fore, early operative therapy consisted of palliative drainage procedures. The morbidity from resection depends on the ability to separate the tumor from adjacent organs and blood vessels. Postoperative retroperitoneal drainage should be considered if there is dissection near the pancreas or if extensive lymphatic disruption occurs. Prognosis for patients with resected mature cystic teratomas of the retroperitoneum is now excellent.[11]

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THE TREATMENT OF RETROPERITONEAL FIBROMATOSIS WITH MEDROXYPROGESTERONE ACETATE

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Wide excision is the recommended primary therapy for retroperitoneal fibromatosis. Radiation therapy and a variety of medications have been used to treat patients with recurrent tumors, but the response to these agents has not been uniform. The patient presented was successfully treated with medroxyprogesterone acetate for recurrent retroperitoneal fibromatosis that was refractory to multiple operative resections and radiation therapy. (*Obstet Gynecol 70:502, 1987*)

Retroperitoneal fibromatosis is a fibrotic process of the retroperitoneum that frequently produces ureteral obstruction.[1] This neoplasm, also known as a desmoid tumor, is generally regarded as benign.[2] Histologically, it is characterized by a proliferation of well differentiated fibroblasts, an infiltrative pattern of growth, and the presence of variable amounts of collagen between the proliferating cells. There are no cytologic features of malignancy, and scant or absent mitotic activity.[3]

While this disease is usually considered idiopathic, several potential etiologies have been proposed. These include retroperitoneal trauma, regional enteritis, diverticulitis, appendicitis, irradiation, abdominal surgery, and genitourinary infections.[1] A relationship between methysergide therapy for migraine headaches and the development of retroperitoneal fibromatosis has been documented.[1,4–7] A similar drug, lysergic acid diethylamide (LSD), has also been implicated as a possible etiologic factor.[4]

The tumor is most commonly diagnosed during the third and fourth decades of life.[3] The reported distribution by gender has been variable. Although male[1] or female[8] predominance has been reported in different series, other authors have noted that the disease is more common in women in the younger age groups, but predominant in men when all age groups are considered.[9]

The recommended therapy for retroperitoneal fibromatosis is wide excision of the tumor.[1,2,8,10–12] Other treatments have included radiation therapy,[2,11,13] corticosteroids,[1,12,14,15] antibiotics,[15] phenylbutazone,[15] indomethacin,[10] and ascorbate.[10] While primary excisio:. is usually curative, recurrence rates of 24% have been noted.[2] The lack of a firm understand-

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ing of the pathophysiology of this disease has made the treatment of patients with recurrent tumor difficult. Recently, it has been postulated that the growth of these neoplasms may be hormonally controlled.[8,16,17] Despite the suggested benign nature of retroperitoneal fibromatosis, mortality rates of 13% have been reported.[9]

A patient with retroperitoneal fibromatosis is presented. Despite multiple operative resections and radiation therapy, the tumor continued to recur.

Case Report

The patient was a 22-year-old woman, gravida 2, para 1, abortus 0, at 20 weeks' gestation when a right-sided pelvic mass and right ureteral obstruction were discovered. She underwent exploratory laparotomy with partial resection of a solid retroperitoneal right pelvic sidewall mass, which required removal of a segment of the ureter. A right ureteroneocystostomy was performed, and a percutaneous nephrostomy tube was inserted. The uterine examination was consistent with a 20-week gestation, and the ovaries and fallopian tubes were normal. Histologically, the mass was a desmoid tumor composed of relatively bland-appearing fibroblasts in a dense collagenous matrix (Figure 1). There was no evidence of necrosis and a paucity of mitoses. It appeared to be an aggressive localized neoplasm that infiltrated and entrapped the normal retroperitoneal structures. Assessment of the margins of resection was difficult because the tumor blended imperceptibly with the surrounding fibrous tissue. No postoperative therapy was administered. The nephrostomy tube was removed after six weeks because of poor urine production. The pregnancy progressed normally, and the patient underwent an uncomplicated vaginal delivery at term. Previously, the patient used a diaphragm for contraception, although oral contraceptives had been used for a short time. She had a history of acne vulgaris since age 14 and has received tetracycline intermittently for this condition. Otherwise, she had no significant medical history and took no regular medications.

Five months postpartum, the patient was found to have a nonfunctioning right kidney on renal scan. Physical examination revealed a 5 × 8-cm right pelvic mass. Exploration showed a solid retroperitoneal mass involving the right pelvic sidewall. A total abdominal hysterectomy, right salpingo-oophorectomy, right nephrectomy, and resection of the tumor were performed. The mass was incompletely excised, with tumor remaining at the bifurcation of the right common iliac artery. Histologic examination of the neoplasm confirmed a recurrence of the retroperitoneal fibromatosis. Postoperatively, the patient received 4500-rad whole pelvis irradiation with an additional 500-rad boost to the right pelvic sidewall.

