The Role and Clinical Effectiveness of Multiline Chemotherapy in Advanced Desmoplastic Small Round Cell Tumor

Hye hyun Jeong1, Yong Sang Hong1, Young-Hoon Kim2, Chan Wook Kim3, Si Yeol Song4, Joon Seon Song5, Kyung-Ja Cho5, Jeong Eun Kim1* and Jin-Hee Ahn1*

1Department of Oncology, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Republic of Korea. 2Division of Kidney and Pancreas Transplantation, Department of Surgery, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Republic of Korea. 3Department of Colon and Rectal Surgery, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Republic of Korea. 4Department of Radiation Oncology, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Republic of Korea. 5Department of Pathology, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Republic of Korea.

ABSTRACT

BACKGROUND: A multimodal approach is the standard treatment for desmoplastic small round cell tumor (DSRCT); however, many patients are diagnosed with inoperable disease, which leaves chemotherapy as the only treatment option. There are limited data on the effectiveness of palliative chemotherapy, especially when used after first-line treatment. Here, we evaluated the clinical outcomes of patients with DSRCT treated with multiple lines of chemotherapy.

METHODS: We reviewed medical records of 14 patients with pathologically confirmed DSRCT at Asan Medical Center between 2004 and 2018.

RESULTS: The median age at diagnosis was 25, with males comprising 92.9% of patients. All patients had inoperable disease at presentation and received chemotherapy as the initial treatment. Four patients (28.6%) were treated with surgery, and complete resection was achieved in 1 patient. Median overall survival (OS) was 23.9 months, and 1-, 2-, and 3-year survival rates were 92.9%, 48.6%, and 19.5%, respectively. In patients receiving first- (N = 14), second- (N = 10), and third-line (N = 8) chemotherapy, median time-to-progression was 9.9, 3.5, and 2.5 months, respectively, and the disease control rates were 100%, 88.9%, and 75.0%, respectively. Factors associated with longer OS in the univariable analysis were ≤2 metastatic sites at presentation (27.0 vs 14.7 months; P = .024) and surgery with intended complete resection (43.5 vs 20.1 months; P = .027).

CONCLUSIONS: Although advanced DSRCT may initially respond to chemotherapy after first-line treatment, the response becomes less durable as the disease progresses. Individualized treatment decisions focused on palliation should be made.

KEYWORDS: Desmoplastic small round cell tumor, palliative chemotherapy, multiline chemotherapy

Background

Desmoplastic small round cell tumor (DSRCT) is a rare and aggressive form of sarcoma. Because of its rarity, the disease was recognized only recently in 1989. Since then, a handful of small-to-medium-sized retrospective studies have been conducted, and the overall incidence of DSRCT is estimated at approximately 0.2 to 0.5 cases/million people. The peak age of incidence is 20 to 24 years old, and it predominantly occurs in males. Clinically, these tumors usually present as multiple peritoneal soft tissue masses. Most patients have advanced disease at presentation, and approximately 60% of patients initially present with extra-abdominal metastases. Histologically, DSRCT is characterized by nests of small, round, blue cells with a desmoplastic stroma, and molecularly, these tumors show a characteristic translocation, t(11;22) (p13;q12), with fusion of the EWSR1 and WT1 genes. Other heterogeneous genetic alterations have been reported, but no actionable targets have been established.

The disease is frequently detected at an inoperable stage, and complete resection is not feasible in many cases. Favorable initial responses to combination chemotherapy, such as alkylating agent-based regimens, are not durable. A multimodal approach including combination chemotherapy, aggressive surgical resection, radiotherapy, and hyperthermic
intraperitoneal chemotherapy (HIPEC) has been reported in the literature, showing clinical benefits in the management of DSRCT. However, the survival outcomes of these patients are still poor, with a reported median overall survival (OS) of less than 3 years and with long-term, disease-free survival unlikely to be achieved in many cases because of recurrence and/or disease progression. In these patients, palliative chemotherapy often becomes the only viable treatment option. Currently, there is limited information on the effectiveness of palliative chemotherapy, especially after first-line treatment. Here, we analyzed the treatment response and survival outcomes of patients with DSRCT treated in our institution with an emphasis on the effectiveness of multiline chemotherapy.

