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Original Article

Results of multimodal treatment for desmoplastic small round cell tumor of the abdomen and pelvis

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Abstract: Purpose: Desmoplastic small round cell tumor (DSRCT) is a rare aggressive malignancy that occurs in a young population with a male predominance. We studied the clinical and pathological characteristics of DSRCT and investigated the effects of multimodal therapy including aggressive surgical resection, induction chemotherapy, and external beam radiotherapy. Methods: We retrospectively reviewed and analyzed our experience with 11 histologically proven cases of DSRCT between March 2004 and October 2014. The clinical information, histological, immunohistochemistry and survival data of the patients were collected. Results: The median age at diagnosis was 31.4 years (range, 14-64 years) and nine (82%) of the patients were males. The most common presenting complaint was abdominal pain (72.7%). Surgical resection was attempted in five patients and included macroscopic total resection in two patients and debulking in three patients. Six patients underwent biopsy only. Eleven patients received multiagent chemotherapy. Five patients (45.5%) received radiotherapy. The median survival of patients who underwent surgical resection was 34.5 months, whereas the patients who underwent biopsy alone was 24.5 months ($P<0.05$). The median survival was 40.8 months in radiotherapy group, and 19.2 months in non-radiotherapy group ($P<0.05$). The 3-year progression-free survival rate was 27.2%. The median survival was 29 months, and the median time to local failure was 8.8 months. Cox regression analysis showed surgery and radiotherapy were highly significant in prolonging patients survival. Conclusion: Multimodal therapy consists of combination of surgical resection, chemotherapy and radiotherapy results in improved survival in patients with DSRCT. For unresectable DSRCT, we recommend radiotherapy combined with anthracycline-based chemotherapy.

Keywords: Desmoplastic small round cell tumor, surgery, radiotherapy, prognosis

Introduction

Desmoplastic small round cell tumor (DSRCT) is a rare and highly aggressive disease, which mainly affects adolescents and young adults [1]. The clinical presentation is most often marked by a large abdominal and/or pelvic mass with peritoneal seeding. Less frequently, these tumors can be found in the thoracic cavity, intracranially, and hands. The histogenic origin of these tumors is unclear, it has been suggested that it might arise from the primitive mesothelium or submesothelial mesenchyme [2]. Organ involvement is capricious and secondary, liver and lung are two common sites for metastatic disease beyond the peritoneum [3]. Immunohistochemical staining shows nests of small round cells within desmoplastic stroma with no glycogen deposit in the cytoplasm. Genetic expression observed constantly in DSRCT reveals a particular chromosomal translocation t (11; 22) (p13; q12) and its assayable fusion transcript of the Ewing sarcoma (EWS) and Wilms tumor (WT1) genes has assisted diagnosis [4].

Nowadays, the treatment strategies for this tumor include intensive multi-agent chemotherapy, aggressive debulking surgery (>90% resection), adjuvant abdominal pelvic radiation, and myeloablative chemotherapy with stem cell rescue. Despite multimodal managements, optimal treatment strategies remain disputable and the prognosis is poor. Because most of our cognitive regarding the pathologic and clinical nature of this disease has been based on case reports and small series of patients, the development of standard methods for its diagnosis

Table 1. Clinical findings of the 11 cases of DSRCT

Clinical findings	Positive	Positive rate (%)
Abdominal pain	8/11	72.7%
Abdominal distension	6/11	54.6%
Weight loss	4/11	36.4%
Diarrhea	3/11	27.3%
Constipation	1/11	9.1%
Hematuria	1/11	9.1%
Itchy skin	1/11	9.1%

and management has been complicated. Consequently, to our knowledge there is no clear consensus regarding the optimal therapeutic modalities for treating these highly aggressive disease. Herein, we presented our experience in the diagnosis and management of this disease at our institution, moreover, whether surgery and radiotherapy were closely related to the prognosis were analyzed.

