Original Article

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En bloc resection for intra-abdominal/retroperitoneal desmoid-type fibromatosis with adjacent organ involvement: A case series and literature review

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Summary **Surgical treatment for intra-abdominal/retroperitoneal desmoid-type fibromatosis (IA/RPDF) is still controversial. Studies regarding en bloc resection in IA/RPDF with adjacent organ involvement are scanty. This study aims to evaluate the safety and effectiveness of en bloc resection in IA/RPDF with adjacent organ involvement. This retrospective clinical study included 21 patients who were diagnosed with IA/RPDF and underwent tumor resection at a single center between March 2013 and June 2018. All patients included in the study underwent surgery with curative intent, and IA/RPDF with adhesive organs was removed en bloc. The safety of surgical treatment was verified by the analysis of intraoperative bleeding, postoperative morbidity and perioperative mortality. The efficacy of surgical treatment was evaluated based on the status of tumor infiltration of adjacent organs and patient follow-up results. Complete macroscopic (R0 or R1) resection was achieved in all cases. A median of 2 (range, 1-7) organs were resected. The median operating time was 300 (90-650) minutes. The median intraoperative bleeding was 300 (20-4,500) milliliters. For postoperative pathological diagnosis at our center, tumor infiltrated at least one organ in each patient. Infiltration was noted in 45 resected organs (45/57, 78.9%). Grade III-V postoperative morbidity developed in one patient (4.8%). During the follow-up, one patient developed local recurrence. No DF-related death was noted during the follow-up. The 3-year disease-free survival rate was 94.1% (95% confidence interval: 83.6-100%). Therefore, en bloc resection of the tumor and involved adjacent organs is a safe and effective treatment modality for IA/RPDF.**

Keywords: Desmoid-type fibromatosis, retroperitoneum, surgery

1. Introduction

Desmoid-type fibromatosis (DF) is a rare monoclonal, fibroblastic proliferation characterized by locally infiltrative but rarely metastatic lesions (*1,2*). The local recurrence rate is high and varies between 15% and 77% (*3,4*). DF can occur in many locations, the most common being the extremities and abdominal wall. It can also occur in the abdominal cavity and retroperitoneum. Desmoid tumors have been reported to occur in 7.5% to 16% of patients with familial adenomatous polyposis (FAP) and the relative risk of developing desmoid tumors is much higher in patients with FAP than in the general population (*5,6*). FAP-related tumors more commonly arise in the intra-abdominal region (*7*).

The consensus for treatment of DF has changed over the past decade, with most centers moving away from primary radical surgery toward a front-line "watchful waiting" policy (*1,2,8-11*). However, most of the results of previous studies were based on the analysis of extra-abdominal DF (*1,2,8-11*). There are limited studies on intra-abdominal/retroperitoneal DF (IA/RPDF), and most previous studies were case reports. The effectiveness of surgical treatment for IA/RPDF remains controversial. Given the lower risk of complications and recurrence rates than extremity DF

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following surgical resection, some investigators have suggested that there should be a low threshold for surgery in the treatment of intra-abdominal DF (*2,12*). However, other investigators have recommended that surgical treatment should be chosen with caution according to the changing consensus regarding extra-abdominal DF therapy (*1,13*).

Several studies show that margin status does not affect recurrence and has no prognostic significance in desmoid tumors (*14-16*). However, some large retrospective studies have reported higher recurrence rates for patients with positive margins when compared to negative margins (*15,17,18*). Considering that DF is characteristically locally infiltrative, *en bloc* resection in IA/RPDF with adjacent organ involvement may help reduce relapse and improve prognosis. Studies regarding *en bloc* resection in IA/RPDF with adjacent organ involvement are scanty.

Hence, to assess the safety and efficacy of *en bloc* resection in IA/RPDF with adjacent organ involvement, we conducted a retrospective case series analysis and literature review.

2. Materials and Methods

2.1. *Data source and patient selection*

A retrospective clinical study was conducted, with data retrieved on 21 consecutive patients with IA/RPDF who were treated at the Peking University Cancer Hospital Sarcoma Center between March 2013 and June 2018. All patients included in the study underwent surgery with curative intent, and IA/RPDF with adhesive organs were removed *en bloc*. The median follow-up period was 24 (range, 4-68) months. The ethics committee of Peking University Cancer Hospital and Institute approved the study.

2.2. *Pathological diagnosis*

All resected tumors were delivered to the Department of Pathology after the operation. Overall tumor size was defined as the sum of the perpendicular maximum diameters. All margins were perpendicularly sampled, with two or more sections taken from all margins. Additional sections were taken from the closest margin. Serial sampling of all resected organs and surrounding fat was performed. Two sarcoma pathologists independently diagnosed the tumor pathology.

