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RESEARCH

# Analysis of Clinicopathological Features and Prognostic Factors of Desmoplastic Small Round Cell Tumor

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**Abstract** Desmoplastic small round cell tumor (DSRCT) is a relatively uncommon and highly aggressive malignancy in young males. It is associated with a poor outcome, due in part to missed diagnosis. To characterize the clinical pathological features of DSRCT in Chinese patients and to find out the characteristics of treatment and prognostic factors, the authors collected and analyzed the clinical information of 48 cases. A total of 48 cases of DSRCT between March 1995 and March 2012 were retrospectively reviewed and analyzed. The clinical information, histological results and survival data of the patients were collected. Median age was 26.96±14.09 years with a range of 6–66 years. Thirty-three patients (68.75 %) were seen before 30 years old, and 15 patients (31.25 %) were diagnosed after 30 years old. The male-to-female ratio is 3.36 :1. Among them, 37 cases presented with tumors in the abdominal or pelvic cavity; the other 11 cases had extra-abdominal tumors. The most common symptoms were abdominal pain (19/48, 39.58 %) and palpable mass (12/48, 25.00 %). The percentage of patients received surgery, complete surgery, and chemotherapy was 79.17 %, 37.50 %, and 52.08 %, respectively. Median follow-up duration was 2.67 years. Median overall survival for all patients was 24.33 months (95 % CI: 9.74–38.92 months) and median event-free survival for all patients was 8.00 months (95 % CI: 5.13–10.89 months). Univariate analysis revealed that surgery, effective debulking surgery, chemotherapy and any two or more combined therapeutics were significant prognostic factors for longer overall survival ($p<0.05$). Cox regression analysis showed complete surgery was an independent prognostic factor. Standard therapy for DSRCT consists of combination of surgical resection and postoperative chemotherapy. Complete surgery is an independent prognostic factor and should be further investigated.

**Keywords** Desmoplastic small round cell tumor · Prognosis

## Introduction

Desmoplastic small round cell tumor (DSRCT) is a member of small round cell tumor family and was first described by Gerald and Rosai in 1989. It is an exceedingly rare malignancy with a predilection for adolescent and young male, moreover, it typically presents as an intra-abdominal mass with multiple intra-peritoneal implants, and pathologically composed of undifferentiated small round cells with unknown origin and fibrous stroma. The main diagnostic feature of the tumor cells is multi-lineage potential with co-expression of epithelial, mesenchymal, and neural markers. A specific chromosomal translocation, t (11; 22) (p13; q12), has been documented in DSRCT and is increasingly used to confirm the diagnosis. 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. Since rare diseases are often not diagnosed due to the

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inexperience of the physicians, optimal treatment strategies and prognosis of DSRCT remain controversial. Using the data from our hospitals, an attempt was made to identify clinical pathological features and prognostic factors in this study.

## Materials and Methods

### Collection of Individual Patient Data

The medical records of patients diagnosed with DSRCT between March 1995 and March 2012 in China-Japan Union Hospital, Suzhou Kowloon Hospital, Shanghai Yangpu District Center Hospital, Shanghai Ruijin Hospital, Shanghai Cancer Hospital of Fudan University and et al. were reviewed for patients' clinical characteristics, histological and immunochemical data, treatments and survival time. Records of patient characteristics included sex, age, tumor sites, size and stage, disease progression and postoperative disease recurrence were also analyzed.

### Statistical Analysis

The findings were analyzed using SPSS for Windows, Version 16. Survival outcomes were estimated 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.

## Results

### Clinical Presentation

Forty-eight patients with DSRCT were identified. Of these 37 were male (77.08 %) and 11 were female (22.92 %), with male: female ratio of 3.36:1 and age ranging from 6 to 66 years (mean, 26.96 ±14.09). The courses of the disease were range from 1 to 365 days, with mean 86.76 days. Clinically, the most common complaint was abdominal pain (19/48, 39.58 %), followed by symptoms related to mass (12/48, 25.00 %), and the other symptoms were weight loss (4/48, 8.33 %), cough (3/48, 6.25 %), back pain, blurred vision, and leg pain (2/48, 4.17 %), urinary irritation symptoms, anemia, bloody stool and constipation (1/48, 2.08 %). On radiological imaging 77.08 % (37/48) of tumors involved multiple sites within the abdominal cavity and 11 (22.92 %) tumors occurred in extra-abdominal sites. Intra-abdominal tumors were identified in the abdomen (27), pelvic region (4), left kidney (2), liver (2), pancreas (1) and uterus (1). The extra-abdominal tumors were detected in the testis (3), lung (2), thoracic cavity (2), legs (2) and ethmoid sinus (2). Tumor size varied from 0.3 to 18.0 cm in maximum dimension with 31.25 % more than 10 cm in maximum dimension, 56.25 % between 5 cm and 10 cm and 12.50 % less than 5 cm (As shown in Table 1). Distant metastasis was observed in 16.67 % (8/48) of these patients at diagnosis and local involvement was observed in 64.58 % (31/48). The most common site of metastasis was liver (six patients). Metastasis were also found in the lungs (two patients). Recurrence during the follow-up period was observed in 17 of the 28(60.71 %) patients, and 20 of the patients without definite data for tumor recurrence.