Materials and methods

After approval from the institutional review board, a retrospective review was conducted of the records of 11 DSRCT patients treated at Shandong Cancer Hospital and Institute between March 2004 and October 2014. Tissue diagnosis was determined by immunohistochemistry (IHC) in all patients. The data collected included age, sex, clinical presentation, radiologic findings, pathologic findings, type of surgery performed, postoperative course, adjuvant chemotherapy, radiotherapy, and follow-up.

Macroscopic total resection was defined as surgical resection of all macroscopically visible disease, debulking was defined as removal of >90% of the tumor burden but with macroscopically residual intraabdominal disease, and biopsy was defined as the removal of tissue for diagnosis only. Because of multiple lesions or diffuse nature of tumors infiltrating adjacent vital organs, it is often impossible to complete resection of all tumors with negative microscopic margins. Subsequently, we suggested all cases received adjuvant chemotherapy. Response to chemotherapy based on the degree of tumor volume reduction was denied as follows: complete response (CR), partial response (PR), stable disease (SD), and progressive disease (PD).

The actuarial survival rates were calculated from the initiation of treatment to the date of death or last follow-up using the Kaplan-Meier method, and compared between groups by use of log-rank test. A multivariate analysis was performed using Cox model. *P* value <0.05 was considered to indicate statistical significance; all statistical tests were carried out utilizing SPSS, version 17.

Results

Patient characteristics, clinical presentation, and diagnosis

The median age at diagnosis was 31.4 years (range, 14-64 years) and 9 of the patients were males (81.8%). All patients were symptomatic at the time of presentation with a mean duration of symptoms of 2 months (range, from 5 days to 5 months). The most frequent symptom was abdominal pain, usually cramping in nature (72.7%). Abdominal distension, weight loss, diarrhea, constipation, itchy skin, and hematuria were less common symptoms at presentation (**Table 1**).

All cases underwent abdominal and pelvic CT examination with contrast-enhanced CT scan. The most common imaging finding were multiple nodular peritoneal soft-tissue masses with variable sizes in abdominal and pelvic space (8/11, 73%), retroperitoneal and mesenteric lymphadenopathy (7/11, 67%), and hepatic lesions (4/11, 36%). Tumor size varied from 1.0 to 20.0 cm in maximum dimension with 36.4% more than 10 cm in maximum dimension, 36.4% between 5-10 cm and 27.3% less than 5 cm. Distant metastasis was observed in 54.5% (6/11) of these patients at diagnosis and local involvement was observed in 72.7% (8/11). The most common site of metastasis was liver (5/11). Metastasis was also found in the lungs (2/11).

Once the diagnosis of DSRCT is considered, a diagnostic biopsy should be carried out. In some cases a suspicious mass can be completely removed, but more frequently the tumor was so extensive that a biopsy is indicated. Fine needle aspiration cytology has been used correlated with histological findings from open biopsies and may be helpful to confirm recurrence. In addition to standard histopathology, cytogenetics and immunohistochemical studies also help confirm the diagnosis.

Table 2. The IHC analysis of various markers in the present series of DSRCT

Markers	Positive	Positive rate (%)
Cytokeratin (CK)	9/11	81.8%
Epithelial membrane antigen (EMA)	9/11	81.8%
Desmin	8/11	72.7%
Vimentin	8/11	72.7%
Neuronspecific enolase (NSE)	6/11	54.5%
CD99	1/6	16.7%
Synaptophysin	2/7	28.6%
Chromogranin A	4/9	44.4%
SMA	6/8	75%
HBME1	2/4	50%
NF	1/4	25.0%
CD20	0/2	0%
HMB45	0/4	0%
Calretinin	0/2	0%