2.3. *Definitions*

Different from the DF located in extremities and abdominal wall, it is difficult to achieve a reliable assessment of the margin status of IA/RPDF (*19*). Thus, surgical resection was described as macroscopically complete (R0 or R1) or incomplete (R2) (*19*). Postoperative complications were graded according to the Clavien-Dindo classification (*20*). Postoperative pancreatic fistulae (POPF) were defined according to the International Study Group on Pancreatic Fistula definition (*21*).

2.4. *Follow-up*

At our center, patients were prospectively followed up via clinical examination, chest radiography, and abdominopelvic computed tomography or magnetic resonance imaging every 3 months for the first year, every 6 months for the subsequent 4 years, and yearly thereafter.

2.5. *Statistical analysis*

Data extracted from the database, computerized hospital notes, and pathology records were analyzed. Data are presented as median and range or number and percentage, as appropriate. We analyzed the local disease-free survival (DFS) from the date of operation to the date of last follow-up. Statistical analyses were performed using SPSS version 24.0 (IBM Corp., Armonk, NY, USA) and R version 3.4.0 (*http://www.r-project.org*).

3. Results

3.1. *Patient characteristics*

Data from 21 patients, comprising 11 male (52.4%) and 10 female (47.6%), were retrospectively analyzed in our study. The median age was 35 (range, 21-73) years. DF was located in the abdominal cavities of 6 (28.6%) patients, and in the retroperitoneum of 15 (71.4%) patients. Seventeen (81.0%) patients received primary surgical treatment, while 4 patients underwent surgery after tumor recurrence. Meanwhile, DF in 3 patients (14.3%) was considered to be related to FAP due to a history of colon polyposis. All the patients achieved macroscopically complete resection (R0 and R1). The clinicopathologic characteristics of all the patients are displayed in Table 1.

3.2. *Details of surgery*

All patients included in the study underwent surgery with curative intent, and the surgical policy was to remove tumors with adhesive organs *en bloc* (Figure 1). Complete macroscopic (R0 or R1) resection was achieved in all cases. The median number of resected organs was 2 (range, 1-7). The median operating time was 300 (90-650) minutes. The median intraoperative bleeding was 300 (20-4,500) milliliters. The tumor of the patient with the largest amount of intraoperative bleeding was located in the pelvic cavity, and the

amount of bleeding in the presacral venous plexus was large. Except in this patient, the maximum volume of intraoperative blood lost was 1,000 milliliters. The details of operating time and intraoperative bleeding are displayed in Table 2.

The small intestine, including the duodenum, was the organ most commonly resected (14/21, 66.7%), followed by the colon (12/21, 57.1%). Of the 4 patients (4/21, 19.0%) who underwent nephrectomy, 1 had a tumor that invaded the kidney and 3 had a tumor that encapsulated the proximal ureter for a long length, making it difficult to reconstruct the ureters. Two patients (2/21, 9.5%) underwent distal pancreatectomy, and 1 patient (1/21, 4.8%) underwent partial resection of the uncinate process. Three patients (3/21, 14.3%) underwent resection of the iliac vessels and artificial vessels replacement because the vessels adhered to the resected tumor; among them, 1 patient underwent resection of the iliac artery and vein, while the remaining 2 patients underwent resection of the iliac vein alone. The details of resected organs are displayed in Table 2 and Table 3.

Table 1. Patient characteristics

Characteristics	*n* (%)
Age, (years; median [range])	35 (21–73)
Sex	
Male	11 (52.4)
Female	10 (47.6)
Presentation	
Primary	17 (81.0)
Recurrent	4 (19.0)
Tumor site	
Abdomen	6 (28.6)
Retroperitoneum	15 (71.4)
Number of tumors	
Single	17 (81.0)
Multiple	4 (19.0)
Tumor size	
≤ 10 cm	11 (52.4)
> 10 cm	10 (47.6)
FAP-related desmoid-type fibromatosis	
Yes	3 (14.3)
No	18 (85.7)
Number of organs resected [median (range)]	2 (1–7)
Resection margins	
Macroscopically complete	21(100.0)
Macroscopically incomplete	0 (0)

FAP, familial adenomatous polyposis.

