

## CASE REPORT

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# An Aggressive Retroperitoneal Fibromatosis

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## ABSTRACT

**Introduction:** Aggressive fibromatosis (AF) is a heterogeneous group of mesenchymal tumors that have locally infiltrative growth and a tendency to relapse. The clinical picture is often conditioned by the obstruction of the ureter or small intestine. Diagnosis is based on clinical, radiological and histological parameters. **A case report:** We report a case of male patient, aged 35 years, with the retroperitoneal fibromatosis. He reported to the physician because of frequent urination with the feeling of pressure and pain. Computed tomography revealed the tumor mass on the front wall of the bladder with diameter of 70mm with signs of infiltration of the musculature of the anterior abdominal wall. Endoscopic transurethral biopsy showed proliferative lesion binders by type of fibromatosis. The tumor was surgically removed in a classical way. The patient feels well and has no recurrence thirty-six months after the operative procedure. **Conclusion:** The complete tumor resection is the therapeutic choice for the primary tumor as well as for a relapse.

**Key words:** aggressive fibromatosis, desmoid, surgery, radiation therapy, relapse.

## 1. INTRODUCTION

Fibromatosis (desmon tumor—from the Greek word desmos—bar), also known as aggressive fibromatosis (AF) or musculoaponeurotic fibromatosis is a heterogeneous group of mesenchymal tumors whose histological picture characterize monoclonal proliferation of fibroblasts and myofibroblasts with the production of intracellular collagen (1). The disease was first described by Mueller in 1838 as a band, or a cyclic structure, and the term fibromatosis as a group of similar changes, was first used by Stout in 1961 (2).

Fibromatosis is a rare disease and makes 0.03% of the tumors (3) and 3% of all the soft tissue tumors (4). The incidence is 2.4–4.3 new cases per million people per annum. It occurs most often between 25 and 35 years of age (5). Most cases are sporadic but there is also a link with familial adenomatous polyposis and Gardner's syndrome and it is related to mutations in the APC gene chromosome 5q22 (6). There is also mentioned the trauma and long-term use of estrogen (7). Reports on gen-

der predominance are different but till now have been reported more frequently in females (8).

It is a histological entity that can have characteristics of benign fibrosis since it does not give metastases and fibrosarcoma for possible locally aggressive growth (1). The etiology of the disease is not known. Irrespective of the way it causes benign significant morbidity and mortality. In case of recurrent disease, unlike sarcoma, non-differentiated in the high-ranking malignancies (9). Based on clinical images, patient's age and the nature of the change AF is divided into two large groups with several subgroups (Appendix 1). The clinical picture is often conditioned by the obstruction of the ureter or small intestine.

Diagnosis is based on clinical, radiological and histological features. The main characteristic of AF is the infiltration of muscles and deep tissue. MSCT is the method of choice for evaluation of intra-abdominal changes, and MRI for changes in the extremities, head and neck and the abdominal wall and thorax (10).

Superficial (fascial) fibromatosis
Palmar fibromatosis (Dupuytren's Disease)
Plantar fibromatosis (Ledderhose's disease)
Knuckle pads
Deep (musculoaponeurotic) fibromatosis
Extra-abdominal fibromatosis (extra - abdominal desmoid tumor)
Fibromatosis of the abdominal wall (abdominal desmoid tumor)
Intra-abdominal fibromatosis (intra-abdominal desmoid tumor)
Pelvic fibromatosis
Mesenteric fibromatosis
Mesenteric fibromatosis in Gardner's syndrome

Table 1. Classification of fibromatosis

**2. CASE REPORT**

A man aged 35, works as a laborer, admitted because of problems with urination as a feeling of pressure and pain during micturition, which persist for the last few months. In the regional medical institution treated as acute prostatitis. Good general condition, and the physical examination findings orderly. The laboratory findings of elevated CRP: 145mg / l, while the other parameters were of the reference values. On the ultrasound examination of the urinary tract, in the bladder was revealed tumor change the largest diameter to 70mm, which covers the front wall of the bladder, and whose origin cannot be determined. Computed tomography revealed that in the pelvic there was edge well vascularised formation that covers the front wall of the bladder, with visible

