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Clinicopathological characteristics, treatment, and survival outcomes of retroperitoneal desmoid-type fibromatosis

A single-institution experience in China

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Abstract

Retroperitoneal desmoid-type fibromatosis (RPDF) is a rare mesenchymal neoplasm, and it covers a broad spectrum of aggressive monoclonal, fibroblastic proliferation. There is no evidence-based or established optimal treatment available for this intriguing disease yet. Therefore, we here investigated the clinicopathological characteristics, surgical, and survival outcomes in RPDF among Chinese patients.

Patients with histologically confirmed RPDF were retrospectively studied from 2010 to 2018 within the West China Hospital of Sichuan University. Demographics, clinicopathological characteristics, treatment, and survival outcome data were collected.

Of the 29 cases of RPDF, 19 were females. Tumor diameter ranged from 4 to 40 cm, with a median of 10 cm. Of these patients, surgical resection was the primary treatment adopted for RPDF in 26 cases; while 3 patients underwent watchful waiting. In surgical group, complete and incomplete macroscopic resection was achieved in 21 (80.77%) and 6 (19.23%) cases, respectively. Totally, 21 (80.77%) cases underwent multi-visceral resection. With a median follow-up duration of 48 months, 11 patients experienced tumor progression for the entire cohort. Tumor progression was observed for those patients with incomplete and complete macroscopic resection in 2/5 (40.0%) and 6/21 (28.6%) cases, respectively. In the watchful waiting group, there were no documented cases of RPDF regression. The progression-free survival rate was 86.1%, 71.5%, and 62.3% at 1-, 2-, and 3-years, respectively.

RPDFs are rare types of tumor, which have characteristically varied natural histories. Surgical resection had a relative favorable outcome, but some patients were associated with burden of significant surgical complications.

Abbreviations: CNB = core needle biopsy, CT = computed tomography, DF = desmoid-type fibromatosis, FAP = familial adenomatous polyposis, MRI = magnetic resonance imaging, NSAIDs = nonsteroidal anti-inflammatory drugs, PFS = progression-free survival, SPSS = statistical package for the social science, RPDF = retroperitoneal desmoid-type fibromatosis, TKI = tyrosine kinase inhibitors.

Keywords: aggressive fibromatosis, desmoid, outcome, retroperitoneum, surgery

1. Introduction

Desmoid-type fibromatosis (DF) is a subtype of mesenchymal neoplasia, which covers a broad spectrum of benign fibrous tissue proliferation.[1] The term desmoid was first described by Mueller in 1838, based on the morphologically bland mayo-fibroblastic cells which make up the desmoid tumor. DF is a distinct rare entity that accounts for 0.03% of the tumors and 3% of all soft-tissue tumors with an incidence of 5 to 6 per million of the population per annum.[2–4] It can occur as a superficial or deep form, with a tendency towards recurrence, but an inability to

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metastasize.[5] The local recurrence rate is high and varies from 18% to 56%.[6,7] DF may occur in many locations, such as the extremities and abdominal wall, but the solitary occurrence is rare in retroperitoneal space.[8]

Due to the rarity of the disease and a considerable variation in the natural history of DF, there are few evidence-based consensuses available for this disease. Historically, immediate surgery was favored by many scholars with reported local control rates of up to 80% at 5 years. More recent long-term researches have demonstrated that many DFs remain stable or even regress (20%–30% of cases) with observation.[9] Moreover, with a development of the understanding of DF's natural history, an approach of initial watchful waiting has been advocated especially for asymptomatic tumors.[6] Of note, clinical management varies based on tumor location and size. In 2017, an updated guideline published by the European desmoid working group supports that medical therapy should be the first therapeutic option for retroperitoneal desmoid-type fibromatosis (RPDF).[10] In case of further progression, surgery, radiotherapy, or medical therapy would be an option, but with a tendency toward surgery if acceptable. However, data on RPDF are scanty yet, and a majority of previous studies were case reports.[11–13] Moreover, the effectiveness of surgical and feasibility of watchful waiting for RPDF also remain unknown.