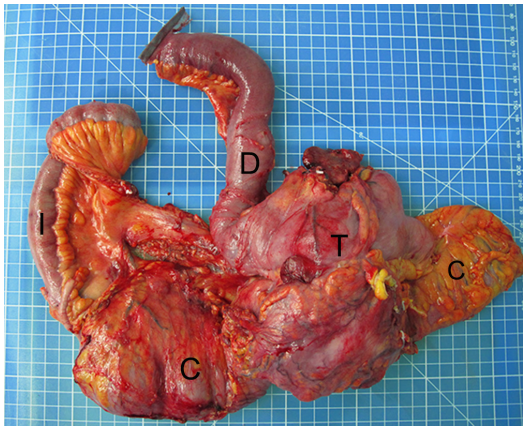

Figure 1. The resected specimen of *en bloc* resection of intra-abdominal/retroperitoneal desmoid-type fibromatosis and involved adjacent organs. The tumor and adjacent organs were resected *en bloc*. The tumor invaded the duodenum and colon. **T**, tumor; **C**, colon; **D**, duodenum; **I**, ileum.

Table 2. Details of resected organs, intraoperative bleeding, duration of operating time, infiltrated organs and postoperative morbidities of patients

Items	Resected organs	Intraoperative bleeding (mL)	Duration of operating time (minutes)	Infiltrated organs	Post-operative morbidities
Case 1	Colon, ovary, kidney, AG, ureter, IA, IV	1,000	650	IA, IV, ureter, colon	Gastroparesis
Case 2	SI	300	135	SI	
Case 3	Colon, ureter, SI	200	300	Colon, ureter, SI	Grade A POPF
Case 4	Colon, pancreas, spleen	500	300	Colon, pancreas	
Case 5	Pancreas, stomach, spleen	50	210	Pancreas, stomach, spleen	
Case 6	Colon, SI, ureter	100	190	SI, ureter	
Case 7	SI	800	180	SI	
Case 8	Colon, SI	1,000	210	Colon, SI	
Case 9	SI	100	90	SI	
Case 10	SI	50	480	SI	
Case 11	Colon, stomach	50	300	Stomach	Hemorrhage
Case 12	Pancreas, SI	50	480	Pancreas, SI	
Case 13	SI	900	180	SI	Gastroparesis
Case 14	Colon, SI	200	210	Colon, SI	Incomplete intestinal obstruction
Case 15	Colon, SI	50	150	Colon, SI	
Case 16	SI, ureter, kidney	800	210	SI, ureter	
Case 17	Kidney, AG, ureter, ovary, AW, colon, IV	20	600	Kidney, ovary, AW, colon	
Case 18	Colon, kidney, ureter	300	330	Colon, ureter	
Case 19	Colon, SI, AW	800	460	Colon, SI, AW	
Case 20	Rectus, uterus, ovary, ureter, IV	4,500	630	rectum, ureter, IV	
Case 21	Colon, SI	1,000	380	Colon, SI	

AG, adrenal gland; AW, abdominal wall; IA, iliac artery; IV, iliac vein; POPF, postoperative pancreatic fistula; SI, small intestine.

3.3. *Infiltration details*

All tumors were diagnosed as DF, and tumor infiltrated at least one organ in each patient. The median number of infiltrated organs was 2 (range, 1-5) in each patient. Infiltration was noted in 45 resected organs (45/57, 78.9%). The tumors commonly infiltrated the small intestine, and all the resected small intestines were infiltrated by DF while some of the DF lesions even infiltrated the submucosae of the small intestines (Figure 2A). DF infiltrated all the resected abdominal walls, stomachs, rectums, pancreata (Figure 2B), and iliac vessels (Figure 2C). Among 4 patients who underwent nephrectomy, only 1 (1/4, 25%) kidney infiltration by DF, and the remaining 3 had tumor infiltration of the ureters. The details of infiltration are displayed in Table 2 and Table 3.

3.4. *Postoperative morbidity and follow-up*

Grade II postoperative morbidity developed in 4 patients (19.0%); 2 of them experienced gastroparesis, 1 experienced incomplete intestinal obstruction, and 1 experienced grade A POPF. Grade III postoperative morbidity developed in 1 patient (4.8%). This patient experienced postoperative hemorrhage and required reoperation. No patient died of surgery.

During the follow-up, 1 patient developed local recurrence in the 15th month after surgery, and the tumor progressed despite non-steroidal anti-inflammatory drug treatment. Thus, the patient underwent secondary surgery, and no recurrence was noted during follow-up (20 months). The 3-year DFS rate was 94.1% (95% confidence interval: 83.6-100%). No DF-related death was noted during the follow-up. The patients' functional ability was scored according to the Barthel Index (*22*), with all patients scoring 100.