**Methods**

**Patients**

Medical records of patients who were pathologically diagnosed with DSRCT between 2004 and 2018 at Asan Medical Center, a tertiary referral center in Seoul, South Korea, were identified and reviewed. All patient data were retrieved from the retrospective sarcoma registry maintained by the Center for Cancer Data Management of the Asan Cancer Institute, which extracts de-identified research data from the hospital electronic medical records. Patients who were under 18 years of age were excluded because the retrospective registry used in this study did not include pediatric records. Treatment response was assessed according to the revised Response Evaluation Criteria in Solid Tumors version 1.1 (RECIST 1.1). This study was approved by the institutional review board of Asan Medical Center (approval number: #2018-0751) and was performed in accordance with the ethical standards of the institutional research committee and the Declaration of Helsinki.

**Statistical analysis**

Baseline characteristics were analyzed using descriptive methods. Overall survival was defined as the time period from the date of diagnosis to the date of death from any cause. Progression-free survival (PFS) for each line of chemotherapy was defined as the time period from the date of the start of each line of chemotherapy to the date of disease progression or death from any cause, whichever occurred first. Time-to-progression (TTP) was defined as the time period from the date of the start of each line of chemotherapy or the date of surgery to the date of disease progression. Survival outcome estimation and univariable risk factor analysis were performed using the Kaplan-Meier method. A P-value < .05 was considered to indicate statistical significance. All analyses were performed using R version 3.5.3 (R Foundation for Statistical Computing, Vienna, Austria).

**Results**

**Patient characteristics**

A total of 14 patients who were pathologically diagnosed with DSRCT were identified and included in the analysis (Figure 1). The median follow-up duration was 17.4 months (range,
Of the 14 patients, 13 (92.9%) were men. The median age at diagnosis was 25 years (range, 19-39). All 14 patients had unresectable disease at presentation, and 10 patients (71.4%) had extra-abdominal disease. The most commonly involved sites were the lymph nodes (13 patients, 92.9%) and the peritoneum (12 patients, 85.7%). Baseline patient characteristics are shown in Table 1.

**Treatment overview**

Treatment details are summarized in Table 2. All patients received chemotherapy as the initial treatment after diagnosis, except for one patient who received palliative colostomy before pathologic diagnosis. Four patients (28.6%) underwent surgery during the course of treatment; all surgeries were preceded by chemotherapy. Among those 4 patients, the median number of surgeries per patient was 1.5 (range, 1-4). Surgery with the intent of macroscopic complete resection was performed in 2 patients. Complete resection was achieved in one patient, and this patient proceeded to receive adjuvant chemotherapy. Another patient had residual small metastatic lymphadenopathies and proceeded to second-line chemotherapy after watchful waiting until definite disease progression was observed. Remaining 2 patients underwent palliative surgery to alleviate debilitating symptoms caused by intra-abdominal masses.

The median TTP from surgery to disease progression was 7.4 months (95% confidence interval [CI], 0.5-not estimated [NE]). The 2 patients with the longest TTP were those who underwent surgery attempting complete resection (Supplementary Table S1).

**Chemotherapy effectiveness**

In all 14 patients, the first-line chemotherapy regimen was alkylating agent-based P6 protocol which combined vincristine, doxorubicin, cyclophosphamide, ifosfamide, and etoposide.\(^{15,16}\) The disease control rate (DCR) and overall response rate (ORR) to first-line chemotherapy were 100% and 57.1%, respectively. Median time-to-progression to first-line chemotherapy (TTP\(_1\)) was 9.9 months (95% CI, 7.2-11.7). During follow-up, 13 patients (92.9%) developed radiologically confirmed progressive disease (PD) after first-line chemotherapy despite initial response, except one patient who underwent cytoreductive surgery after chemotherapy and currently on treatment holiday.