In the present study, we; therefore, investigated the clinicopathological features, diagnosis, treatment, and survival outcomes of these patients based on data obtained from 29 consecutive RPDF patients in our institution.

2. Materials and methods

2.1. Patient selection and data source

The study was approved by the Institutional Review Board of West China Hospital. Operation consents were obtained from each patient in this cohort. All patients within our institution were pathologically diagnosed with RPDF (primary/recurrent) between August 2010 and November 2018 were identified. The type and timing of the treatment were recorded. Surgical specimens were examined and stained immunohistochemically for β-catenin, CD117, CD34, DOG-1, S-100, desmin, and smooth muscle actin (SMA). Computed tomography (CT) and/or magnetic resonance imaging (MRI) scan of the chest and abdomen, electronic gastrointestinal endoscopy, renal and liver function, and so on. were routinely performed preoperatively. All data were abstracted from the electronic medical chart, including age, gender, tumor site, size, clinical manifestation, surgical information, and prior surgical history were carefully reviewed.

2.2. Surgical treatment and watchful waiting

Patients with RPDF underwent surgical treatment with curative intent. In the case of rapid tumor progression or if RPDF threatens life function, surgery was performed by an anterior or posterior approach. CT and MRI scan were preoperatively performed at predicting tumor resectability, which allowed tumor location, size, and infiltration to be determined. Multivisceral resection was performed for those who invaded adjacent organs, but function preservation should also be an essential goal. Finally, the surgery was classified into 2 categories: macroscopically incomplete (R2) and macroscopically complete (R0/R1). Short/long-term postoperative complications were recorded. A total of 3 patients managed with a conservative "watchful waiting" approach, who were asymptomatic or denied surgery and medical therapy. The diagnosis of RPDF was established on core needle biopsy (CNB) guided by CT for those patients. The radiologic re-evaluation was done every 3 to 6 months, and response evaluation criteria in solid tumors score were also recorded. Patients were deemed as progression if so determined by the radiological evaluation or by the clinician based on worsening symptoms.

2.3. Follow-up and statistical analysis

Follow-ups were conducted through office visit, telephone calls, or outpatient clinic visits from May 2019 to June 2019. Abdominopelvic CT/MRI was performed every 3 to 6 months for the first year, every 6 months up to the fifth year. Progression-free survival (PFS) refers to the duration from the start of any treatment until disease progression. Calculations statistical analysis was performed with the Statistical Package for the Social Science (SPSS) version 21.0 for Windows (SPSS Inc, Chicago, IL). Measurement data were expressed as mean ± standard deviation, and enumeration data were described as a percentage. Cumulative survival was determined using the Kaplan–Meier method. Due to the limited sample size, predictors of progression by the multivariate analyses were thus not attempted.

3. Results

3.1. Demographic and clinicopathological characteristics

Until November 2018, a consecutive series of 29 patients with RPDF at our institution were retrospectively enrolled. The clinicopathological data are summarized in Table 1. This entire cohort comprised 10 (34.48%) males and 19 (65.52%) females, with a female-to-male ratio of 1.9. The ages of the patients ranged

Table 1

Demographic and clinicopathological characteristics of RPDF (n = 29).

Parameters	N (%)
Gender	
Male	10 (34.48)
Female	19 (65.52)
Age (yr; median [range])	26 (2–65)
Clinical symptoms	
Abdominal pain/discomfort	7 (24.14)
Accidental discovery	6 (20.69)
Swelling of lower extremities	3 (10.34)
Mass	8 (27.59)
Others*	5 (17.24)
Previous retroperitoneal surgery	
Yes	7 (24.14)
No	22 (75.86)
Primary/recurrent RPDF	27 (93.10)/2 (6.90)
Tumor size (cm; median [range])	10 (4∼40)
Treatment options	
Surgical resection	26 (89.66)
Watchful waiting	3 (10.34)
Hospital stay (days; mean ± SD)	22.69 ± 15.38
FAP-related RPDF	2 (6.90)

FAP = familial adenomatous polyposis, RPDF = retroperitoneal desmoid-type fibromatosis, SD = standard deviation.