### Tumor Pathology and Immunohistochemical (IHC) Analysis

Histopathologically, tumors generally displayed nests of tumor cells and abundant desmoplastic stroma (Fig. 1a). Necrosis was noted in 17 tumors (17/48, 35.42 %). Tumor cells had hyperchromatic nuclei with indistinct nucleoli (Fig. 1b).

Panel of primary antibodies were used for immunohistochemical staining. The tumor cells showed focal to diffuse positivity for cytokeratins (CK)(37/42, 88.10 %), epithelial membrane antigen (EMA) (33/41, 80.49 %), desmin (45/46, 97.83 %), vimentin (43/45, 95.56 %), CD99(6/20, 30.00 %), neuron-specific enolase (NSE) (38/45, 84.44 %), synaptophysin (2/15, 13.33 %) and chromogranin (4/19, 21.05 %). Moreover,

**Table 1** Analysis between clinical pathological characteristics and overall survival in patients with DSRCT (48 cases)

| Characteristic | Status | N | Number of events | Log-rank p |
|---|---|---|---|---|
| Age of onset | <30 years | 33 | 10 | 0.237 |
| | ≥30 years | 15 | 5 | |
| Sex | Male | 37 | 10 | 0.715 |
| | Female | 11 | 5 | |
| Site | Intra-abdominal | 36 | 10 | 0.301 |
| | Extra-abdominal | 12 | 5 | |
| Surgery | Negative | 10 | 2 | 0.026 |
| | Positive | 38 | 13 | |
| Complete surgery | Non-complete S | 20 | 4 | 0.004 |
| | Complete S | 18 | 9 | |
| Size | <5 cm | 6 | 2 | 0.858 |
| | 5–10 cm | 27 | 9 | |
| | ≥10 cm | 15 | 4 | |
| Stage | Gilly2 | 2 | 0 | 0.428 |
| | Gilly3 | 2 | 1 | |
| | Gilly4 | 44 | 14 | |
| Necrosis | Negative | 4 | 1 | 0.438 |
| | Positive | 17 | 3 | |
| Chemotherapy | Negative | 23 | 3 | 0.026 |
| | Positive | 25 | 12 | |
| Combined two or more therapies | Negative | 28 | 5 | 0.006 |
| | Positive | 20 | 10 | |

