Surgery for Sporadic Abdominal Desmoid Tumor: Is Low/No Recurrence an Achievable Goal?

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ABSTRACT: **Background:** Sporadic abdominal desmoid tumors are rare and data on these tumors as a distinct disease entity are lacking. Previous abdominal surgery, trauma, pregnancy and estrogen intake are considered risk factors. Although desmoids are benign, invasion and a high recurrence rate are common.

Objectives: To evaluate outcomes of surgery for this rare disease.

Methods: Since 1995, 16 patients with pathologically confirmed desmoid tumor were operated on in our center. All familial adenomatous polyposis patients were excluded. A retrospective analysis of data was performed.

Results: Of the 16 patients 12 (75%) were females. Mean age was 40.5 years (range 24–70). Thirteen patients were symptomatic and 3 were incidentally diagnosed. All patients presented with an isolated mass; 7 (50%) originated in the abdominal wall, 6 (37.5%) were retroperitoneal and 3 were (18.8%) mesenteric. All tumors except one were completely excised. Morbidity was low with no mortality. One patient was reoperated due to involved margins. None of the patients had recurrence within a median follow-up of 64 months (range 5–143).

Conclusions: The perception of sporadic abdominal desmoids as tumors with a high recurrence rate (20–70%) is probably incorrect. Adequate surgery with wide margins leads to a very low recurrence rate; cure is a legitimate goal.

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Desmoid is derived from the Greek word "desmos," which means band/tendon-like. These neoplasms are benign fibrous tissue tumors arising in the musculo-aponeurotic structures throughout the body. Desmoid tumors can be divided into five subgroups: extraabdominal, intraabdominal, multiple, multiple familial, and as part of Gardner's syndrome [1]. The association of desmoid tumors and familial adenomatous polyposis is well recognized. Desmoid tumors occur in 3.5–29% of patients with FAP, whereas the incidence of sporadic desmoids in the general population is very rare [2,3].

Surgeons encounter a miniscule number of these tumors during their career. These tumors occur primarily in the abdominal wall and extraabdominally, primarily in the trunk and extremities. These "benign" tumors are locally aggressive; they have no distinct capsule, and their margins are ill defined [3]. They can infiltrate surrounding structures, mainly muscles and fascial planes. Desmoids tend to recur locally, but they never metastasize [4]. Most of the reported cases were associated with previous trauma, prolonged estrogen intake, and pregnancy [5,6]. Abdominal desmoid tumor usually presents as a mass that is sometimes associated with pain and weight loss [7]. The treatment of choice is surgical removal with broad 'clean' margins in order to reduce the expected high local recurrence rate of 25–50% [8,9]. Often, removal of these tumors with safe negative margins requires resection of adjacent structures such as muscles, nerves, blood vessels and intraabdominal organs; involvement of vital structures may prevent complete resection. Due to the rarity of the disease there are no large series in the literature of sporadic abdominal desmoid tumors. In the largest reported series of desmoid tumors 77 were located in the trunk; 47 were intraabdominal, of which 7 were FAP related [10]. Data for this group were not reported as a separate disease entity, although their microenvironment and their sporadic nature may differentiate them from other desmoids. We present our experience with the surgical management and outcome of 16 patients diagnosed with sporadic abdominal desmoid tumor.

PATIENTS AND METHODS

We conducted a retrospective review of patients who underwent surgery for abdominal desmoid tumorin our facility in the last 10 years. Included were patients operated for a pathologically confirmed diagnosis of abdominal desmoid. All underwent a preoperative colonoscopy in order to rule out polyposis; known FAP patients were excluded. Patients were evaluated for gender, age, clinical presentation, site and size of the desmoid tumor, operative course, pathology and outcome. None of the patients in this

FAP = familial adenomatous polyposis

series received radiotherapy and/or systemic therapy. Recurrence was excluded by physical examination and abdominal computed tomography. To confirm the diagnosis and to verify the status of resection margins, histological slides were revised by a pathologist who was an expert in soft tissue sarcoma.

RESULTS

Between January 1995 and April 2007, 16 patients with sporadic abdominal desmoid tumor were identified – 12 women (75%) and 4 men (25%). Mean age at diagnosis was 40.5 years (range 24–70). Five of the 16 patients (31.3%) had a history of previous abdominal surgery: 3 cesarean sections, one appendectomy, and one right colectomy for benign adenomatous polyp. Three patients (18.8%) had a history of in vitro fertilization treatment before the diagnosis of desmoid tumor. Two patients were diagnosed during pregnancy and three others at the postpartum period. A summary of individual patient characteristics is depicted in Table 1.