* Including pain in the inguinal area, dysuria, lumbar pain/discomfort.

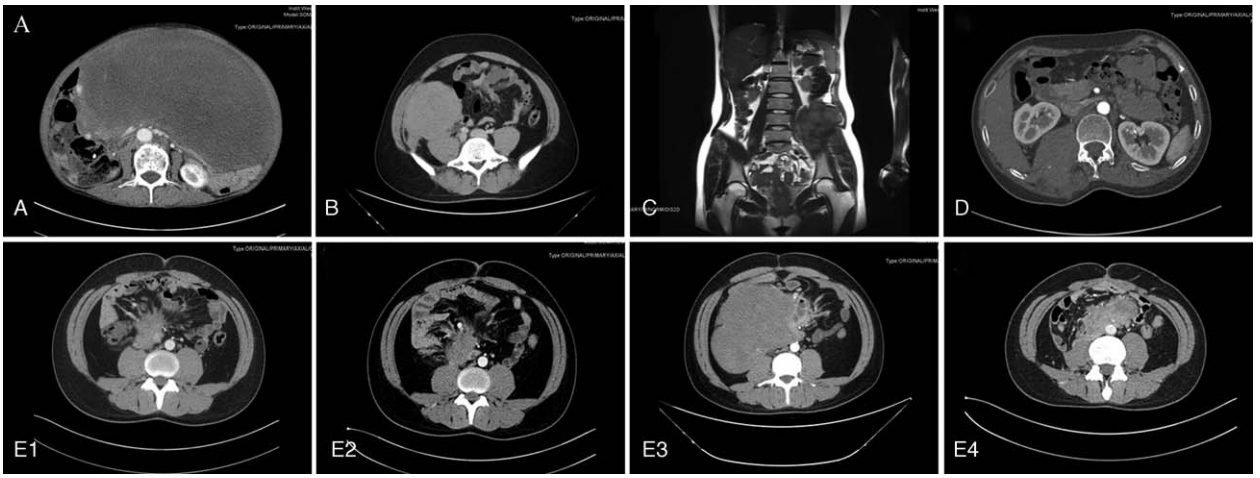

Figure 1. (A) Giant cystic and solid mass (40*30 cm) originating from the retroperitoneal space. Liquefaction necrosis occurred, and compression of adjacent organs can be seen. (B) Soft tissue mass located in the right lower abdomen and pelvic retroperitoneal region, and infiltrate the right psoas, iliac muscles, and ureter. (C) MRI showing that the tumor located in the left retroperitoneum with a size of 7.6*9.0 cm. (D) The tumor compressing the right kidney. (E1) Tumor with 7.0*3.0 cm at the initial diagnosis (September 2011). (E2) No progression was observed (October 2012). (E3) Rapid growth of tumors was observed after 27 months (December 2013). And then surgical resection was performed. (E4) Stable disease was noted until now. MRI = magnetic resonance imaging.

from 2 to 65 years at initial diagnosis (median: 26 years). In patients reporting main symptoms upon initial presentation, 8 patients exhibited mass, 7 patients with abdominal pain and discomfort, 6 cases were incidentally discovered. Tumor diameter ranged from 4 to 40 cm, with a median of 10 cm. In total, there were 7 (24.14%) patients who had previously underwent retroperitoneal surgery. Of these patients, surgical resection was the primary treatment adopted for RPDF in 26 cases, whilst 3 patients underwent watchful waiting. In the present study, 2 patients had a concurrent diagnosis of familial adenomatous polyposis (FAP).