Tumor pathology and immunohistochemical analysis

Histopathologically, the characteristic findings are well-defined nests or clusters of undifferentiated small round cells, surrounded by a prominent desmoplastic stroma. The tumor cells are characterized by small hyperchromic nuclei with scanty cytoplasm. The results of IHC staining of 11 patients were summarized in **Table 2**. The tumor cells showed positive for cytokeratins (CK) (9/11, 81.8%), epithelial membrane antigen (EMA) (9/11, 81.8%), desmin (8/11, 72.7%), vimentin (8/11, 72.7%), neuron-specific enolase (NSE) (6/11, 54.5%). In addition, the tumors of all patients were negative for calretinin, human melanoma antibody (HMB45) and CD20. Molecular evidence of t (11; 22) (p13; q12), the defining cytogenetic abnormality of DSRCT, was certified in 4 patients of our cases. The 4 patients were confirmed with positive molecular results by fluorescent in situ hybridization (FISH).

Treatment setting

In five patients, a surgical resection was performed (**Table 3**). Primary macroscopic total resection of disease was accomplished in 2 of these patients, and the remaining 3 patients underwent major (>90%) debulking of their intraabdominal disease. Surgical resections were incomplete because of extensive peritoneal attachments and vessel involvement. The resection percentage was determined by review of operative notes and comparative imaging. Attempts were not made to achieve microscopic negative margins. Omentectomy, stripping of the parietal and visceral peritoneum, splenectomy for hilar involvement, and local resection of the diaphragm were often required. Two patients received preoperative chemotherapy with ifosfamide, doxorubicin, and cisplatin (IAP) for four cycles and one patient received a combination of doxorubicin, cyclophosphamide and cisplatin (CAP) for three cycles, which resulted in partial tumor regression in two patients and a minor response in the one patient who received the latter regimen. The three patients then underwent surgical resection (macroscopic total resection in 1 patient and debulking in 2 patients) followed by adjuvant chemotherapy plus EBRT (5000 cGy) to the abdomen and pelvis in 1 patient and postoperative chemotherapy alone in 2 patients. Two other patients underwent primary surgical excision (macroscopic resection in one patients and debulking in one patient) as initial treatment. One patient who received CAP chemotherapy for four cycles also received 4500 cGy of preoperative external beam radiation therapy (EBRT). One patient refused to receive any kind of chemotherapy in the courses of the diseases.

Six patients underwent biopsy only, because their intraabdominal disease was too extensive for primary surgical resection (**Table 3**). All 6 patients began treatment with multiagent chemotherapy, which involved a combination of doxorubicin, ifosfamide, and cisplatin (IAP) in 2 patients for four-six cycles; 3 patients were treated with regimens of concurrent doxorubicin, cyclophosphamide and cisplatin with irradiation using conventional fractions (200 cGy) followed by three cycles of adjuvant CAP chemotherapy. The conventional technique was used for administration of a total dose of 5000 to 6000 cGy. Etoposide, cisplatin, and doxorubicin (EAP) were applied in 1 patient for six cycles.

After 3 to 4 cycles of chemotherapy, response was assessed by computed tomography scan. Three patients had a good response with marked diminution of the tumor. Due to the stromal composition of these tumors, a minority showed little response to chemotherapy. In

Table 3. Patient and disease characteristics and treatments in DSRCT

No.	Age/Sex	Tumor localization	Treatment	Follow-up (Months)	Results
1	14/M	Lower abdominal/pelvic mass, peritoneal and omental implants	MTR	21	Death
2	52/M	Lower abdominal/pelvic mass	Debulking surgery + RCT (CAP)	25	Death
3	30/M	Lower abdominal/pelvic mass; Peritoneal, and omental implants	IAP + MTR + IAP	16	Death
4	25/F	Pelvic mass, peritoneal and serosal implants	IAP + debulking surgery + IAP	38	Death
5	23/M	Lower abdominal/pelvic mass; Peritoneal, serosal, and omental implants	CAP + debulking surgery + RCT (CAP)	72	Survival
6	38/M	Lower abdominal/pelvic mass, peritoneal and omental implants	Biopsy + RCT (CAP)	24	Death
7	26/M	Lower abdominal/retroperitoneal and pelvic mass; Periaortic lymphadenopathy	Biopsy + EAP	11	Death
8	28/M	Peritoneal implants, liver metastasis	Biopsy + RCT (CAP)	40	Survival
9	64/F	Pelvic mass; Liver metastasis	Biopsy + IAP	9	Death
10	24/M	Pelvic mass; Mesenteric, retroperitoneal, and pelvic lymphadenopathy	Biopsy + RCT (CAP)	43	Death
11	21/M	Lower abdominal/pelvic mass; Peritoneal implants	Biopsy + IAP	20	Death