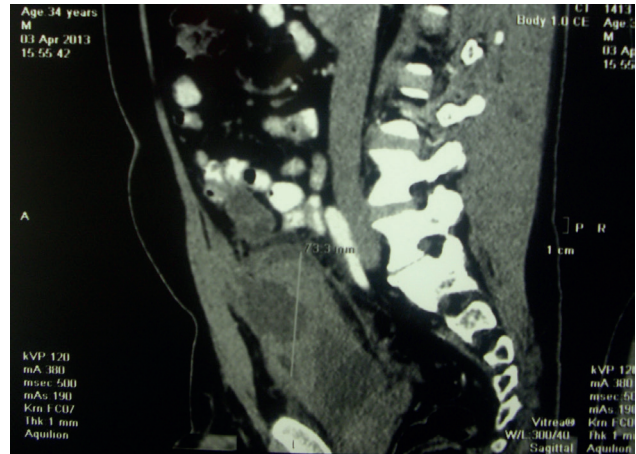


Figure 1. CT - tumor mass of the front wall of the urinary bladder

signs of infiltration of the musculature of the anterior abdominal wall measuring 75x80mm, which was seen as a change of bladder tumor (Fig. 1,2,3). On ureterocystoscopy the capacity of bladder easy decreased and on the front wall of the urinary bladder more over the left side of the suspicious bullous edema and external signs of infiltration of the wall of the bladder. Colonoscopically, except for internal hemorrhoidal plexus other findings were normal.

Endoscopic transurethral revealed on the front wall of the bladder solid tumor diameter 40x50x10mm entering the lumen of the bladder, and mucosa above the change is largely intact and marginally more toward the back wall bullous and cerebriform. It was taken biopsy of change which as microscopic showed the elements of proliferative lesion binders most by type of fibromatosis.

The patient underwent resection of pelvic tumors with partial cystectomy and Right Hand iliac lymphadenectomy. Intraoperatively, dilated small bowel diameter twice large compared to the usual size was seen and identified tumor of pelvic diameter 70x80mm, which invaded the anterior wall of the bladder, m.rectus abdominis from the

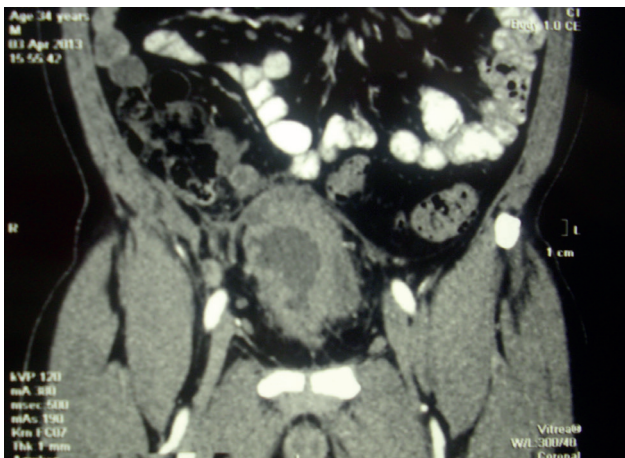


Figure 2. CT - tumor mass on the front wall of the urinary bladder

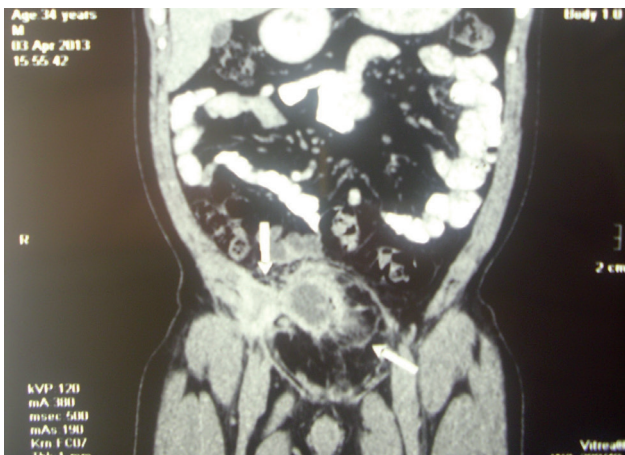


Figure 3. CT - infiltration of the musculature of the anterior abdominal wall



Figure 4. MRI of the abdomen and pelvis - no signs of recurrence of the underlying disease

right and parietal peritoneum anterior lateral abdominal wall, rest on the iliac vessels on the right hand and right ureter, but did not infiltrate them. Tumor engage omentum and convolutions of the ileum without involvement of the small and large intestines. Right iliac lymph nodes in ex tempore PH survey pointed to inflammation and necrosis. The operation lasted 120 min. It was accessed to tumor tissue with lower median laparotomy. In the postoperative period there came to subocclusion which was resolved with nasogastric suction 5 days with stimulation peristalsis neostigmine. Postoperatively, until the patient's discharge from the hospital, the patient was on parenteral antibiotic therapy as well as on the therapy with low molecular heparin. The patient was discharged on the twelfth postoperative day in good general condition.