3.2. Detailed outcomes of different treatment strategies

A total of 26 patients were treated by immediate surgical resection due to tumor size, choice of doctors and patients, and aggravation of related symptoms. Eleven patients had a perioperative blood transfusion. Complete and incomplete macroscopic resection was achieved in 21 (80.77%) and 6 (19.23%) cases, respectively. There was 1 patient who received celecoxib postoperatively. In total, 21 (80.77%) cases underwent multi-visceral resection. Infiltration was noted in 24/26 (92.31%) patients. The median number of infiltrated organs was 2 (0~5) in each patient, and the median number of resected organs was 1.5 (0~4). Of note, the RPDF commonly infiltrated the retroperitoneal vessels (34.62%) and small intestine (34.62%), followed by colon, bladder, and others (psoas, diaphragm, ureter, liver, pancreas, kidney and the abdominal wall; Fig. 1). Ten patients experienced postoperative complications, such as retroperitoneal abscess ($n=2$), wound dehiscence ($n=1$), wound infection ($n=2$), intestinal obstruction ($n=2$), nerve injury ($n=1$), ureteral fistula ($n=1$) and hydronephrosis ($n=1$). No operation-related death occurred in this cohort. The details can be seen in Table 2. Three patients underwent watchful waiting. However, all 3 patients experienced tumor progression. One of them eventually underwent surgical resection (R2), which was combined with partial duodenectomy, right hemicolectomy and right nephrectomy (Fig. 1E1–E4).

3.3. Pathological and immunohistochemical characteristics

All excised or biopsy specimens were examined by pathology. Histologically, it is characterized by a uniform spindle cell proliferation with a moderate amount of collagen fibers; which are usually arranged in a wavy interlaced arrangement (Fig. 2). No pathological mitosis was noted in all cases. β-catenin staining showed that cytoplasmic reaction was diffuse, and the nuclear

Table 2

Surgery information and postoperative complications (n=26).

Parameters	N (%)
Perioperative blood transfusion	
Yes	11 (42.31%)
No	15 (57.69)
Resection margins	
Macroscopically complete	21 (80.77)
Macroscopically incomplete	5 (19.23)
Multivisceral resection	
Yes	21 (80.77)
No	5 (19.23)
No. of organs infiltrated (median [range])	2 (0–5)
No. of organs resected (median [range])	1.5 (0–4)
Resected organs	
Vessel	9 (34.62)
Small intestine	9 (34.62)
Colon	5 (19.23)
Bladder	3 (11.54)
Others*	14 (53.85)
Postoperative complications	
Retroperitoneal abscess	2 (7.69)
Wound dehiscence	1 (3.85)
Wound infection	2 (7.69)
Intestinal obstruction	2 (7.69)
Nerve injury	1 (3.85)
Ureteral fistula/hydronephrosis	2 (7.69)

* Including pancreas, spleen, ureter, vagina, and ilium.