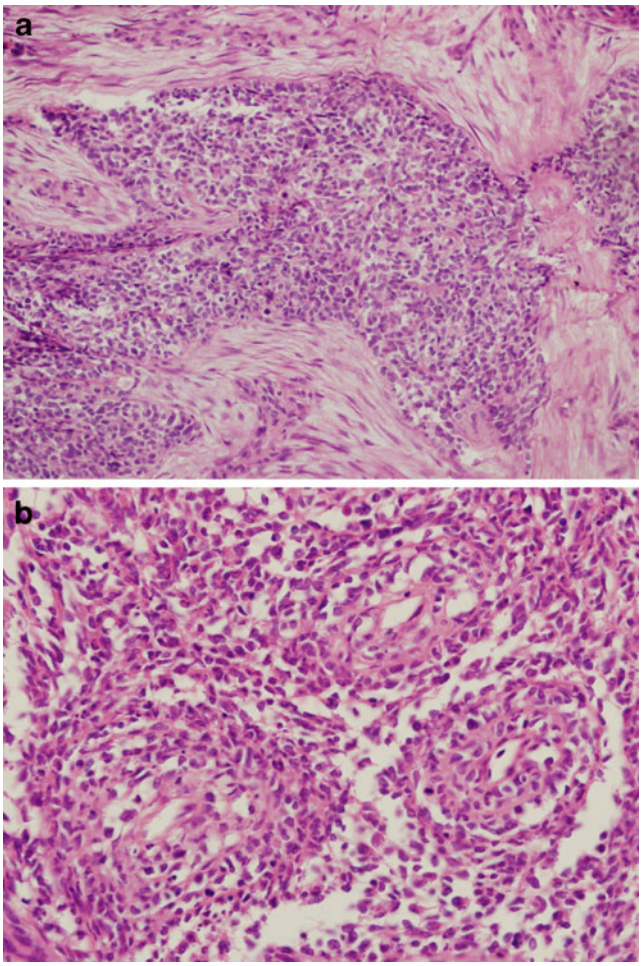


**Fig. 1** The imaging of our patient with DSRCT in abdominal cavity. (**a**) Routine histological hematoxilin and eosin (HE) staining and assay of DSRCT. Nests of small round cells separated by desmoplastic stroma (×200). (**b**) Small round cells with hyperchromatic nuclei, and indistinct nucleoli (×400)

tumor or stromal cells were also positive for SMA (10/13, 76.92 %) and HBME1 (2/2, 100.00 %). In addition, the tumors of all patients were negative for calretinin, human melanoma antibody (HMB45), nuclear factor (NF) and CD20. The results of immunohistochemical staining of 48 patients were summarized in Table 2. In one of our patients, the tumor had been found to co-express epithelial, mesenchymal and neural cell markers. The IHC results were listed in Fig. 2a–f.

Molecular evidence of t(11;22) (p13;q12), the defining cytogenetic abnormality of DSRCT, was demonstrated in few of the patients in this cohort. Only four of the 48 tumors were confirmed with positive molecular results by fluorescent in situ hybridization (FISH). These included tumors involving abdomen (1), renal (1), pancreas (1), paratestis (1).

As far as tumor markers were concerned, there was no tumor-specific marker for DSRCT. Clinically, an elevated level of serum CA 125 or NSE concentration was found in some patients as previous reports. In the present study, serum alpha-fetoprotein (AFP), carcinoembryonic antigen (CEA), CA l9-9,

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

| No. | Antibody marker | Positivity | ± | Negative | Percentage |
|---|---|---|---|---|---|
| 1 | Cytokeratin(CK) | 37 | 1 | 4 | 37/42 88.10 % |
| 2 | Epithelial membrane antigen(EMA) | 33 | 6 | 2 | 33/41 80.49 % |
| 3 | Vimentin | 43 | 2 | 0 | 43/45 95.56 % |
| 4 | Desmin | 45 | 0 | 1 | 45/46 97.83 % |
| 5 | Neuron specific enolase (NSE) | 38 | 1 | 6 | 38/45 84.44 % |
| 6 | Synaptophysin | 2 | 4 | 9 | 2/15 13.33 % |
| 7 | Chromogranin A | 4 | 0 | 15 | 4/19 21.05 % |
| 8 | S-100 | 3 | 4 | 17 | 3/24 12.50 % |
| 9 | CD56 | 1 | 0 | 1 | 1/2 50.00 % |
| 10 | LCA | 1 | 0 | 25 | 1/26 3.85 % |
| 11 | Calretinin | 0 | 0 | 6 | 0/6 0 % |
| 12 | CD99 | 6 | 6 | 8 | 6/20 30.00 % |
| 13 | SMA | 10 | 0 | 3 | 10/13 76.92 % |
| 14 | HMB45 | 0 | 0 | 14 | 0/14 0 % |
| 15 | Actin | 5 | 0 | 5 | 5/10 50.00 % |
| 16 | CD34 | 1 | 0 | 2 | 1/3 33.33 % |
| 17 | CD117 | 1 | 0 | 3 | 1/4 25.00 % |
| 18 | NF | 0 | 2 | 3 | 0/5 0 % |
| 19 | CD20 | 0 | 0 | 8 | 0/8 0 % |
| 20 | HBME1 | 2 | 0 | 0 | 2/2 100.00 % |

CA l25, human chorionic gonadotrophin (HCG), and LDH were measured in some of the patients at presentation. Tumor markers of AFP, CEA and LDH values were always normal. Serum CA125 was elevated in six out of nine cases (66.67 %). Serum CA19-9 was obtained in eight cases before therapy and was elevated in one (12.50 %). The HCG was also elevated in one patient (1/5, 20.00 %).