Ten out of 13 patients (76.9%) who developed PD after first-line therapy proceeded to second-line chemotherapy. All regimens used in the second-line were cytotoxic chemotherapies, including ICE (ifosfamide, carboplatin, etoposide) and VIP (etoposide, ifosfamide, cisplatin) (Table 2). The DCR and ORR to second-line chemotherapy were 88.9% and 0%, respectively. Median TTP to second-line chemotherapy (TTP\(_2\)) was 3.5 months (95% CI, 1.4-8.6). Third-line chemotherapy was administered in 8 patients, all of whom received cytoreductive surgery after chemotherapy and currently on treatment holiday. Ten out of 13 patients (76.9%) who developed PD after first-line therapy proceeded to second-line chemotherapy. All regimens used in the second-line were cytotoxic chemotherapies, including ICE (ifosfamide, carboplatin, etoposide) and VIP (etoposide, ifosfamide, cisplatin) (Table 2). The DCR and ORR to second-line chemotherapy were 88.9% and 0%, respectively. Median TTP to second-line chemotherapy (TTP\(_2\)) was 3.5 months (95% CI, 1.4-8.6). Third-line chemotherapy was administered in 8 patients, all of whom received cytoreductive surgery after chemotherapy.

Patients with longer TTP\(_1\) duration also tended to have longer TTP\(_3\) (median 3.6 months [95% CI, 2.5-NE] vs 1.6 months [95% CI, 0.9-4.8]). Of the 14 patients, 13 (92.9%) were men. The median age at diagnosis was 25 years (range, 19-39). All 14 patients had unresectable disease at presentation, and 10 patients (71.4%) had extra-abdominal disease. The most commonly involved sites were the lymph nodes (13 patients, 92.9%) and the peritoneum (12 patients, 85.7%). Baseline patient characteristics are shown in Table 1.

**Table 1. Baseline characteristics at presentation.**

| Characteristic                              | TOTAL N = 14 |
|--------------------------------------------|--------------|
| Age at diagnosis                           | Median (range) 25 (19-39) |
| Sex                                        | Male 13 (92.9%) Female 1 (7.1%) |
| ECOG PS at diagnosis                      | 0-1 12 (85.7%) ≥2 2 (14.3%) |
| Primary tumor extent                       | Intra-abdominal lesion only 4 (28.6%) Extra-abdominal lesion only 0 Both 10 (71.4%) |
| Biopsy method used for diagnosis           | Needle biopsy 13 (92.9%) Open surgical biopsy 1 (7.1%) |
| Maximum tumor diameter at diagnosis (cm)   | Median (range) 12.5 (4.5-18.5) |
| Number of metastatic sites at diagnosis    | 1 3 (21.4%) 2 6 (42.9%) ≥3 5 (35.7%) |
| Involved sites                             | Peritoneum 12 (85.7%) Lymph node (any) 13 (92.9%) Extra-abdominal lymph node 8 (57.1%) Liver 8 (57.1%) Lung 3 (21.4%) Bone 1 (7.1%) Others 7 (50.0%) |

Abbreviation: ECOG PS, Eastern Cooperative Oncology Group Performance Status.
Table 2. Treatment summary.

| A. SURGERY          |               |               |
|---------------------|---------------|---------------|
| Surgery             | N = 14        |               |
| Received            | 4 (28.6%)     |               |
| Not received        | 10 (71.4%)    |               |
| Chemotherapy before surgical tumor resection | N = 4 |               |
| Yes                 | 4 (100%)      |               |
| Best surgical outcome |               |               |
| Microscopic complete resection | 0 |               |
| Macroscopic complete resection | 1 (25.0%) |               |
| Remaining macroscopic lesions | 3 (75.0%) |               |
| Total number of surgeries received per patient | N = 4 |               |
| Median (range)      | 1.5 (1-4)     |               |

