor. Hence, it was thought that the etiology of retroperitoneal fibrosis might also be idiopathic.

Optimal therapy for the idiopathic retroperitoneal fibrosis is unknown.¹ The therapy aims to relieve the obstruction caused by fibrosis, stop the progression of the fibrotic process, and to prevent recurrence.¹ It has been advocated to use medical therapy alone for patients with idiopathic retroperitoneal fibrosis with only mild or no ureteral involvement.³,⁵ If significant renal dysfunction due to obstruction of the urinary tract is present, immediate relief of obstruction is generally advised, with open surgical, percutaneous or endoureteral interventions.

The management of secondary retroperitoneal fibrosis includes the removal of any identifiable inciting agents. The surgical approach provides simultaneous relief of mechanical obstruction and an open biopsy, and exploration to exclude lymphoma or metastatic cancer as the etiology of the fibrotic process in the patients in whom a differential diag-
nosis was not made. In the presented case, the ovarian tumor which could be the inciting agent was removed and ureterolysis was performed. The clinical course of retroperitoneal fibrosis improved rapidly. Even if retroperitoneal fibrosis seemed to be associated with ovarian malignancy, the possibility of an idiopathic etiology should be considered.

Although the diagnosis of idiopathic retroperitoneal fibrosis relies primarily on imaging studies and histopathology to rule out alternative conditions such as malignancy, many aspects of the disease are poorly understood. In the present case, it was not possible to differentiate idiopathic or secondary form of disease.

REFERENCES

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