MTR: macroscopic total resection; IAP: ifosfamide, doxorubicin, and cisplatin; CAP: cyclophosphamide, doxorubicin, and cisplatin; EAP: etoposide, doxorubicin, and cisplatin; RCT: radiochemotherapy.

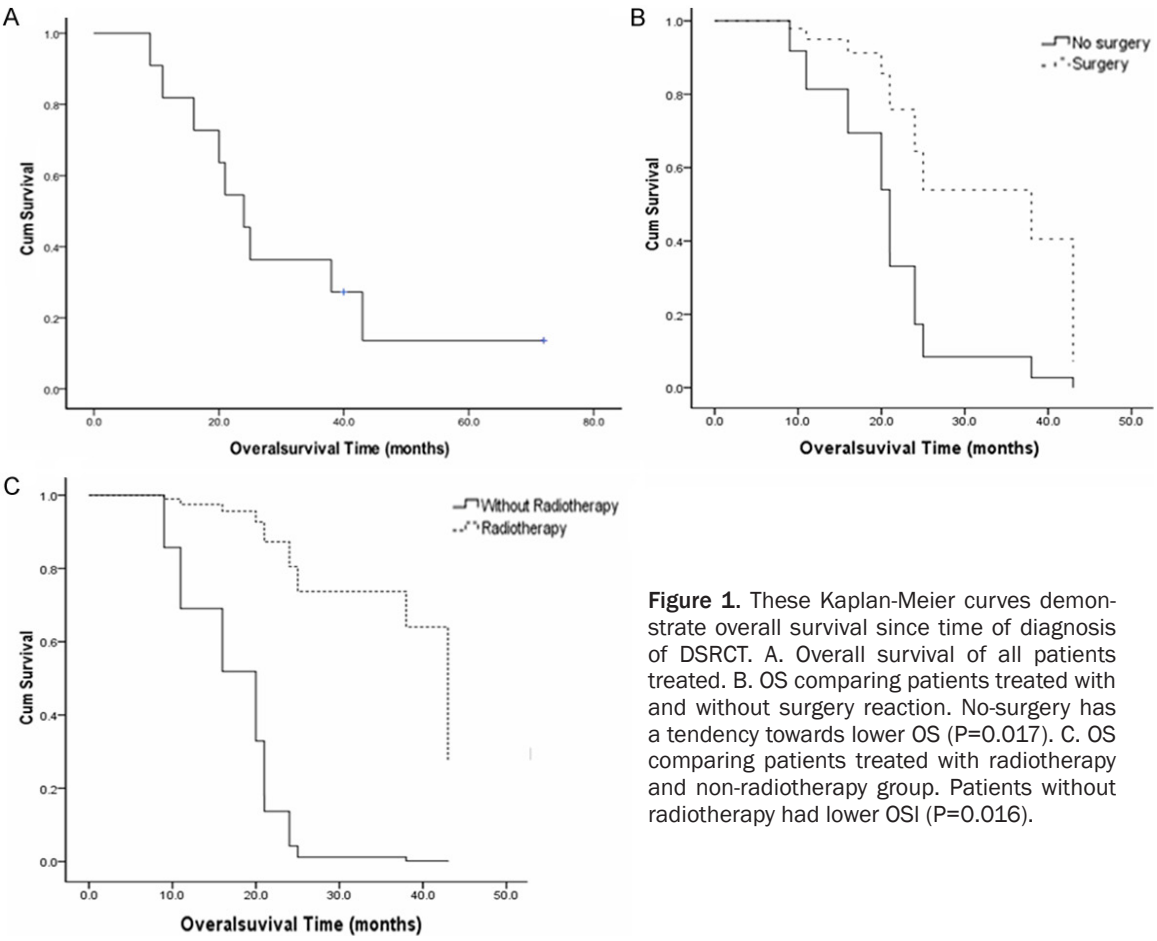

Figure 1. These Kaplan-Meier curves demonstrate overall survival since time of diagnosis of DSRCT. A. Overall survival of all patients treated. B. OS comparing patients treated with and without surgery reaction. No-surgery has a tendency towards lower OS (P=0.017). C. OS comparing patients treated with radiotherapy and non-radiotherapy group. Patients without radiotherapy had lower OSI (P=0.016).

the operation group, three patients were evaluable for response, 1 patient had PR, 1 patient had stable disease, and 1 patient had progressive disease. In nonoperation group, all six patients were evaluable for response. 1 patient had CR, 1 patient had PR, 2 patients had stable disease, and 2 patients had progressive disease.

Outcome and survival

The 3-year and 5-year overall survival (OS) rates were 36.4 % and 10.0 % (**Figure 1A**). Of the 5 patients who received surgery, the complete surgery application rate was 40.0 % (2/5). Median OS was 34.5 months in surgery patients, and 24.5 months in non-surgery patients. A statistically significant difference was observed between the two groups (**Figure 1B**, P=0.017). Radiotherapy application rate was 45.5% (5/11). Median OS was 40.8 months in radiotherapy group, and 19.2 months in non-radiotherapy group. The patients with radiotherapy have better overall survival (**Figure 1C**, P=0.017). Nine patients died of disease progression or widespread metastases. Two patients were alive at last follow-up including one patient who survived 72 months after macroscopic