| B. CHEMOTHERAPY     |               |               |
|---------------------|---------------|---------------|
| First-line chemotherapy regimen | N = 14 |               |
| P6                  | 14 (100%)     |               |
| BOR to first-line chemotherapy |               |               |
| CR                  | 0             |               |
| PR                  | 8 (57.1%)     |               |
| SD                  | 6 (42.9%)     |               |
| PD                  | 0             |               |
| Reason for first-line chemotherapy cessation |               |               |
| Progression/minimal response | 9 (64.3%) |               |
| Watchful waiting/patient refusal | 4 (28.6%) |               |
| Toxicity/intolerability | 1 (7.1%) |               |
| Second-line chemotherapy regimen |               |               |
| ICE or IE           | 3 (21.4%)     |               |
| VIP or IP           | 3 (21.4%)     |               |
| Gemcitabine/docetaxel | 1 (7.1%) |               |
| Others*             | 3 (21.4%)     |               |
| Not given           | 4 (28.6%)     |               |
| BOR to second-line chemotherapy With measurable lesion N = 8 |               |
| CR                  | 0             |               |
| PR                  | 1 (12.5%)     |               |
| SD                  | 5 (62.5%)     |               |
| PD                  | 2 (25.0%)     |               |
| Fourth-line chemotherapy regimen |               |               |
| Pazopanib           | 3 (21.4%)     |               |
| Others**            | 2 (14.2%)     |               |
| Not given           | 9 (64.3%)     |               |

| C. RADIOTHERAPY      |               |               |
|---------------------|---------------|---------------|
| Abdominal radiotherapy |               |               |
| No                  | 12 (85.7%)    |               |
| Yes                 | 2 (14.3%)     |               |

(Continued)

Abbreviations: BOR, best overall response; CR, complete response; CYVADIC, cyclophosphamide, vincristine, doxorubicin, dacarbazine; ICE, ifosfamide, carboplatin, etoposide; IE, ifosfamide, etoposide; IP, ifosfamide, cisplatin; P6, cyclophosphamide, vincristine, doxorubicin, ifosfamide, etoposide; PD, progressive disease; PR, partial response; SD, stable disease; VIP, etoposide, ifosfamide, cisplatin.

*Include trabectedine, dacarbazine/cisplatin, and vincristine/dactinomycin/ cyclophosphamide.

**Include goserelin/flutamide and gemcitabine/docetaxel.

[0.9-NE], P = .23), but these differences were not statistically significant.

Five patients proceeded to fourth-line chemotherapy. Of note, pazopanib was used in 3 out of 5 patients as a fourth-line treatment, and the best overall response for pazopanib was PD in 2 patients, and the response was not evaluable in one patient.

Survival outcomes and risk factor analyses

The median OS for all patients was 23.9 months (95% CI, 13.5-27.0). The survival rates at 1, 2, and 3 years were 92.9%, 48.6%, and 19.5%, respectively. The PFS of patients receiving first-line
(N = 14), second-line (N = 10), and third-line (N = 8) chemotherapy was 9.9 months (95% CI, 7.2-11.7), 4.2 months (95% CI, 0.9-8.6), and 3.0 months (95% CI, 0.9-6.3), respectively (Figure 2). In the univariable analysis, ≤2 metastatic sites at presentation (27.0 vs 14.7 months; \( P = .024 \)) and surgery with the intent of complete resection (43.5 vs 20.1 months; \( P = .027 \)) were associated with longer OS. The presence of extra-abdominal metastases or the presence of any specific metastatic sites did not show association with OS. In patients who had measurable diseases, those with better tumor response to each line of chemotherapy tended to have longer OS (for first-line chemotherapy, 24.0 and 14.1 months for partial response [PR] and stable disease [SD], respectively, \( P = .630 \); for second-line chemotherapy, 24.7 and 20.1 months for SD and PD, respectively, \( P = .025 \); for third-line

**Figure 2.** Kaplan-Meier survival estimate of the overall patient population. (A) The median overall survival (OS) was 23.9 months (95% confidence interval [CI], 13.5-27.0). (B) The median progression-free survival (PFS) for first-line (N = 14), second-line (N = 10), and third-line (N = 8) chemotherapy was 9.9 months (95% CI, 7.2-11.7), 4.2 months (95% CI, 0.9-8.6), and 3.0 months (95% CI, 0.9-6.3), respectively.
chemotherapy, 38.1, 24.7 months, and not estimated for PR, SD, and PD, respectively, \( P = .900 \), although the differences were not statistically significant except for the second-line response. Supplementary Table S2 shows the results of the univariable analyses of clinical factors associated with survival outcomes.