A definitive histological diagnosis was retroperitoneal fibromatosis with purulent inflammation of local adipose tissue and chronic lymphadenitis.

Five months after the operative procedure the patient feels well and has no problems. In laboratory findings CRP: 2.85 mg / l, MRI of the abdomen and pelvis with no signs of recurrence of the underlying disease (Figure 4).

### 3. DISCUSSION

Although the majority of tumors are sporadic, approximately 8% of patients with AF have cases of colorectal cancer in the family history which is significantly higher than in the general population and suggests a genetic predisposition of both conditions (11).

Fibromatosis has locally infiltrative growth and a tendency of recurrence (12) and that in 39% to 79% of the cases (13). Tumor growth is not constant and may eventually be regressive, but in some periods and rapid too (14). In our view was intraoperatively incorporated infiltrative growth of tumor.

In relation to the anatomical localization we distinguish superficial and deep subtypes. Superficial changes are usually slow-growing changes, which rarely involve the deeper structures. Superficial fibromatosis includes the structures of the face and neck (Fibromatosis coli), palms (Dupuytren's contracture), feet (Ledderhose's disease), penis (Peyronie Mb.), shoulder, thigh, gluteal region and trunk. (15) On the other hand, the second the group includes changes of larger size with greater tendency of recurrence after treatment. This subgroup covers the muscles of the trunk and extremities. It is important to emphasize the significance of tumor size at initial surgical treatment, as in patients with a tumor smaller than 50 mm in diameter was better survival period of fifty years. The patient presented to our work falls into the category of high risk for recurrence, so he would therefore be strictly followed in the next five years. In the case of disease recurrence, the size of tumor does not affect the survival period (16).

Deep AF may occur at any soft tissue localization, but patients with familial AF frequently have tumor in mesentery of bowels, and with sporadic AF at their extremities, and the folds (17). Through medical history we were not able to confirm a positive family history in terms of

the existence of AF to our patient. The deep variant of AF includes abdominal wall, mesentery, retroperitoneum, mediastinum and abdominal cavity (18). Intra-abdominal fibromatosis as subtype of deep fibromatosis is often associated with familial adenomatous polyposis (19). To young women a deep AF occurs during or after pregnancy. Extra-abdominal fibromatosis derives from muscular connective tissue and of fascia close fitting.

The main therapeutic strategy is to prevent the invasion of surrounding tissues. The optimal time of surgical treatment is not defined.

Despite the well-known clinical picture of the disease, prognostic factors have not been clearly identified. The marginal status was not a significant prognostic factor for this tumor subtype (16), but despite the microscopic appearance of the surgical margins of about 75% of patients who underwent surgery for primary aggressive fibromatosis, are cured (16).

In previous studies that have examined the results of surgical treatment of extra-abdominal fibromatosis in 203 patients after primary resection of the ten-year survival was 76% and after resection of recurrent disease 59% for the same period (16).

Current reports indicate that in 5% of cases there was a spontaneous tumor regression (20), mainly in recurrent disease

Deep fibromatosis originates from mesenchymal tissue. Several authors have showcased extra-abdominal fibromatosis in patients with familial adenomatous polyposis (Gardner's syndrome) (21).

Findings on MDCT chest abdomen and pelvis showed that the fibromatosis originated of the muscles of the anterior abdominal wall.

Complete resection of the tumor is the therapeutic choice for both the primary tumor (22) and the recurrence (16).

Adjuvant therapies of NSAIL, with tamoxifen, anti-neoplastic, tyrosine kinase inhibitors, isolated limb perfusion, cryoablation with radiation and their combination have been mainly a therapeutic option for patients with unresectable tumors (10,23), and the effect of this therapy is not possible to assess the available literature. After incomplete resection, radiation therapy is a possible treatment option (24). There are researches on the possible use of preoperative radiation therapy for local control of the disease (25).

### 4. CONCLUSION

Early diagnosis of these tumors which are often asymptomatic is a key factor in the outcome of treatment, and wide resection of tumor is the best form of treatment which significantly reduces the risk of recurrence by the available literature.

• Conflict of interest: none declared.

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