Giant desmoid tumor in a case of ileal neobladder

Tahir Karadeniz, MD, Caner Baran, MD, Medih Topsakal, MD
Department of Urology, Okmeydani Research Hospital, Istanbul, Turkey

KARADENIZ T, BARAN C, TOPSAKAL M. Giant desmoid tumor in a case of ileal neobladder. The Canadian Journal of Urology. 2010;17(1):5038-5039.

Aggressive fibromatosis or desmoid tumor, is a histologically benign entity with unknown etiology that may present a serious clinical course. Due to its high tendency to recur and local aggressive behavior and as there is no established effective medical treatment, complete surgical excision remains the sole management. To our knowledge, we describe the first case of giant desmoid tumor which arised from ileal neobladder mesenterium in a bladder cancer patient with orthotopic substitution.

Key Words: aggressive fibromatosis, desmoid, bladder cancer, cystectomy, urinary diversion

Introduction

The fibromatoses are a group of fibroblastic proliferations distinguished by their tendency to grow in an infiltrative fashion and, in many cases, to recur after surgical excision. They are classified as the superficial fibromatoses and the deep (aggressive) fibromatoses (desmoid tumors).[1] The superficial fibromatoses, (Dupuytren contracture, Peyronie's disease), arise in the superficial fascia. The deep fibromatosis, is a mesenchymal neoplasm characterized by fibrous connective tissue proliferation and develops from muscle connective tissue, fasciae and aponeuroses.[1] Due to location, they can be divided into several groups: extraabdominal - shoulder and pelvic girdle, chest and neck muscles and in extremities; abdominal - abdominal wall muscles; intra abdominal - concerning small intestine mesentery connective tissue, pelvic or retroperitoneal space.[2] The deep fibromatoses have greater tendency to recur; although they grow in a locally aggressive manner they do not metastasize and rapid growth is occasionally seen.[1,3] Desmoid tumor (DT) often presents a problem in recognition and management because of: the lack of diagnostic awareness, as a consequence of its rarity, and the striking discrepancy between its benign histological appearance and its local aggressive behavior.[4] Mortality rate may be as high as 10% because of its tendency to infiltrate surrounding tissue and its recurrence rate (40%).[2,5,6] We present a new case of giant DT which recurred 3 years after the initial attempt for excision.

Accepted for publication August 2009

Address correspondence to Dr. Caner Baran, Nevbahar Mah. Topcueminbey Sok Kahyaoglu Apt. 1/4 Daire:5 Fatih Istanbul 34096 Turkey

Case report

A 6 cm right presacral mass was diagnosed in a 60 years male after radical surgery for muscle invasive bladder cancer. He had previously undergone TUR of bladder followed by pelvic irradiation. In 2002 he was offered radical cystoprostatectomy and bilateral extended pelvic lymph node dissection with ileal neobladder reconstruction due to muscle invasive transitional cell carcinoma of bladder. The specimen yielded pT1G3 TCCa with diffuse CIS, lymph nodes were not involved. Without further treatment he was free of disease for 2 years until the presacral mass was detected on pelvic computed tomography (CT), Figure 1a. CT guided biopsy intervention failed as the lesion lied between promontorium and the afferent limb of the neobladder. Radical surgical excision was unsuccessful due to invasion of the mesentery of ileal neobladder and the right common iliac vessels. Histopathology yielded a hypocellular lesion with thick collagen and spindle cell component composed of active fibroblasts with no cytological atypia. It was classified among pseudosarcomatous reactive stromal lesions and final diagnosis was aggressive desmoid tumor, Figure 2.

After 3 years without remarkable change of the mass and no evidence of bladder cancer recurrence, he presented with a right abdominal mass of 30 cm in diameter. Abdominal CT showed a soft tissue mass at the mesentery of neobladder which obliterated vena cava and compressed abdominal aorta, Figure 1b. Complete surgical removal with ileal conduit diversion and resection of 15 cm intestinal segment was performed. On the second postoperative day he died of massive pulmonary thromboembolism. Pathological examination of the specimen again revealed a diagnosis of aggressive desmoid tumor.

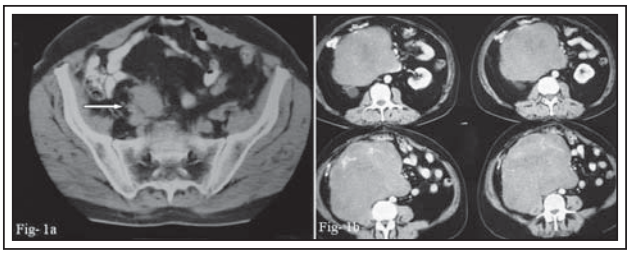

Figure 1a, 1b. CT appearance of the lesion initially and 3 years after.