Discussion

In this retrospective study, we found that initial response to chemotherapy was noted in patients with advanced-stage DSRCT from first-line to subsequent lines of chemotherapy, but the response duration for each successive line of chemotherapy became shorter as the disease progresses. The DCR after first-, second-, and third-line chemotherapy in this study was 100%, 89%, and 75%, respectively, and the median TTP for each line was 9.9, 3.5, and 2.5 months, respectively.

Currently, a multimodal approach including chemotherapy, aggressive surgery, and other adjunctive methods, such as radiotherapy and HIPEC, is widely accepted for the management of DSRCT. Improved survival outcomes have been reported after complete tumor resection combined with perioperative chemotherapy. However, earlier studies have found that complete resection was not possible in a majority of cases, and even in completely resected patients, long-term survivors were rare due to frequent recurrence. Consistent with these reports, only 2 out of 14 patients underwent surgery with the intent of macroscopic complete resection in this study, and these patients showed the best survival outcomes (although 1 of these 2 patients did not achieve “complete” resection because small metastatic lymph nodes were not removed). However, all patients eventually experienced disease progression or recurrence, including the patient who achieved complete resection and proceeded to receive adjuvant chemotherapy.

In circumstances where complete resection is not feasible, few studies have documented the effectiveness of palliative chemotherapy alone. An early report by Kushner et al showed a favorable response rate with an intensive alkylating agent-based P6 protocol in a first-line setting, achieving a DCR > 90%. Other retrospective studies have shown DCRs of >90% and the disease-free survival of 8 months using various regimens including the P6 protocol in a first-line setting. Our results were consistent with these findings with a DCR of 100% and a TTP of 9.9 months. Because all patients received chemotherapy in this study, it was not possible to directly estimate the survival benefit of chemotherapy in this study. A prior retrospective analysis of 187 patients suggested the clinical benefit of chemotherapy by identifying an association between better chemotherapy response and improved survival outcome, both in a neoadjuvant setting and the overall chemotherapy-treated population. In line with these results, our patients tended to have somewhat longer OS with better overall responses to first- to third-line chemotherapy regimens, although these differences were not statistically significant, possibly due to the small sample size.

Evidence as to whether or not chemosensitivity is retained beyond first-line chemotherapy, let alone its survival benefit, is scarce. In a small retrospective study of 41 patients in the United Kingdom where most patients were treated with cytotoxic chemotherapy, median TTP of second-line and third-line chemotherapy regimens was 2.3 and 1.1 months, respectively. This is comparable with our results, where the median TTP was 3.5 months and the median TTP was 2.5 months. The duration of response became shorter after each line of chemotherapy despite initial response to subsequent lines of chemotherapy. Considering most patients present with symptoms due to tumor mass, subsequent lines of chemotherapy might lead to temporary clinical benefits in terms of symptom palliation. Therefore, treatment decisions should be individualized based on potential risks and benefits.

Despite recent improvements in outcomes with multimodal approaches, the survival of patients with advanced DSRCT is still limited. Therefore, the development of novel, effective, and durable regimens is necessary. Several studies have explored new chemotherapeutic options for DSRCT. A phase II trial of imatinib mesylate for DSRCT with activated platelet-derived growth factor receptor (PDGFR) expression failed, and antiangiogenic agents have only shown a limited benefit.