total resection and postoperation adjuvant radiotherapy and chemotherapy. One patient was survived 40 months after radiotherapy and chemotherapy. The 3-year progression-free survival rate was 27.2%. The median survival was 29 months, and the median time to local failure was 8.8 months. First recurrences most often involved the liver (66.7%) and/or the abdomen or retroperitoneum (36.4%). Isolated distant failure involving the lungs and/or mediastinal lymph nodes was seen in only 2 cases.

Discussion

DSRCT is an uncommon and highly aggressive malignant tumor. It is acknowledged with relatively specific pathology, unique molecular characteristics, and multidisciplinary therapeutics.

The onset of DSRCT is very unintelligible. Clinically it is with a predilection for young male. As the previous reports, the mean age at diagnosis is approximately 22 years and the male to female ratio is 4:1 [5]. In our study of 11 patients, the mean age was 31.4 years ranging from 14 to 64 years with male to female ratio of 4.5:1, and 63.6% of the patients were diagnosed before 30 years. It usually arises from abdominal or pelvic peritoneum as a diffuse mass and occasionally can also be found in solid organs such as the livers, ovaries, pancreases, kidneys, bones, and brains [6]. Lal et al. [7] indicated that the most common presenting complaint was an intra-abdominal mass (64%). In a literature composite analysis of 71 patients indicated that pain (52.1%) and increased abdominal girth (8.4%) were the major initial symptom or sign [6]. DSRCT has a tendency for peritoneal and omental spread and hematogenous metastasis, especially to the liver, lung and bone [8]. Hepatic or lung involvement and regional or distant nodal metastasis are relatively common.