Table 3. Resection and infiltration details of intra-abdominal/retroperitoneal desmoid-type fibromatosis at our center

Items	Resected number	Infiltrated number (%)
Small intestine	14	14 (100)
Colon	12	10 (83.3)
Ureter	7	6 (85.7)
Vessel	4	4 (100)
Kidney	4	1 (25)
Pancreas	3	3 (100)
Ovary	3	1 (33.3)
Stomach	2	2 (100)
Abdominal wall	2	2 (100)
Spleen	2	1 (50)
Adrenal gland	2	0 (0)
Rectum	1	1 (100)
Uterus	1	0 (0)
Total	57	45 (78.9)

FAP, familial adenomatous polyposis.

4. Discussion

The consensus for treatment of DF has changed over the past decade, with most centers moving away from primary radical surgery toward a front-line "watchful waiting" policy (*1,2,8-11*). However, most of the results of previous studies were based on the analysis of extra-abdominal DF (*1,2,8-11*). There are limited studies on IA/RPDF, and most previous studies were case reports (Table 4) (*23-42*). A case series of IA/RPDF treated with *en bloc* resection of the tumor with adhesive organs is reported for the first time in this study. Complete macroscopic resection was achieved in all cases.

Although the "watchful waiting" policy was recommended as the first-line management for extra-abdominal DF, it is a controversial one for IA/RPDF (*2,12,43*). Several previous studies have reported tumor location as a risk factor for recurrence after surgery, and a lower rate of recurrence for intra-abdominal DF than for tumors located in the extremities (*1,18*). Lev *et al.* reported that the most common sites of local recurrence were the extremities (16/57 [28%]), superficial trunk (7/71 [10%]), and viscera (4/47 [9%]) (*18*). The response rate to medical therapy varies from 15-60% (*44-47*), and grade 3-4 toxicities occurred in approximately 13-43% of patients (*46,48-50*). Considering the uncertain effect of medical therapy and the occurrence of adverse effects, there is insufficient evidence that drug therapy is better than surgery in the treatment of IA/RPDF. Furthermore, for patients with symptoms, awaiting spontaneous regression or drug onset is intolerable. Therefore, there should be a low threshold for surgery in the treatment

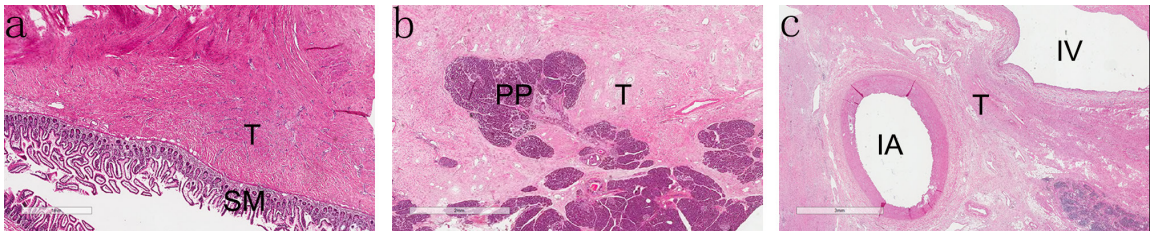

Figure 2. Intra-abdominal/retroperitoneal desmoid-type fibromatosis infiltrating the adjacent organs. (A) Desmoid-type fibromatosis infiltration of the submucosa of the small intestine. **(B)** Desmoid-type fibromatosis infiltration of the pancreatic parenchyma. **(C)** Desmoid-type fibromatosis infiltration of the iliac vessels. **T**, tumor; **SM**, submucosa of the small intestine; **PP**, pancreatic parenchyma; **IA**, iliac artery; **IV**, iliac vein.

Table 4. Summary of 23 cases of intra-abdominal/retroperitoneal desmoid-type fibromatosis published in the PubMed database between 2006 and 2017