A recent study also showed interesting results of irinotecan, temozolomide, and bevacizumab combination chemotherapy, which yielded a 3-year OS of 61%. Pazopanib as a subsequent line of chemotherapy resulted in a DCR of 78% and a median PFS of 9.2 months in a multicenter retrospective study, although we did not observe a meaningful clinical response in the limited number of patients treated with pazopanib as fourth-line chemotherapy in this study. The benefit of immunotherapy in DSRCT has been rarely reported, which may be related to the low immunogenicity of the disease. However, several clinical trials of newer immunotherapeutic agents such as enoblituzumab and Omburtamab, the combination of ipilimumab and nivolumab, and chimeric antigen receptor-T cell therapy are currently planned or in progress.

This study had several limitations due to the small sample size and the single-center retrospective design. However, to the best of our knowledge, there are few studies demonstrating the effectiveness of subsequent lines of chemotherapy in DSRCT, especially in Asian population. We believe that our findings could provide useful information on the real-world clinical effectiveness of palliative chemotherapy for the treatment of advanced DSRCT. Also, although our study focused on evaluating the effectiveness of the palliative chemotherapy, it should be noted that a multimodal approach including combination chemotherapy as well as radiotherapy and complete surgical resection of the tumor whenever possible is important in achieving long-term disease control in DSRCT.

In conclusion, although advanced DSRCT may initially respond to subsequent lines of chemotherapy beyond first-line treatment, the response becomes less durable as the disease progresses. Therefore, individualized treatment decision focused on palliation should be made.
Author Contributions

HJ analyzed the patient data and drafted the manuscript. YSH, Y-HK, CWK, and SYS contributed to the interpretation of the results and revised the manuscript. JSS and K-JC performed the histological examination, JEK and J-HA supervised the current study and revised the manuscript. All authors read and approved the final manuscript.

Availability of Data and Materials

All data analyzed during this study are included in this published article and its supplementary information files.

Research Ethics and Patient Consent

This study was approved by the institutional review board of Asan Medical Center (approval number: #2018-0751) and was performed in accordance with the ethical standards of the institutional research committee and the latest Helsinki declaration. The requirement for informed consent for this study was waived because all data were retrieved from our de-identified retrospective sarcoma registry from the Center for Cancer Data Management of the Asan Cancer Institute.

ORCID iD

Hyeyun Jeong https://orcid.org/0000-0001-7277-6463

Supplemental Material

Supplemental material for this article is available online.