One year later, at age 24, the patient had a normal pelvic examination, but a surveillance computed tomography (CT) scan showed a 10 × 15-cm mass occupying the right renal bed. Fine-needle aspiration was consistent with recurrent retroperitoneal fibromatosis. At exploration, the tumor was found to be attached to the vertebral bodies posteriorly, invading the inferior vena cava medially, and extending to the pelvic brim inferiorly. There was no gross pelvic involvement. During the dissection, including resection of the vena cava, all visible tumor was removed. The left fallopian tube and ovary were excised. Intraoperatively, the patient received 3000-rad electron beam irradiation directly to the tumor bed.

The patient remained disease-free until age 26, when a 5 × 10-cm right pelvic sidewall mass was palpated and clearly defined by CT scan (Figure 2). Recurrent retroperitoneal fibromatosis was diagnosed by fine-needle aspiration. A regimen of 1 g of medroxyprogesterone acetate intramuscularly every two weeks was instituted. Subsequent examinations and CT scans substantiated a decrease in the size of the tumor. After one year of therapy, the regimen was changed to 1 g of medroxyprogesterone acetate intramuscularly each

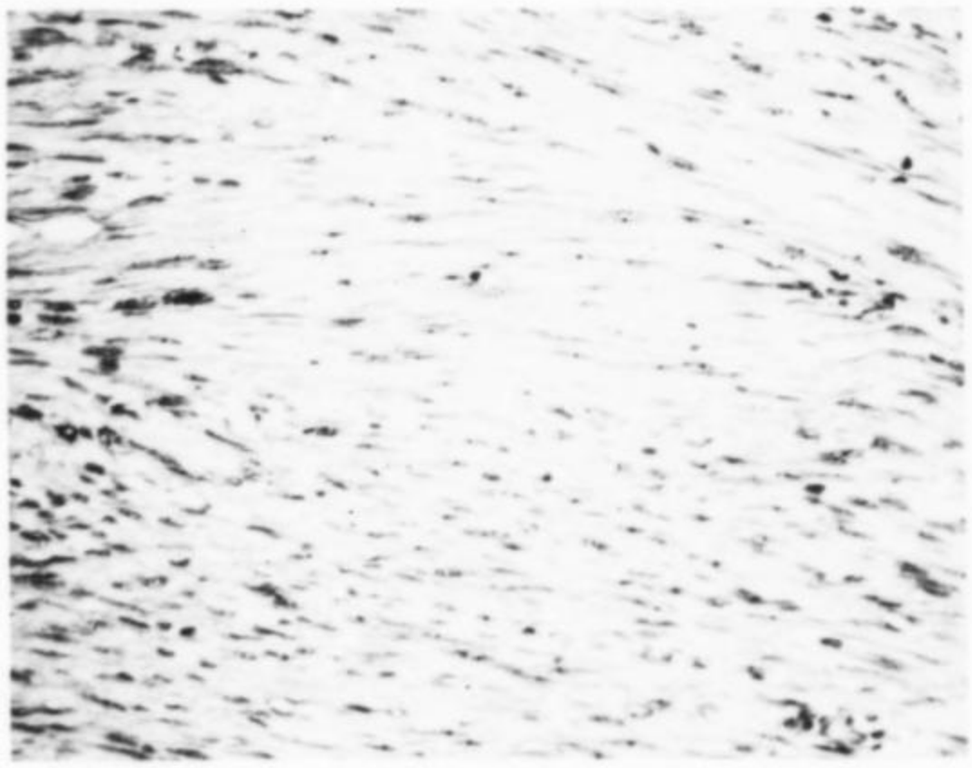

Figure 1. Photomicrograph demonstrating the histologic features of retroperitoneal fibromatosis (×250).