## Treatment Setting

Therapeutic management of DSRCT remains challenging with low efficacy and no proper consensus, despite the combination of aggressive treatments such as debulking surgery, polychemotherapy, whole abdominal radiation, hyperthermic intraperitoneal chemotherapy (HIPEC), bone marrow transplantation (BMT) and targeted therapy. Aggressive surgical debulking is the mainstay of the therapeutic strategy. Debulking surgery is defined as definitive removal of at least 90 % of the tumor burden, for complete resection is rarely possible as to extensive dissemination. In this study (As shown in Table 3), 20.83 % (10/48) of the patients did not receive any surgery except for biopsy as to extensive distant metastasis and widespread dissemination. Thirty-eight cases received surgery, but surgical debulking was performed initially only in 47.37 % (18/38) of the patients, and measurable

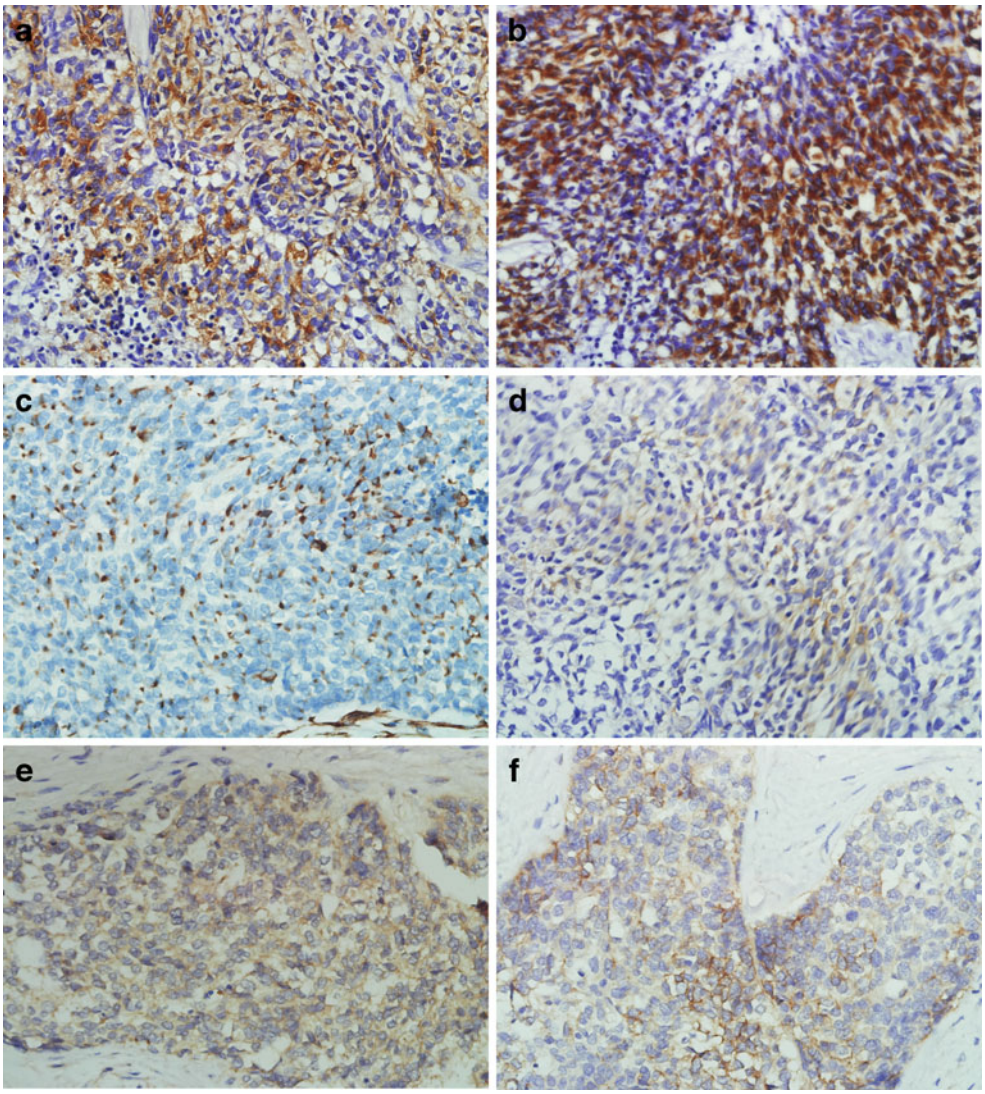


**Fig. 2** Immunohistochemical staining in DSRCT. (**a**) EMA (DAB, original magnification ×400). (**b**) Desmin (DAB, original magnification ×400). (**c**) Vimentin (DAB, original magnification ×400). (**d**) NSE (DAB, original magnification ×400). (E) CD99 (DAB, original magnification ×400). (**f**) CD56 (DAB, original magnification ×400)