Most patients, 13 of the 16 (81.3%), were symptomatic; six (37.5%) had abdominal pain, 7 (43.8%) were seeking clinical advice due to a palpable mass, and 2 (12.5%) described minor weight loss. Three patients (18.8%) were incidentally diagnosed: two during a routine gynecological examination and one on abdominal CT performed for evaluation of bladder cancer. All masses were palpable upon presentation. Table 2 depicts individual clinical presentation.

Abdominal CT with intravenous contrast was performed in all cases; the site of the desmoid tumor at the time of presentation was as follows: 7 (50%) originated from the rectus abdominis muscle, 6 (37.5%) were retroperitoneal, and 3 (18.8%) were mesenteric [Table 1]. All patients had sizable tumors; the mean tumor diameter was 14 cm (range 5–30) [Figure 1]. Preoperative core needle biopsies were taken

Table 1. Individual patient characteristics of 16 patients with sporadic abdominal tumors

Patient no.	Age (yrs)	Origin	Tumor size	Risk factors
1	36	Mesentery	7	Cesarean section
2	27	Retroperitoneum	30	IVF
3	66	Retroperitoneum	15	–
4	70	Abdominal wall	5	Right colectomy
5	34	Retroperitoneum	18	IVF, pregnancy
6	38	Abdominal wall	10	–
7	42	Mesentery	5	–
8	24	Retroperitoneum	15	–
9	49	Abdominal wall	8	Appendectomy
10	28	Abdominal wall	5	Pregnancy, cesarean section
11	31	Abdominal wall	10	Pregnancy
12	39	Abdominal wall	16	Pregnancy
13	56	Mesentery	30	–
14	28	Abdominal wall	8	Cesarean section
15	28	Retroperitoneum	15	IVF, pregnancy
16	42	Retroperitoneum	17.5	–

Table 2. Signs and symptoms at initial presentation*

	No. of patients
Abdominal mass	7
Abdominal pain	6
Weight loss	2
Incidental finding at CT scan	1
Incidental finding on routine gynecological examination	2
Early satiety, dysphagia	1

*Patients may have had more than one sign and symptom at presentation

Figure 1. CT images of desmoid tumors. **[A]** Abdominal wall desmoid. **[B]** Retroperitoneal desmoid tumor. A celiac trunk is embedded within the tumor.

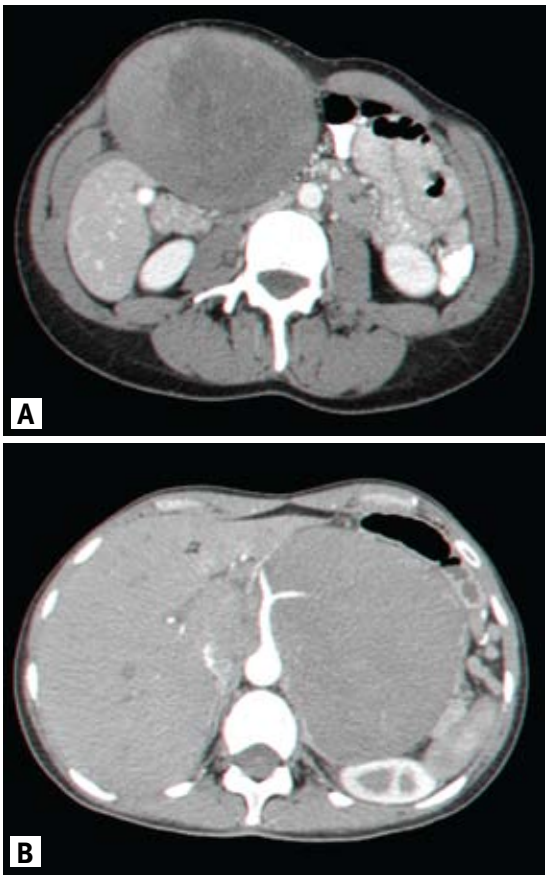
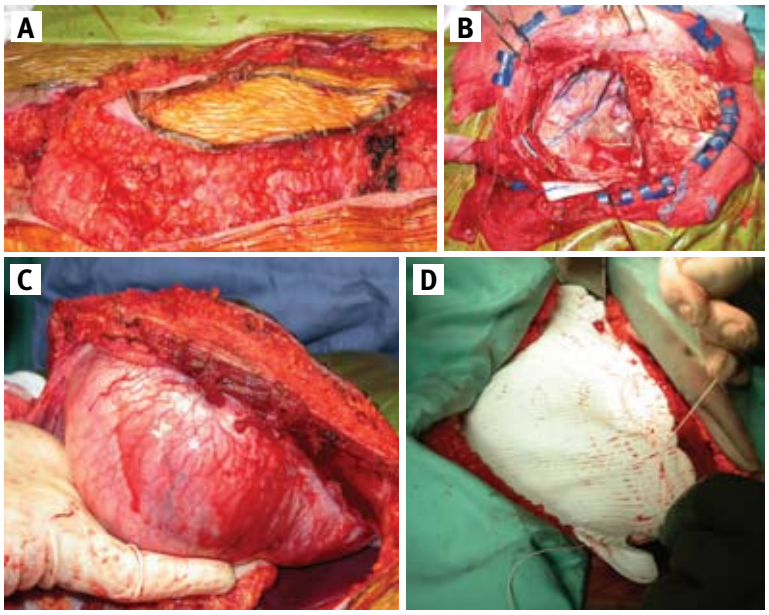