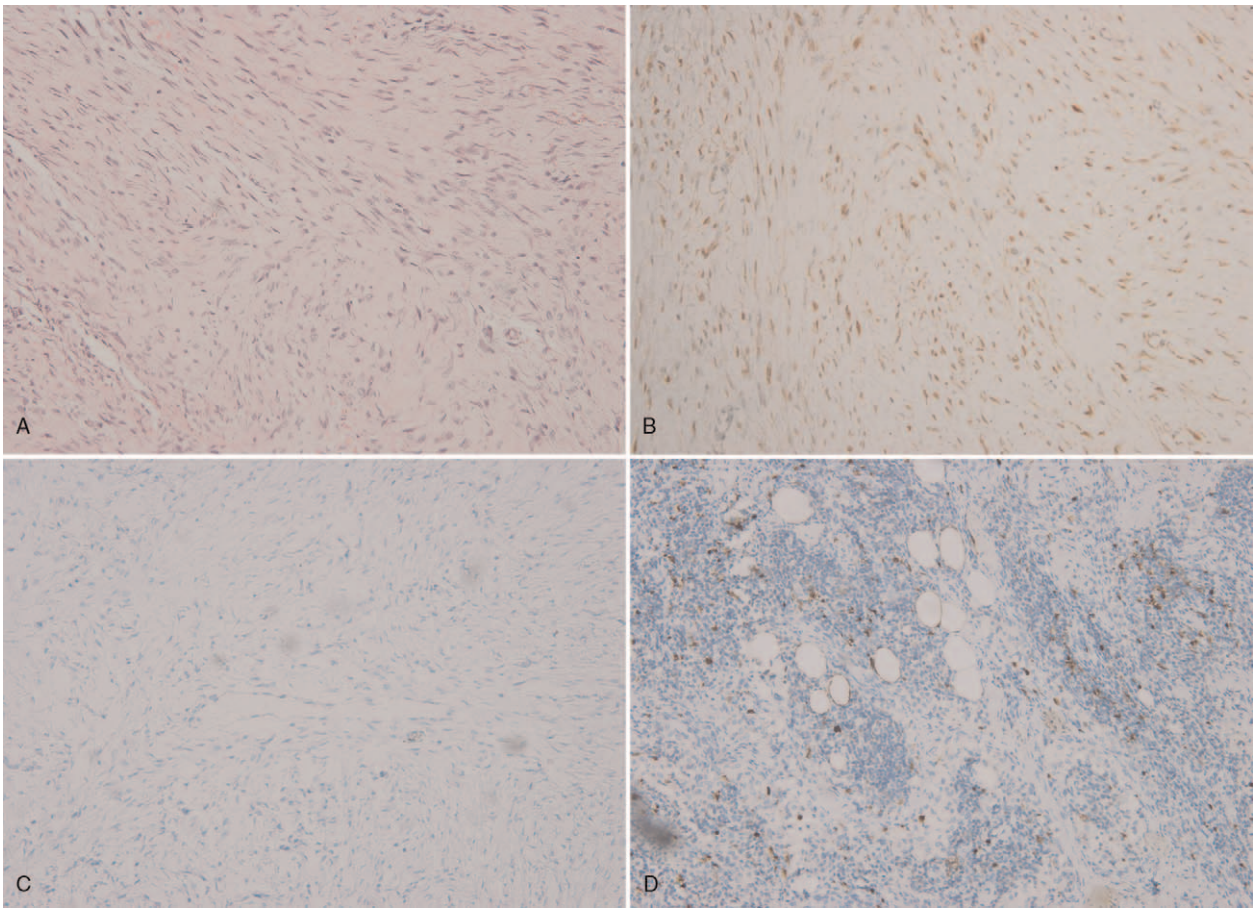

Figure 2. (A) The tumor composed of uniform spindled proliferation (hematoxylin-eosin staining, ×200). (B) β-catenin-positive staining in most of the nuclei (×200). (C) Negative desmin staining (×200). (D) S-100-negative staining (×200).

reaction was observed in about 50% of the cells, whilst CD117, DOG-1, CD34, S-100, desmin, and SMA staining was negative.

3.4. Survival outcomes

With a median follow-up duration of 48 months (range: 7-106 months), 11 patients experienced tumor progression for the entire cohort. The PFS rate was 86.1%, 71.5% and 62.3% at 1-, 2- and 3-years, respectively (Fig. 3). No RPDF-related death was observed by the end of follow-up. During the follow-up, 8/26 patients who have had immediate surgery experienced tumor progression, and 3/8 cases finally underwent secondary surgery. Tumor progression was noted for those patients with incomplete and complete macroscopic resection in 2/5 (40.0%) and 6/21 (28.6%) cases, respectively. In the watchful waiting group, there were no documented cases of DF regression; Of which, surgery was performed after progression in 1 case, but recurrence occurred 35 months postoperatively; whilst 2 continued observation without further progression after further observation. In total, tumor-bearing and tumor-free patients were observed in 13 and 16 patients, respectively, before the conclusion of the study.