REFERENCES

1. Gerald WL, Rosai J. Case 2. Desmoplastic small cell tumor with divergent differentiation. Pediatr Pathol. 1989;9:177-183.
2. Gani F, Goel U, Canner JK, Meyer CF, Johnston FM. A national analysis of patterns of care and outcomes for adults diagnosed with desmoplastic small round cell tumors in the United States. J Surg Oncol. 2019;119:880-886.
3. Honore C, Delhorme JB, Nassif E, et al. Can we cure patients with abdominal desmoplastic small round cell tumor? results of a retrospective multicentric study on 100 patients. Surg Oncol. 2019;29:107-112.
4. Stiles ZE, Dickson PV, Glazer ES, et al. Desmoplastic small round cell tumor: a nationwide study of a rare sarcoma. J Surg Oncol. 2018;117:1579-1576.
5. Lerttiiri CK, Garcia-Filion P, Hingorani P. Incidence and outcomes of desmoplastic small round cell tumor: results from the surveillance, epidemiology, and end results database. J Cancer Epidemiol. 2014;2014:680126.
6. Morani AC, Barthala TK, Surabhi VR, et al. Desmoplastic small round cell tumor: imaging pattern of disease at presentation. AJR Am J Roentgenol. 2019;212:W45-W54.
7. Scheer M, Vokuhl C, Blank B, et al. Desmoplastic small round cell tumors: multimodality treatment and new risk factors. Cancer Med. 2019;8:527-542.
8. Gerald WL, Miller HK, Battifora H, Miettinen M, Silva EG, Rosai J. Intra-abdominal desmoplastic small round-cell tumor. Report of 19 cases of a distinctive type of high-grade polyphenotypic malignancy affecting young individuals. Cancer. 1989;64:1219-1226.
9. Ladanyi M, Gerald W. Fusion of the EWS and WT1 genes in the desmoplastic small round cell tumor. Cancer Res. 1994;54:2837-2840.
10. Biegel JA, Conard K, Brooks J. Translocation (11;22)(p13;q12): primary change in intra-abdominal desmoplastic small round cell tumor. Genes Chromosomes Cancer. 1993;9:119-121.
11. Rodriguez E, Sreekantaiah C, Gerald W, Reuter VE, Motzer RJ, Chaganti RS. A recurring translocation, t(11;22)(p13;q12), characterizes intra-abdominal desmoplastic small round-cell tumors. Cancer Genet Cytogenet. 1993;69:17-21.
12. Xu J, Bulbul A, Kashad S. Potential therapeutic genomic alterations in desmoplastic small round blue cell tumor. J Clin Oncol. 2016;34:299-304.
13. LaL DR, Su WT, Wolden SL, Loh KC, Modak S, La Quaglia MP. Results of multimodal treatment for desmoplastic small round cell tumors. J Pediatr Surg. 2005;40:251-255.
14. Kushner BH, LaQuaglia MP, Wollner N, et al. Desmoplastic small round cell tumor: prolonged progression-free survival with aggressive multimodality therapy. J Clin Oncol. 1996;14:1526-1531.
15. Weeler LH, Delaney TF, Tiokos M, et al. Eosinophil and eosinophil plus vincristine, doxorubicin, and cyclophosphamide for newly diagnosed Ewing’s sarcoma family of tumors. Cancer. 1996;78:901-911.
16. Kolb EA, Kushner BH, Gielick R, et al. Long-term event-free survival after intensive chemotherapy for Ewing’s family of tumors in children and young adults. J Clin Oncol. 2003;21:3423-3430.
17. Hayes-Jordan A, LaQuaglia MP, Modak S. Management of desmoplastic small round cell tumor. Semin Pediatr Surg. 2016;25:299-304.
18. Honore C, Amrout K, Vilicot L, et al. Abdominal desmoplastic small round cell tumor: multimodal treatment combining chemotherapy, surgery, and radiotherapy is the best option. Ann Surg Oncol. 2015;12:1073-1079.
19. Subbiah V, Lamhamedi-Cherradi SE, Cuglievan B, et al. Multimodality treatment of desmoplastic small round cell tumor: chemotherapy and complete cytoreductive surgery improve patient survival. Clin Cancer Res. 2018;24:4865-4873.
20. Wong HH, Hatcher HM, Benson C, et al. Desmoplastic small round cell tumour: characteristics and prognostic factors of 41 patients and review of the literature. Clin Sarcoma Res. 2013;3:14.
21. De Sanctis R, Bertuzzi A, Bisogno G, et al. Imatinib mesylate in desmoplastic small round cell tumors. Cancer Genet Cytogenet. 1993;78:901-911.
22. Betrian S, Bergeron C, Blay JY, et al. Antiangiogenic effects in patients with progressive desmoplastic small round cell tumor: data from the French national registry dedicated to the use of off-labeled targeted therapy in sarcoma (OUTC’s). Clin Sarcoma Res. 2017;7:10.
23. Magnan HD, Price A, Chou AJ, et al. A pilot trial of irinotecan, temozolomide and bevacizumab (ITB) for treatment of newly diagnosed patients with desmoplastic small round cell tumors (DSRCT). J Clin Oncol. 2017;35:11005.
24. Freeza AM, Benson C, Judson IR, et al. Pazopanib in advanced desmoplastic small round cell tumours: a multi-institutional experience. Clin Sarcoma Res. 2014;4:7.
25. A phase II of Nolivumbol Plus ipilimumab in non-resectable sarcoma and endometrial carcinoma. https://ClinicalTrials.gov/show/NCT02982486.
26. A study of the drug I131-omburtamab in people with desmoplastic small round cell tumors and other solid tumors in the peritoneum. https://ClinicalTrials.gov/show/NCT04022213.
27. EGFR806 CAR T cell immunotherapy for recurrent/refractory solid tumors in children and young adults. https://ClinicalTrials.gov/show/NCT03618381.