Figure 2. Intraoperative pictures of surgery for abdominal wall desmoid tumor. **[A]** Abdominal wall with tumor. **[B]** Tumor excised with all the layers of the abdominal wall. **[C]** Large abdominal wall defect after resection of tumor. **[D]** Abdominal wall after Gortex mesh repair.



in 5 of the 16 patients (31.3%). Of these, three were diagnosed as desmoids, one as a questionable myxoma, and one showed elongated spindle cells suspicious for malignancy. Three patients (18.8%) had undergone previous surgery with curative intent in another hospital; two of them were considered inoperable. All 16 patients were operated on in an attempt to achieve cure; seven abdominal wall desmoids were widely resected including all the layers of the abdominal wall followed by a Gortex mesh reconstruction [Figure 2]. Three mesenteric desmoids were resected with an adjacent small bowel loop. Retroperitoneal desmoids were radically excised with adjacent structures that were involved; left upper abdominal exenteration (including resection of the stomach, body and tail of pancreas, spleen and left kidney) was performed in one patient. The celiac trunk was embedded within the tumor [Figure 1B] and was sacrificed; this was possibly due to a replaced right hepatic artery and backflow perfusion from the gastroduodenal artery.

One patient was referred for a third attempt to resect a huge left retroperitoneal desmoid after two operations with failure to achieve complete resection. During these previous interventions nephrectomy was performed due to ureteral encasement. Eventually her desmoid was completely resected, with a partial resection of pelvic floor muscles, left colon, femoral nerve and the left iliac artery and vein. A vascular femorofemoral bypass graft was required for vascular reconstruction.

There was no mortality, and complications occurred in 5 patients (31.3%); one (7%) had a major complication – anastomotic leak from a colocolostomy requiring a diverting colostomy that was reversed 2 months later. Wound infection occurred in two patients with mesh reconstruction of the abdominal wall; they were treated with antibiotics and recovered without further intervention. All desmoids but one, which originated in the rectus abdominis, were completely excised with clear histological margins.

None of our patients received complementary radiation therapy. No recurrence occurred in any of the patients during a median follow-up of 64 months (range 5–143). Fifteen of the 16 patients (93.8%) are alive and disease free. One patient, who never had a recurrence of his disease, died from an acute myocardial infarction 6 years after his operation. The one patient who was reoperated shortly after the first operation due to residual disease has been disease free for 5 years. Three patients gave birth 2 and 4 years after recovery, with no evidence of a new or recurrent desmoid. No patient was diagnosed with FAP during the follow-up period.

DISCUSSION

Abdominal desmoid tumors are rare, and most develop as an extracolonic manifestation of FAP. Sporadic abdominal desmoid tumors are extremely rare, the vast majority arising in the abdominal wall. Sporadic presentation mandates a thorough gastrointestinal tract study to rule out FAP. The basic underlying cause for desmoid tumor formation is a defect in connective tissue formation [11]. Histologically, mature fibroblasts of uniform size and shape are seen; mitoses are unusual [3]. Although defined as benign, these are aggressive tumors with a high propensity for local invasion and recurrence, yet they do not metastasize [4].