residual tumor persisted in most of intra-abdominal cases. Chemotherapy was initiated in 52.08 % (25/48) of the patients and the rest of others did not receive any kind of chemotherapy in the courses of the diseases. Most patients underwent a combination of multi-chemotherapy drugs as reformed P6 regimen or PAVEP regimen and without adjuvant radiotherapy. External beam radiotherapy was delivered at the end of chemotherapy in only one patient. 41.67 % patients received any kind of the combined therapeutics as surgery plus chemotherapy or radiotherapy.

## Prognostic Analysis

The median follow-up time was 32 months (range 1–123 months). Median overall survival for all patients was 24.33 months (95 % CI 9.74–38.92 months) and median

**Table 3** Analysis of OS in DSRCT patients with different treatment setting

| Group | Status | N | x ± s | lower | 95%CI upper | Log-rank p |
|---|---|---|---|---|---|---|
| Surgery | Negative | 10 | 7.70±1.45 | 4.87 | 10.54 | 0.026 |
| | Positive | 38 | 28.21±9.04 | 10.49 | 45.93 | |
| Complete surgery | Negative | 20 | 11.87±2.32 | 7.33 | 16.40 | 0.004 |
| | Positive | 18 | 49.03±19.19 | 11.43 | 84.64 | |
| Chemotherapy | Negative | 23 | 19.16±7.73 | 4.01 | 34.32 | 0.026 |
| | Positive | 25 | 22.81±3.70 | 15.56 | 30.06 | |
| Combined two or more therapies | Negative | 28 | 18.69±7.33 | 4.33 | 33.06 | 0.006 |
| | Positive | 20 | 25.02±4.00 | 17.19 | 32.85 | |

event-free survival for all patients was 8.00 months (95 % CI 5.13–10.88 months). Univariate analysis revealed that surgery, complete surgery(effective surgical debulking), chemotherapy and any two or more combined therapeutics were significant independent prognostic factors for longer overall survival ($p<0.05$). There was no statistical OS difference in age, sex, site, size, stage and with necrosis groups ($p>0.05$, Table 1). Significant differences were found between the groups subdivided by treatment (Table 3, Fig. 3). Ten cases without surgery (20.83 %) were diagnosed by biopsy or needle aspiration cyto-diagnosis. Of the 38 patients who received surgery, the complete surgery application rate was 47.37 % (18/38). Median OS was 28.21±9.04 months (95 % CI, 10.49–45.93) in surgery patients, and 7.70±1.45 months (95 % CI, 4.87–10.54) in non-surgery patients, and a statistically significant difference was observed between the two groups (Table 3, Fig. 3, $p=0.026$) . OS of patients with complete surgery was statistically higher than that of the non-complete surgery patients ($p=0.004$); Adjuvant chemotherapy application rate was 52.08 % (25/48). Median OS was 19.16±7.73 months (95 %