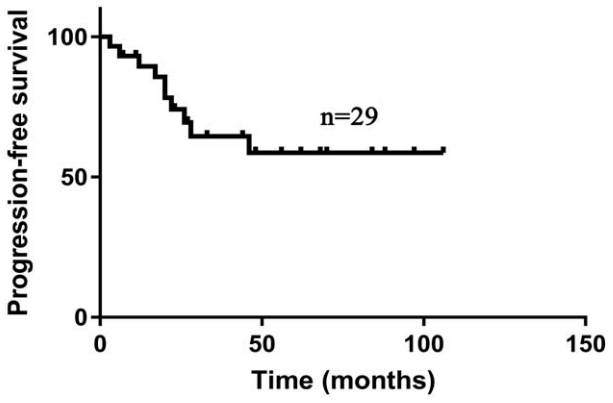

Figure 3. Kaplan–Meier progression-free survival for patients with RPDF (n = 29). RPDF = retroperitoneal desmoid-type fibromatosis.

4. Discussion

According to the World Health Organization, DF is defined as an intermediate soft-tissue tumor characterized by monoclonal fibroblastic proliferation with a variable and unpredictable clinical course. But limited availability of data specifically targeting patients with RPDF is described in clinical trials, and most investigations consist of case series based on relatively small numbers of patients. As such, more research on such tumors is needed. This retrospective study describes the clinicopathology, treatment, and outcomes of RPDF with a relatively large sample in a single institution. DF occurs mainly between the age of

15 and 60 years, with a peak age of 30 to 40 years.[10] The present series gives similar results, which was diagnosed with a median age of 26 years. Sporadic DF predominantly affects young females and is more than twice in female than male patients,[14] which is in concert with our finding. However, there was no gender preference in older patients.

The aetiopathogenesis of the DF is not yet clear, but it is reported to be associated with trauma (including prior surgery), long-term use of estrogen, pregnancy/puerperium and also related with abnormalities of Wnt signaling mediated by the APC/β-catenin pathway[15]; the latter explains the association of DF with FAP (also called Gardner syndrome). Approximately 5% to 10% DF arises in the context of FAP, and is predominantly located within the abdomen.[8] In our series, FAP-related RPDF occurred in 2 (6.90%) male patients, which was in accordance with the previous result.[16,17] Intra-abdominal DF patients may present in multiple ways, such as pain, mass, bowel perforation, hematochezia, or symptoms caused by tumor compression.[18,19] Of note, 20.69% of cases were incidentally discovered in the present study. Furthermore, DF sometimes may mimic cancer recurrence following surgery.[20]

Given the rarity of this disease, DF diagnosis is often hampered by misdiagnosis; the misdiagnosis rate can be as high as 30% to 40%.[4,21] Thus, the histopathologic confirmation of DF is mandatory before any treatments. A diagnosis of DF can be readily established on CNBs in most cases; an excisional or incisional biopsy is not needed. In the present study, a total of 3 cases who underwent watchful waiting were confirmed by biopsy. Imaging can be used to guide clinicians to locate lesions safely and facilitate the success of biopsy; CT is normally used for deep biopsies, while ultrasound can be used for superficial lesions. Clinically, the diagnosis of DF is not always simple. Histological diagnosis often needs to rule out other probable mesenchymal tumors such as gastrointestinal stromal tumors, or a low-grade leiomyosarcoma.[22] Immunohistochemical staining with CD117, CD34, S-100, and desmin is useful in the differential diagnosis. Noteworthy, nuclear accumulation of β-catenin on immunostaining has been usually expressed in DF.