Most of the reported series are in the adult population; according to these data previous abdominal operation, prolonged estrogen intake, pregnancy, and trauma are described as risk factors for sporadic abdominal desmoid [5,6,12,13]. Reitamo et al. [12] observed an interesting association between age and the occurrence of desmoid tumors. Four age groups demonstrate different varieties of desmoid tumors: the juvenile desmoid tumor is an extraabdominal tumor found in young girls; the fertile variant is an abdominal tumor found in women; the middle age is also overwhelmingly abdominal but the sex ratio approaches equality; whereas in the old age group, both abdominal and extraabdominal tumors are equally frequent and the sex ratio is equal [12]. In this report, fertile women aged 22.8–31.6 displayed the highest susceptibility to desmoids. In our cohort 12 patients (75%) were women, 11 of them in the fertile age (range 24–38). The relatively high prevalence of desmoids among fertile women implies that sex hormones possibly play an important role in the development

and growth of these tumors. This premise is strongly supported by the higher incidence of desmoids among pregnant and IVF-treated women. Reitamo and colleagues [12] showed a correlation between tumor growth rate and estrogen levels; the slowest rate was in young girls, increasing from menarche and declining after menopause. In contrast, growth rate was similar in all male patients unrelated to age. In the older female age group a low growth rate was observed, equal to that of male patients. Similarly, in our group of 11 fertile women 5 desmoids were diagnosed during pregnancy or shortly after labor; 2 women were exposed to high estrogen levels throughout their fertility treatment. Interestingly, three women in our series gave birth within 5 years from desmoid resection with no evidence of recurrence or new primary tumor.

The differential diagnosis of retroperitoneal desmoid tumors includes lymphoma, germ cell tumors, high grade sarcoma, endometriosis in fertile women, and other rare benign tumors; therefore, a preoperative biopsy may be needed to select appropriate therapy in select cases. Although numerous reports [14,15] have shown that a core needle biopsy is safe and effective, there is no consensus regarding its utilization in the diagnostic process of retroperitoneal soft tissue tumors [14]. Given the potential for transperitoneal spread and track implantation, a biopsy taken from a retroperitoneal location may be more complex in comparison to the extremity or abdominal wall locations where it is relatively easier to resect the tract during surgery. In this series, preoperative core needle biopsy was performed in all patients who presented with abdominal wall tumors as well as in younger patients diagnosed with retroperitoneal soft tissue tumors in order to exclude the above mentioned differential diagnoses.

Surgery is the treatment of choice for sporadic abdominal desmoid tumors. Wide radical local excision is mandatory [16,17]. Complete resection is often challenging, especially with vascular and neural involvement. In contrast to retroperitoneal location, abdominal wall desmoids can be managed quite easily by radical resection with safety margins of at least 2–3 cm. Frequently, a sizable defect in the abdominal wall is formed mandating mesh reconstruction [Figure 2]. Intraabdominal and retroperitoneal desmoids require a meticulous preoperative evaluation including careful assessment of vascular, neural and/or other vital structure involvement.

In spite of complete macroscopic resection, desmoid tumors are notoriously known for their high recurrence rate. Previous data show that 70–90% of resected tumors recur within 2 years after surgery [18,19]; a lower rate of 25–68% is reported for extraabdominal tumors [12,20], approximately 40% for abdominal desmoids, and as high as 77% for mesenteric desmoids [8]. Mariani et al. [21] reported a recurrence rate of 43% (median follow-up 23 months) in a series of 7 patients with pelvic desmoid tumors. In two recent series recurrence rates of intraabdominal desmoids in FAP patients were 57% and 85% respectively [22,23]. Abdominal wall desmoids have a significantly lower recurrence rate. In a series of 7 anterior abdominal wall desmoids, Sutton and Thomas [24] found no recurrence. Shao et al. [25] reported a recurrence rate of 5.5% in a large series in China of 42 abdominal wall desmoids. The current series further demonstrates that a very low recurrence rate is achievable and can be expected after adequate surgery without adjuvant therapy. Comparing our data to the literature, this relatively low recurrence rate was most prominent in the group of nine patients who had retroperitoneal and mesenteric desmoids. This may be attributed to two possible factors: The first is that we adopted an aggressive approach according to 'sarcoma surgery principles' – namely, wide clear margins as well as exenteration and sacrifice of vital structures when needed. The second is that this unique series included only sporadic abdominal desmoids excluding FAP and extraabdominal sporadic tumors; these tumors may have a different biological behavior and therefore lower recurrence rates.

CONCLUSIONS

Our series illustrates that although rare, sporadic abdominal desmoids tend to occur in fertile women, mostly in the abdominal wall. Since the role of adjunctive drugs or radiotherapy has yet to be defined for these uncommon tumors, surgery is the only curative treatment. An aggressive surgical approach may enable cure and/or a very low recurrence rate.

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