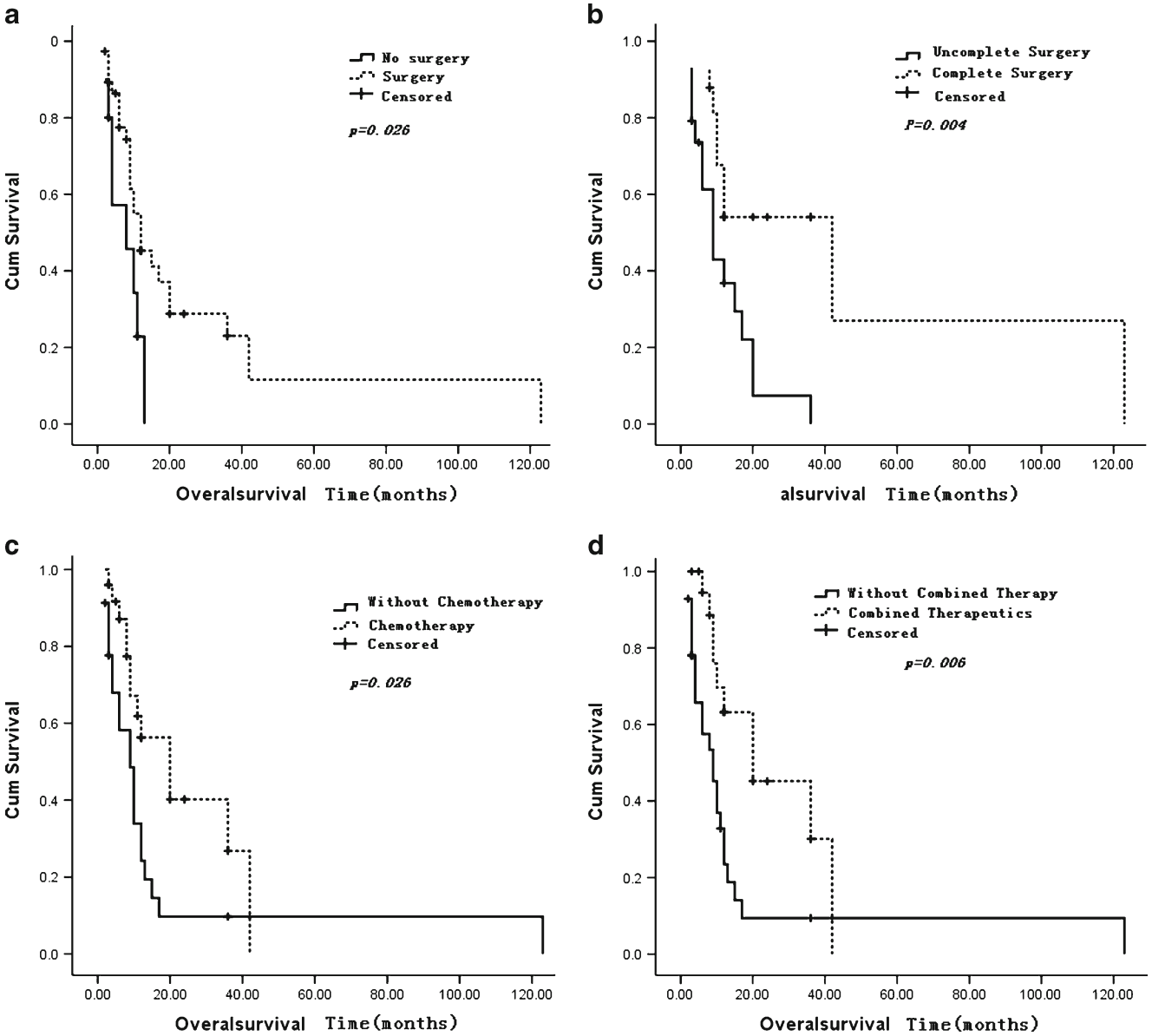


**Fig. 3** Kaplan–Meier survival of DSRCT stratified by different treatment. (**a**) OS for patients with or without surgery. No-surgery was predictive of lower overall survival (p = 0.026) for all patients. (**b**) OS for patients with or without complete surgery. Without complete surgery was correlated with decreased OS (p = 0.004). (**c**) OS for patients with or without chemotherapy. Patients without chemotherapy had lower overall survival (p = 0.026). (**d**) OS for patients with or without combined surgery-chemo-radio therapeutics. Power correlation between the OS and the combined therapeutics (p = 0.006)

CI, 4.01–34.32) in non-chemo patients, and 22.81±-3.70 months (95 % CI, 15.56–30.06) in chemotherapy patients ($p$=0.026). The patients with two or more combined therapeutics with higher OS than those with one kind or no therapy patients (25.02±4.00 vs 18.69±7.33 months, $p$=0.006). Further study by COX regression analysis showed that patients with complete surgery have better overall survival (Table 4), the odds ratio significantly decreased (OR: 0.266); (95 % CI, 0.106–0.670; $p$< 0.05), that is to say, complete surgery will improve OS with 3.76 fold than those without complete surgery patients. In the present study, complete surgery was an independent good predictive marker of prognosis.

## Discussion

DSRCT is an uncommon and highly aggressive malignant tumor. About 300 cases of DSRCT have been reported in the literature since it was initially described by Gerald and Rosai in 1989 [1]. Despite its unknown origin and nonspecific clinical features, DSRCT is acknowledged with relatively specific pathology, unique molecular characteristics, and multidisciplinary therapeutics.