CT scan, the most widely used imaging examination way, frequently shows multiple bulky, lobulated, heterogeneous, and peritoneal soft-tissue masses with a predilection for intraperitoneal spread without obvious primary organ involvement. In advanced cases, abdominopelvic DSRCT can develop into bulky and multiple masses that displace the neighboring organs. In our series, the most common character was found to have multiple lobulated solid nodules with irregular boundary and widely distributed on the peritoneum. The hypodense areas and heterogeneity reflect tumor hemorrhage or necrosis. Ascites, calcifications, nodular peritoneal thickening, lymphadenopathy, hydronephrosis, and bowel obstruction were associated findings. Bulky peritoneal soft-tissue masses without an apparent organ based primary site are characteristic of intra-abdominal desmoplastic small round cell tumor [9]. The radiological exam of DSRCT can provide useful information on the tumor position, size and the efficacy evaluation.

Pathology and IHC provide strong evidences for confirmative diagnosis of DSRCT. The characteristic histological features are undifferentiated small to medium sized uniform round cells with scanty cytoplasm and hyperchromic nuclei, which arrange in nest or spindle cell morphology embedded in dense stroma. These characteristic appearances of DSRCT distinguish it from other small round cell tumors. The immunohistochemical profile of DSRCT shows divergent differentiation, a distinct feature, with the neoplastic cells which typically express epithelial (keratin, EMA), vimentin, desmin, and neural markers (NSE and CD56) in IHC analysis provide further testimonies for confirmative and differential diagnosis [10]. Moreover, DSRCT shows a distinctive chromosomal translocation t (11; 22) (p13; q13), resulting in formation of a specific EWS-WT1 fusion gene transcript [11], which can be detected by reverse transcriptase-polymerase chain reaction, FISH, and molecular assays. A high serum cancer antigen 125 level may be a useful marker for DSRCT, permitting early diagnosis and treatment [12]. Lae et al. [13] reported 32 cases of DSRCTs that 84% of cases were immunoreactive for NSE, 81% for desmin, and 87% for keratin. Also, 91% of cases were positive for WT1. Consequently, immunohistochemical examination in combination with molecular study provides a confirmative diagnosis of DSRCT.

To our knowledge, no consensus has been reached concerning the optimal strategy for managing DSRCT, and the treatment remains a clinical challenge. Furthermore, because of the difference of the therapeutic modalities utilized, it is difficult to compare the effectiveness of various regimens. However, the current literatures and our results suggested that an aggressive approach involving total macroscopic tumor excision combined with radiotherapy and chemotherapy may offer the best opportunity for disease control and disease-free survival.

Aggressive tumor resection is currently accepted to have a primary role in the achievement of prolonged survival of other malignancies involving the peritoneum [14]. Primary complete or partial removal of macroscopic disease was possible in only 60% of the patients in the current series but was associated with a longer median survival. Quaglia et al. [15] reported that 3-year overall survival was 58% in patients with gross total resection in comparison with 0% in the nonresection cohort. In our study, 5 of the 11 patients received surgery. The median OS was 34.5 months in surgery patients,

and 24.5 months in non-surgery patients. There was a statistically significant difference between the two groups (**Figure 1B**, P=0.017). We suggested that an attempt should be made to remove all macroscopic tumor, although macroscopic resection of the tumor often is unfeasible technically, due to the extensive involvement of this disease at the time of presentation. The one patient in our case who had 72 months long-term disease-free survival underwent macroscopic resection of both his primary tumors. Another important role of surgery in DSRCT is the relief of symptomatic gastrointestinal obstruction, which reportedly develops in half of the patients with this disease.