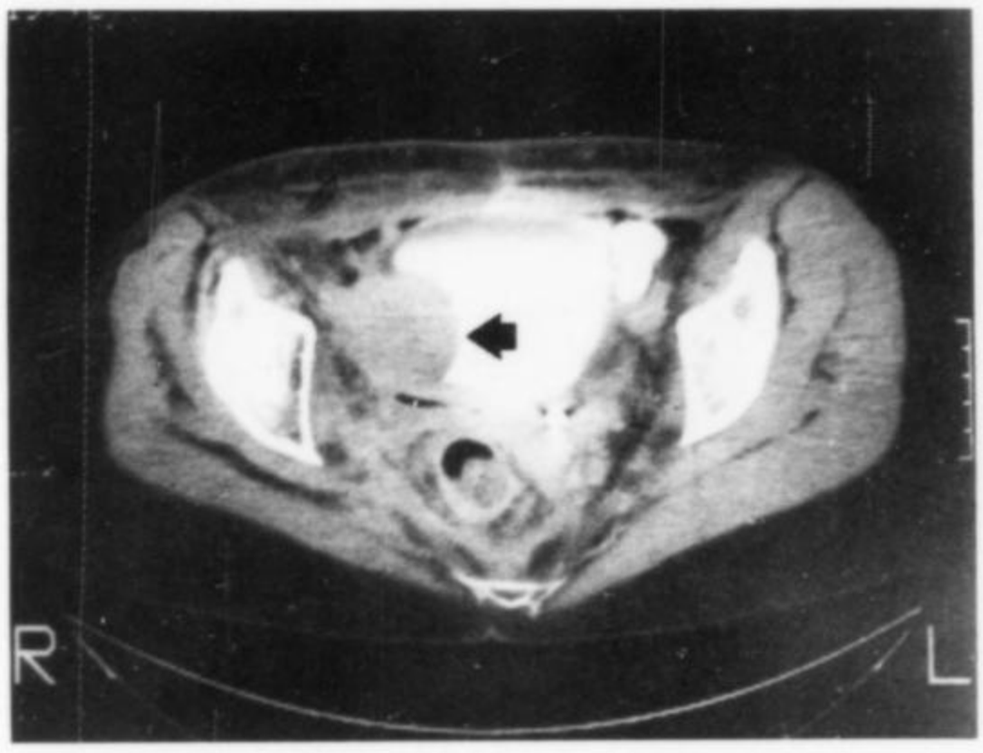

Figure 2. Pelvic CT scan showing recurrent tumor (*arrow*) after multiple operations and radiation therapy.

month. After 18 months of treatment, the patient has no palpable tumor. The CT scan shows only a small density in the area of the tumor (Figure 3). The patient's therapy and surveillance will continue.

Discussion

Patients with retroperitoneal fibromatosis may exhibit a variety of clinical courses. Although no etiology is discovered for most cases, specific agents such as methysergide have been implicated in a significant number of these tumors. Most cases will be cured by wide excision, but 24% of patients will develop recurrent tumor. Of the patients with a recurrence, additional cures have been achieved through further operations and the use of radiation therapy or a variety of agents, including corticosteroids, antibiotics, phenylbutazone, indomethacin, and ascorbate. Therapeutic response to these treatments has not been uniform.

Hayry et al[17] discovered that the retroperitoneal fibromatosis in women in the reproductive age group grew at a much faster rate than in any other subgroup based on age and gender. Using qualitative methods, they found that their study group was predisposed to estrogen predominance, and suggested that there may be an element of hormonal control in the growth of the retroperitoneal fibromatosis in these patients. Our patient continued to develop recurrent tumor despite multiple operations and radiation therapy. It is clear that, in this patient, the retroperitoneal fibromatosis was a persistent and life-threatening process. In thesearch for additional therapy, the theoretic hormonal control of this tumor was considered. This led to the use of a high-dose progestational agent. Tissue was not available for receptor analysis at the institution of treatment. While the results of hormonal therapy in this patient are encouraging, further investigation of these tumors, including estrogen and progesterone receptor determination, is necessary to increase the understanding of the pathophysiology of this potentially fatal disease. This knowledge may lead to the development of consistently successful treatment for patients who fail primary operative therapy.

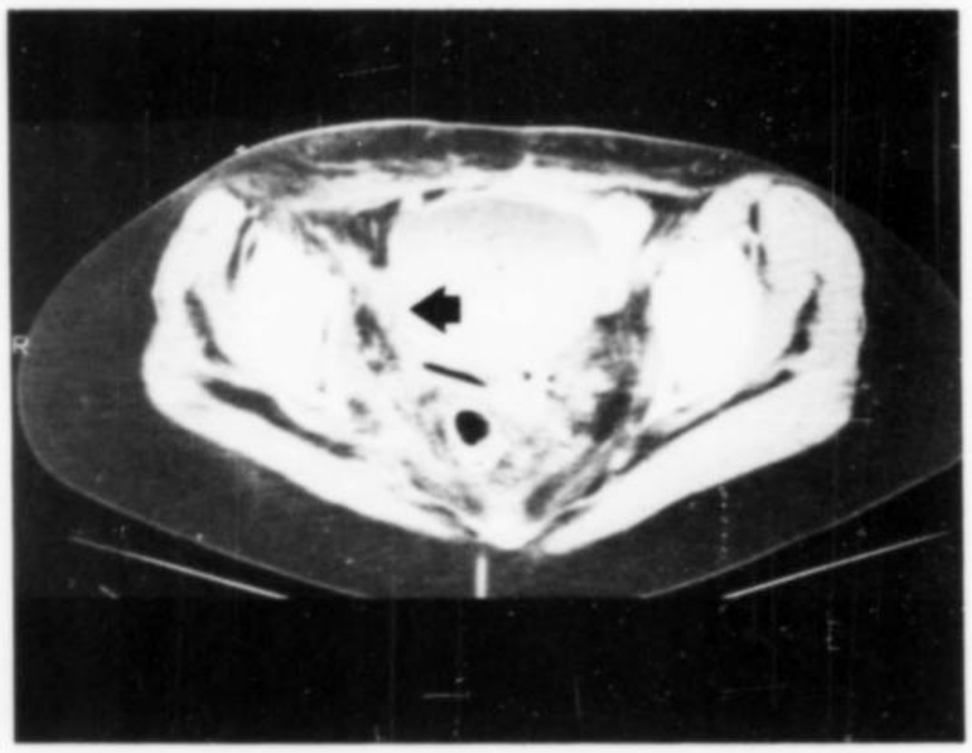

Figure 3. Pelvic CT scan showing only a residual density (*arrow*) in the area of the tumor after treatment with medroxyprogesterone acetate.

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