The onset of DSRCT is very occult. The courses of the disease were range from 1 to 365 days in the present study, with mean time 86.76 days. 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 [2]. In our study of 48 Chinese patients, the mean age was 26.96 ±14.09 years ranging from 6 to 66 years with male to female ratio of 3.36:1, and 68.75 % of the patients were diagnosed before 30 years. The Clinical symptoms and signs of DSRCT are nonspecific and complicated. It usually arises from abdominal or pelvic peritoneum as a diffuse mass and sometimes can also be found in solid organs such as the ovaries, livers, kidneys, pancreases, bones, and brains [3]. Comparison of the site differences, the percentage of DSRCT occurring in the abdomen in the study of Gerald et al. was relatively high (103/109, 94.50 %) [4], while the percentage in our study was lower (37/48, 77.10 %), the inconsistance maybe possibly with different groups of people concerned.

**Table 4** Cox regression analysis for overall survival in DSRCT patients

| Parameter | Chi-squared test | SE | OR | 95 % CI | $p$ value |
|---|---|---|---|---|---|
| Age | 1.450 | .442 | .587 | 0.247–1.397 | .228 |
| Gender | 0.001 | .496 | .986 | 0.373–2.606 | .977 |
| Site | .248 | .528 | .769 | 0.273–2.163 | .618 |
| Size | 1.021 | .355 | .698 | 0.348–1.401 | .312 |
| Complete surgery | 7.906 | .470 | .266 | 0.106–0.670 | .005 |
| Chemotherapy | 2.805 | .380 | .529 | 0.251–1.114 | .094 |

Lal DR et al. indicated that the most common presenting complaint was an intra-abdominal mass (64 %) [5] and a composite analysis of 71 patients in the literature indicated that pain (52.1 %) and increased abdominal girth (8.4 %) were the predominant initial symptom or sign [6]. While the most common complaint in this study was abdominal pain (19/48, 39.58 %), followed by symptoms related to mass (12/48, 25.00 %) . DSRCTs have a tendency for peritoneal and omental spread and hematogenous metastasis, especially to the liver, lung and bone [7]. Hepatic or lung involvement and regional or distant nodal metastasis are relatively common in our study at first presentation.

The imaging examination of DSRCT includes ultrasound, CT scan, magnetic resonance scan and FDG-PET/CT imaging. However, radiological exam of DSRCT is also nonspecific, and can just provide useful information on the tumor site, size and the efficacy evaluation. The diagnosis is mainly based on the pathology, immunohistochemistry and the cytogenetic analysis. Histologically, DRSCT is mainly composed of small round blue cells in nests separated by an abundant desmoplastic stroma [8]. In addition, the neoplastic cells which typically express epithelial (e.g., CK and EMA), mesenchymal (e.g., vimentin), myogenic (e.g., desmin), and neural markers (e.g., NSE) in IHC analysis provide further evidence for confirmative and differential diagnosis [9]. Moreover, DSRCT shows a unique chromosomal translocation t (11; 22)(p13; q13), resulting in formation of a specific EWS-WT1 fusion gene transcript [10], which can be detected by reverse transcriptase-polymerase chain reaction, FISH, and molecular assays.

Therapeutic management of DSRCT remains challenging with low efficacy and poor prognosis, despite the combination of aggressive treatments. Debulking surgery is defined as definitive removal of at least 90 % of the tumor burden [11]. Biswas et al. [12] did conclude that complete surgical excision seems to provide a better survival, but additional adjuvant therapy is urgent due to the high recurrence and aggressive biology of the tumor [13]. The most representative one of chemotherapy was P6 regimen, which reported in 1996 by Kushner et al. [14] and had been approved to be effective against DSRCT. In 2002, Bertuzzi et al. [15] explored a high-dose chemotherapy (HD-CT) approach in poor-prognosis adult small round-cell tumors, but the objective response rate of DSRCT patients was poor than other histologic types. In this study, 20.83 % (10/48) of the patients did not receive any surgery except for biopsy, surgical debulking was performed initially only in 47.37 % (18/38) patients, and chemotherapy was initiated in 52.08 % (25/48) of the patients and one patient underwent radiotherapy, the rest of others did not receive any kind of therapeutics. We postulate that combination surgery and chemotherapy or other therapeutics might benefit patients to achieve the

maximum response and possibly improved survival, but only 41.67 % of the patients received combination therapy in the present study. So improvement of the doctors' concept of multidisciplinary treatment to DSRCT could be considered an approach that is important for patients.