Discussion

A branch of DTs; mesenteric fibromatosis is an infiltrating fibroproliferative process composed of fibroblasts and myofibroblasts with uniformly bland nuclear features.[7] The most common primary tumor of the mesentery is fibromatosis, they are located in the mesentery of the small bowel.[8] Concerning our case, the tumor was located at the mesentery of the ileal neobladder thus enabling us to define it as a mesenteric fibromatosis. Although etiology is unclear previous surgery or trauma as well as radiation may be a triggering factor.

DTs are rare, slowly growing and histologically benign tumors without metastatic potential and lack malignancy signs in nuclear and cytoplasmic characteristics. Tumor cells are generally actin positive and the extent of staining correlates positively with cellularity; desmin and S-100 protein also commonly stain a small part of tumor cells.[9] In this case actin was positive whereas S-100 and desmin were negative, that correlated with literature.

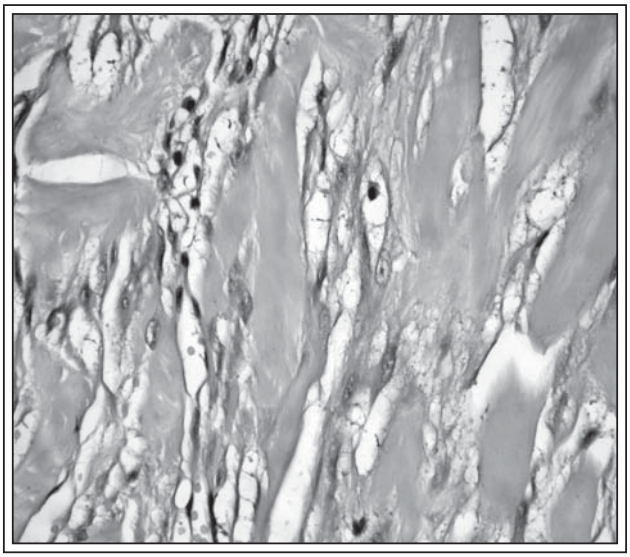

Figure 2. Microscopy showing thick collagen and spindle cell component composed of active fibroblasts with no cytological atypia. (H&E 400x)

Medical or surgical therapies are unsatisfactory due to resistance of drugs and high postsurgical recurrence rates. Consequently, treatment has to be individualized. Owing to its tendency to recur and acceleration of growth after initial surgical attempts, suggested therapy is complete exicision of the mass with negative surgical margins. Yet, there are authors who reserve surgical procedures to complicated cases as intestinal or ureteral obstruction and these interventions are associated with high mortality rates (10%-60%) due to excessive blood loss.[10]

In the case presented, desmoid tumor originated form the mesentery of ileal neobladder, which did not permit complete excision without sacrificing neobladder. No medical therapy was offered initially as it remained silent for 3 years. Although we have succeeded in complete resection with acceptable operative complications and no life threatening bleeding, the patient died of pulmonary embolism.

Occurrence of DTs following major surgical abdominal intervention, especially with a history of irradiation, should be kept in mind. Subsequently, the best treatment modality appears to be complete surgical excision before the lesion gets symptomatic as its tendency to invade local structures that would lead untoward consequences. □

References

1. Robbins & Cotran. Pathologic Basis of Disease, 7th edition pp783-784. Elsevier 2007.
2. Ferenc T, Sygut J, Kopczyński J, Mayer M, Latos-Blelenska A, Dzlki A, Kulig A. Aggressive fibromatosis (desmoid tumors): definition, occurrence, pathology, diagnostic problems, clinical behavior, genetic background. *Pol J Pathol* 2006;57(1):5-15.
3. Rodriguez-Bigas MA, Mahoney MC, Karakousis CP, Petrelli NJ. Desmoid tumors in patients with familial adenomatous polyposis. *Cancer* 1994;74(4):1270-1274.
4. Rakha EA, Kandil MA, El-Santawe MG. Gigantic recurrent abdominal desmoid tumour: a case report. *Hernia* 2007;11(2): 193-197.
5. Nursal TZ, Abbasoglu O. Sporadic hereditary pancreatic desmoid tumor: a new entity*? J Clin Gastroenterol* 2003;37(2):186-188.
6. Kulaylat MN, Karakousis CP, Keaney CM, McCorvey D, Bem J, Ambrus JL Sr. Desmoid tumour: a pleomorphic lesion. *Eur J Surg Oncol* 1999;25(5):487-497.
7. Shields CJ, Winter DC, Kirwan WO, Redmond HP. Desmoid tumors. *Eur J Surg Pathol* 2001;27(8):701-706.
8. Weiss SH, Goldblum JR. Enzinger and Weiss's Soft Tissue Tumors (fourth edition). St. Luis: Mosby 2001, pp. 309-346.
9. Kempson RL, Fletcher CDM, Ewans HL, Hendrickson MR, Sibley RK. Atlas of Tumor Pathology. Tumors of the Soft Tissues. Published by the Armed Forces Institute of Pathology, Washington, D.C. (Bethesda, Maryland 2001), pp. 68-83.
10. Clark SK, Phillips RKS. Desmoids in familial adenomatous polyposis. *Br J Surg* 1996;83(11):1494-1504.