There is no standard chemotherapy regimen for DSRCT. Similar to other small round-cell tumors, DSRCT is alkylator-sensitive and dose-responsive. Kushner et al. [16] reported a 100% response rate using a chemotherapy regimen (P6 protocol) consisting of vincristine, doxorubicin, cyclophosphamide, ifosfamide, and etoposide in 10 patients with DSRCT. Its combination with other treatment modalities such as surgery, radiation, autologous stem cell rescue, or the combination of all of the above was used. The median survival time was 19 months. Some researchers thought that anthracycline-based therapy regimens (doxorubicin, etoposide, cisplatin, and cyclophosphamide) may be best used for recurrent disease [17, 18]. Some patients have been reported to achieve complete radiological remission after chemotherapy [18, 19]. An alternative more tolerable outpatient regimen has more recently been used for DSRCT as documented in a case report [20]. In this report, neoadjuvant chemotherapy included vincristine, ifosfamide, dexrazoxane/doxorubicin, and etoposide. Continuous hyperthermic peritoneal perfusion HIPEC with cisplatin was given after extensive cytoreductive surgery. This was followed by irinotecan + temozolomide monthly 2 and then abdominal radiation 30 Gy with simultaneous temozolomide. A total of 12 cycles of irinotecan and temozolomide were given and provided a disease-free interval of nearly 2 years. In our cases, cyclophosphamide/ifosfamide, doxorubicin and cisplatin were utilized. Tren patients all accomplished the treatment successfully with no grade 4 marrow depression, and gastrointestinal toxicity. The side effects had certainly been handled expeditiously by effective medical treatment.

Goodman et al. [21] reported whole abdominopelvic irradiation in 21 patients who had received chemotherapy followed by debulking operation, and the median time to relapse was 19 months and median overall survival was 32 months. More recently, Pinnix et al. [22] reported 8 patients treated with whole abdominopelvic Intensity Modulated Radiation Therapy (IMRT) after neoadjuvant chemotherapy and debulking surgery (and HIPEC for 7 patients). They concluded that postoperative IMRT is feasible and well tolerated after aggressive surgery with no grade 4 digestive symptoms. Based on these reports, radiotherapy appears feasible in patients with DSRCT. In our study, 5 patient received radiotherapy, and the radiotherapy application rate was 45.5%. The median OS in radiotherapy group improves obviously compared with non-radiotherapy group (**Figure 1C**, P=0.017). Therefore, we considered that whole abdominopelvic irradiation is a effective treatment strategy for DSRCT. Because the tumor has the property of multicentric growth in the abdominopelvic cavity, macroscopic resection of the tumor often is unfeasible technically. Radiotherapy is of great importance in managing unresectable DSRCT.

Despite multiple treatment strategies including systemic chemotherapy regimens active for DSRCT, aggressive debulking surgery, whole abdominal radiation, or even autologous stem cell transplant, the prognosis of DSRCT is poor and most cases die within 3 years [23]. Some tertiary-care center experiences have characterized 3-year and 5-year OS rates of 55% to 71% and 15% [7, 17]. In our series, we have 2 patients that have no evidence of disease with extended long-term follow-up. The patients all received radiotherapy and chemotherapy. Schwarz et al. [24] reported that the median progression-free survival was 2.6 years, and the progression-free survival at 5 years after diagnosis was 18%. Ordonez [25] indicated that 71% (25/35) of patients died in 8-50 months (mean 25.2 months) because of the widespread metastasis of tumors after the diagnosis of DSRCT. In our study, 81.8% (9/11) of the patients died in 9 to 43 months (mean 23.0 months), the data was basically in agreement with the literature. Further study showed that improved survival was correlated with a complete or very good partial response to multimodality therapy, surgical debulking, and use of the chemotherapy [26].

Conclusion

DSRCT is a rare and an aggressive malignancy with poor outcome. Management of DSRCT remains challenging and lack of consensus, thereby emphasizing on multimodality treatment. According to our analyses, we would recommend aggressive multimodal therapy incorporating aggressive surgical debulking, intensive chemotherapy, and radiotherapy for advanced abdominal DSRCT patients. The clinical application of radiotherapy in treating DSRCT should not be overlooked. When the patients can not tolerate an aggressive surgery or the tumors are unable to be resected by operation, a comprehensive therapy included radiotherapy combined with chemotherapy could relieve symptoms and improve the quality of life. More investigations and longer follow-up are warranted to define the optimal therapeutic strategy.

Disclosure of conflict of interest

None.

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