In general, the prognosis of DSRCT patients is poor. Previous clinic-pathologic studies have documented the aggressive nature of DSRCT, but most of the studies have focused on clinical presentation and diagnostic criteria rather than prognostic variables. The study of Schwarz RE et al. suggested that the median progression-free survival was 2.6 years (95 % CI; 1.6–3.5 years), and the progression-free survival at 5 years after diagnosis was 18 % [16]. The study of Ordonez indicates that 71 % (25/35) of patients died in 8 to 50 months (mean 25.2 months) [17]. Though compared with other chemotherapies, the P6 proposal provides a much better curative effect, but the survival rate of DSRCT in a 3-year period is only 29 % [14]. In our study, 68.8 % (33/48) of the patients died in 2 to 123 months(mean 13.63 months), we postulate that the mean overall survival was much lower than that of other's reports for part of the patients in present study without any kind of therapies after surgery. Further study of Schwarz RE showed that improved survival was correlated with a complete or very good partial response to multimodality therapy, surgical debulking, and use of the P6 protocol [12]. Resection was also found to directly affect the prognosis, and complete surgery was found to be an independent favorable prognostic factor in our study, this is consistent with the literature report. Compared with patients who underwent resection, patients who did not have the surgery survived for a shorter period of time; better survival rates were related to complete resection of the tumor, patients with complete surgery had better overall survival, the odds ratio significantly decreased (OR: 0.266); (95 % CI, 0.106–0.670; $p < 0.050$), that is to say, complete surgery will improve OS with 3.76 fold than those without complete surgery patients.

There is number of reports showing that chemotherapy could improve patients' survival [18]. Kushner reported improved progression-free survival after aggressive chemotherapy with a high-dose multiagent regimen and aggressive resection followed by total abdominal radiation [12]. We found in the present study that a trend for better OS in patients with chemotherapy, the median survival of patients with chemotherapy was 20.00±5.22 months, much higher than that of patients without chemotherapy. This is in accordance with most published studies, which have demonstrated chemotherapy is a favorable prognostic factor of DSRCT [19]. Furthermore, it is acknowledged that intensive combination chemotherapy regimens are associated with higher efficacy and greater toxicity. In contrast, the study discussed previously by Bertuzzi A did not find the advantage of intensive chemotherapy [15]. In this retrospective study, only ten patients received adjuvant chemotherapy and 15 patients received first-line salvage chemotherapy with regimens containing two to five drugs of doxorubicin, ifosfamide, cyclophosphamide, etoposide, cisplatin, fluorouracil, or nadaplatin. Only four patients received modified P6 regimen for salvage therapy. So we could not do analysis of P6 regimen compared with reformed relative low dose conventional regimens, for the dose and drugs of regimens was not uniform and too small sample. Given that age and performance status (PS) also have a major impact on the effect of chemotherapy and affect the doctor's choice of regimens, however, our further study indicated that age was not shown to be related with survival and PS was not accurately documented in most of cases and, thus, limited any statistical analysis. We therefore speculate that the multiple drug regimens might have led to the superior clinical outcome observed in patients with good PS, but which kind of combination chemotherapy could improve survival remains unproven and needs to be confirmed in large phase III randomized trials.

It has been suggested that there is a potential advantage for combination chemotherapy with other therapeutics in terms of superior response rates and overall survival. Radiation therapy is helpful in prolonging life but has not resulted in long-term, disease-free survival. According to Lal DR's study [5], 29 of these patients (44 %) underwent induction chemotherapy (P6), surgical debulking, and radiotherapy. Overall, 3- and 5-year survivals were 44 % and 15 %, respectively. Three-year survival was 55 % in those receiving chemotherapy, surgery, and radiotherapy versus 27 % when all three modalities were not used (P <0 .020). We also identified combination therapeutics to be a favorable prognostic factor for survival. In our opinion, if resection is an option, the surgery should be performed as early as possible, then chemotherapy with P6 or modified P6 was recommended.

Our patients in this study, on the other hand, were treated on the basis of clinical need in different clinical centers, complete data on PS in our dataset are lacking, most of the patients with different chemo-regimens and few people received radiotherapy, hence, it may not be directly comparable and could be considered as only a preliminary exploration of prognosis study to this rare tumor subtype and further prospective multicenter random phase III trials should be recommended to do.

## 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. Complete surgical intervention is an independent favorable prognostic factor and further prospective studies in treatment are needed to improve long-term survival.

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