Historically, complete resection is the mainstay treatment for DF. However, the increasing literature supports that a policy of active surveillance has been shown to lead to spontaneous regression in extra-abdominal wall DF.[6,9,23] It is; therefore, reasonable to consider watchful waiting as an initial step in all asymptomatic patients with extra-abdominal DF in non-life threatening locations.[24] As such, asymptomatic DF should be carefully watched without active management, as suggested by the European Society for Medical Oncology (ESMO) guideline.[25] But there is also a concern that delaying treatment might risk the loss of a therapeutic window.[26] Watchful waiting policy is still controversial for RPDF.[27] More corresponding clinical trials should be carried out in the future. Our data have shown that all 3 patients with active observation experienced progression, and surgery was performed in 1; whilst 2 continued observation due to stable disease observed later. Nowadays, there is insufficient evidence yet to suggest that medical therapy/watchful waiting is better than surgery in the treatment of this type of disease. Consequently, surgery should be offered for a large sporadic RPDF due to tumor size and possible related symptoms.[10] In our series, the majority of patients have a huge tumor with evident clinical symptoms. Intra-abdominal DF/RPDF has a high tendency to infiltrate the adjacent organs,[13] and the same phenomenon was observed in the present study. Therefore, en-bloc resection of the tumor is becoming significantly more challenging. Nevertheless, negative surgical margins should be the goal, but should not be pursued at the expense of significant function loss.

There is a large body of literature that supports that both medical therapy and radiotherapy can be considered as single-agent regimens in case of patients with symptomatic/progressive DF.[8] However, intra-abdominal DFs are not candidates for radiotherapy because of the risks of side effects. Systemic therapy comprises hormone therapy, nonsteroidal anti-inflammatory drugs (NSAIDs), chemotherapy, and tyrosine kinase inhibitors (TKI).[28] It is worth noting that successful tamoxifen use in DF was published over 20 years. Although Anti-hormonal agents such as tamoxifen, toremifene, and progesterone can be used in both males and females, response rates have been found to be low.[10] Moreover, Janinis et al in 2003 reported that a partial response in 48% and stable disease in 28% was observed for DF treated by NSAIDs.[29] Therefore, combining hormone therapies and NSAIDs as first medical treatment was recommended by the guideline,[8] which was mainly due to their limited toxicity, rare adverse effects, and effectiveness. Patients treated by NSAIDs in the present study are scarce, and robust conclusions on this question so cannot be formed. The role of chemotherapy in DF is controversial because of the absence of malignant cells and metastatic potential. Based on the identification of c-kit and platelet-derived growth factor receptors in DF tissue, TKI was introduced to treat DF.[24] The exact benefit offered by TKI is not known due to thin literature. No prospective data available yet, TKI should not be routinely used outside of clinical trial.

Several factors have been reported as prognostic indications, such as age, tumor location, and size.[14,30,31] The abdominal wall DF portending a better prognosis, followed by intra-abdominal, and extremity DF had a higher risk of progression. In our series, the PFS rate of RPDF was 86.1%, 71.5%, and 62.3% at 1-, 2-, and 3-years, respectively, which are lower than that of Mullen results.[32] It may attribute to the fact that those patients had a huge tumor in the present study. Additionally, the exact tumor sizes and ages, which may result in a significant increase in the risk of progression, are unknown yet. Incomplete excisions of DF have been linked with post-operative progression for many years,[33] whilst the literature to date is divided on this question.[30] Recently, a population-based study has reported that the microscopic margin had no effect on recurrence (14% of margin negative vs 20% of margin positive, $P=1.0$).[11] Furthermore, the growing body of evidence supports that β-catenin mutational status is correlated with DF recurrence, and S45F mutation may be a clinically useful prognostic factor.

5. Conclusions

In summary, the surgical resection had a relatively favorable outcome for RPDF, but some patients were associated with significant surgical complications burden. Moreover, RPDFs are rare kinds of tumors, which have a characteristically varied natural history. The chief potential criticism of this series is the relatively small number of patients, which prevented us from drawing any strong conclusions. Thus, multicenter researches are warranted and urgently needed in the near future.

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