Authors	Age	Sex	Site	Size (cm)	Resected organs	Complication	Recurrence	Follow up time (months)
Yong W. 2013 (24)	31	Male	Abdomen	10.0	None	No	No	/
Deepak V. 2010 (25)	46	Female	Abdomen	6.2	colon	No	No	3
Koichi T. 2006 (34)	73	Female	Abdomen	6.3	Small intestine	No	No	48
B. Kreuzberg 2007 (26)	48	Female	Abdomen	/	Small intestine, colon, uterus	No	No	/
B. Kreuzberg 2007 (26)	60	Female	Retroperitoneum	/	None	No	Yes	24
B. Kreuzberg 2007 (26)	43	Male	Retroperitoneum	/	None	No	No	/
Mohammad K. 2010 (33)	37	Male	Abdomen	6.0	None	No	No	8
Jae Young C. 2010 (36)	46	Female	Retroperitoneum	5.5	Colon, ureter	No	No	/
Christos N. 2010 (42)	65	Male	Abdomen	12.0	Small intestine	Bowel ischemia	No	/
Coskun P. 2010 (27)	57	Male	Abdomen	10.0	Small intestine, colon	No	No	/
Bouhabl S. 2011 (41)	71	Male	Abdomen	19.0	Small intestine	No	No	12
Sung Hoon J. 2009 (40)	49	Female	Abdomen	7.5	Colon, ovary, oviduct	No	No	/
Liang-Yu S. 2012 (39)	56	Male	Retroperitoneum	3.6	None	No	No	6
Mohammed K. 2012 (28)	47	Male	Abdominal	28.0	Small intestine	No	No	6
Marek W. 2010 (38)	44	Female	Abdominal	10.1	Small intestine	No	No	/
Cemli C. 2011 (37)	35	Female	Abdominal	6.0	Colon	No	No	/
Kinyanjui J. 2012 (32)	41	Male	Abdomen	/	Small intestine	No	No	/
LH Tan 2010 (35)	58	Female	Abdomen	4.5	Small intestine	No	No	6
Menegazzo M. 2013 (23)	35	Female	Abdomen	34.0	Small intestine	No	No	25
Norihito O. 2013 (30)	45	Male	Abdomen	9.0	Pancreas, spleen	No	No	27
Norihito O. 2013 (30)	74	Female	Abdomen	6.0	Small intestine	Bowel obstruction	No	7
Mari M. 2017 (31)	45	Female	Abdomen	5.5	Pancreas, spleen, stomach	No	No	/
Hirotoshi K. 2014 (29)	55	Male	Abdomen	1.2	Small intestine	No	No	/

of intra-abdominal DF, particularly for patients with symptoms that are difficult to endure (*8*).

For postoperative pathological diagnosis, DF infiltrated at least one organ in each patient. Infiltration was noted in 78.9% resected organs. Some of the IA/RPDF lesions even infiltrated the submucosa of the small intestine. All these results indicate that IA/RPDF has a high infiltration tendency, such that it frequently infiltrates organs. Although margin status remains a controversial topic in the management of desmoid tumors, it is often agreed that R0 resections are ideal (*13*). Considering the high infiltration tendency of IA/RPDF, to reduce the local recurrence, achieving negative surgical margins by *en bloc* resection of the tumor and adjacent organs is recommended.

In our present study, grade III-V postoperative morbidity occurred in 1 of the patients, and only 1 patient developed local recurrence during the follow-up. The 3-year DFS rate was 94.1%. No DF-related death was noted during the follow-up. Among the 23 patients previously reviewed, grade III-V postoperative morbidity occurred in only 1 of the patients, and no patient developed local recurrence and died during the follow-up (Table 4) (*23-42*). Based on the above results, *en bloc* resection in IA/RPDF with adjacent organ involvement is a safe and effective treatment modality. However, the varied anatomical locations of the DF within abdominal cavity and retroperitoneum result in local invasion to different adjacent organs, and there is no standard surgical approach or procedure for IA/RPDF treating. We have reported details of anterior approach to *en bloc* resection in left sided retroperitoneal sarcoma with adjacent organ involvement, which also can be applied in left sided retroperitoneal DF (*51*).

Most DF are sporadic, but the incidence of DF associated with FAP has been reported to be 7.5 to 16% (*5,6*). FAP-related tumors more commonly arise in the intra-abdominal region (*7*). FAP-related DF has a high recurrence rate after surgical resection (*12,52*). However, in our study, no recurrence occurred in the patient with FAP-related DF during follow-up, suggesting that *en bloc* resection in IA/RPDF with adjacent organ can effectively control the tumor. This was confirmed by a 10-year review of the management of FAP-associated DF (*53*). However, larger-scale, prospective observational studies with longer follow-up periods are needed to validate the most appropriate treatments for IA/RPDF and FAP-associated DF.

This study has certain limitations. Although it was a case series of IA/RPDF in patients who underwent *en bloc* resection of the tumor and adjacent organs, because of its low incidence, the sample size was relatively small, which might influence the accuracy of our results. Further multicenter and larger sample studies are needed to provide more reliable results.

In conclusion, according to the case series study and literature review, *en bloc* resection of the tumor and adjacent organ involvement is a safe and effective treatment modality for IA/RPDF.

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