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Original research

Multimodal treatment of abdominal and pelvic desmoplastic small round cell tumor with relative good prognosis

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HIGHLIGHTS

- Retrospective analysis of clinicopathological and prognosis data of 18 patients.
- Multivariable and univariate analysis for survival outcomes.
- Sex, tumor localization, and treatment positively affected patient outcomes.
- Age and symptoms or signs did not positively affect patient outcomes.
- Operation can significantly improve the survival outcomes.

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ABSTRACT

Objective: To investigate the clinicopathologic features and survival outcomes of desmoplastic small round cell tumor (DSCRT).
Methods: The retrospective cohort study was performed on clinical and pathological data of 18 DSCRT patients. Among them, two subgroups were classified according to treatment modalities. 10 cases underwent operation and adjuvant chemotherapy (group 1, 10/18, 55.6%) and 8 cases were diagnosed by fine needle aspiration biopsy without surgical intervention (group 2, 8/18, 44.4%). All cases received six courses of multiple agents chemotherapy.
Results: All cases were histologically confirmed as DSRCT and Cox regression revealed that sex, tumor localization and treatment modality affected patient outcomes. Kaplan–Meier analysis revealed that the median survival time was 22.0 ± 4.0 mo in group 1 versus 9.0 ± 0.7 mo in group 2.
Conclusion: DSRCT is highly aggressive malignance with poor prognosis, surgical excision with combination of chemotherapy can significantly improve the survival outcomes.

1. Introduction

Desmoplastic Small Round Cell Tumor (DSRCT) is a rare, highly aggressive and malignant neoplasm which mainly occurs in children and young adult men. It was first reported by Gerald and Rosai in 1898 [1]. It mainly involves the abdominal and/or pelvic peritoneum with widespread intra-abdominal involvement [1,2]. The treatment of DSRCT remains a clinical challenge and the prognosis of the disease is poor [3]. The overall survival of DSRCT is approximately 30%–55% despite chemotherapy, radiotherapy, and aggressive surgical resection [4,5]. DSRCT is commonly fatal with a median survival of 2.5 years in spite of new advances in chemotherapy and stem cell transplantation [6–8].Here, treatment and prognosis data of 18 DSRCT cases were retrospectively analyzed to highlight the modalities of treatment.

1.1. Patients and methods

In our series, 14 males and 4 females were enrolled between March 2002 and May 2013 with the median age of 29.5 ± 12.4 years (ranged from 6 to 54 years).The clinical, pathological and radiological imaging data were available for review. Based on their

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treatment modality, two subgroups were defined: group 1 (surgery group, 10 cases) who underwent exploratory laparotomy and followed by cytoreductive or debulking surgery in eight cases which lesion was distributed in multiple organs including peritoneum, mesentery, pelvic cavity and found as multiple nodes in different sizes. By contrast, en bloc resection was performed only in two cases. In group 2 (nonoperation group, 8 cases), all cases were treated with multiple chemotherapy agents following four to ten courses of multiple agents chemotherapy until tumor progression. Subsequently, all cases were underwent adjuvant chemotherapy. In both groups, the chemotherapy scheme was based on the intensive use of the IVC scheme (ifosfamide 3 g/m^2 days 1 and 2, vincristine 1.5 mg/m^2 day 1, and cisplatin 100 mg/m^2 days 1–3) or vincristine (1.5 mg/m^2, day 1), ifosfamide (3 g/m^2/day × 3), and doxorubicin (30 mg/m^2/day × 2) for four to ten cycles. Survival outcomes were estimated using the Kaplan–Meier method and compared between groups by the use of log-rank test. Multivariable Cox regression analyses addressed the association among age at diagnosis, sex, symptoms or signs, tumor localization, and treatment. *P* value <0.05 was considered to indicate statistical significance, all statistical tests were carried out utilizing SPSS, version 18.

2. Results

2.1. Clinical manifestations

The clinical and radiological characteristics of all patients are summarized in Table 1.In our series, ten cases presented with gastrointestinal symptoms including abdominal pain and distension, with or without nausea and vomiting. Solid tumors were detected in eleven cases during physical examination. Solid masses were detected during digital rectal examination in two cases. One case presented with palpable supraclavicular enlarged lymph node and the other two cases were asymptomatic. Serum CA125 was examined in seven cases and only two cases were increased significantly about 110 U/ML and 124 U/ML respectively (normal range is 1.9 ~ 16.3 U/ML).

2.2. Image analysis

Patients in this study all performed CT examination the striking character were concurrent metastases especially abdominopelvic multiple omental, serosal, or mesenteric masses. The tumors were found with heterogeneous signal mass in abdomen, pelvis, retroperitoneal (n = 16), predominantly intraperitoneal (n = 13), and presented multiple tumor nodules (Fig. 1). Diameter of entity masses in abdomen and retroperitoneal was 1 ~ 4 cm, and the average CT value was 25 ~ 45 HU. Nodular tumors didn't show marked enhancement. CT also showed serosal tumor implants and intraperitoneal spread, and 5 cases' tumors located in the omentum and/or paravesical and pararectal region. What's more, two cases complicated with hydronephrosis (unilateral in one case, bilateral in one case), two cases presented liver matastasis (Fig. 2), and two cases presented entity mass on mediastinum and pleura. Moreover, these disseminated hypoattenuating nodules can occur at other non-serosal surfaces with variable sizes, none of these masses had a definite organ origin.

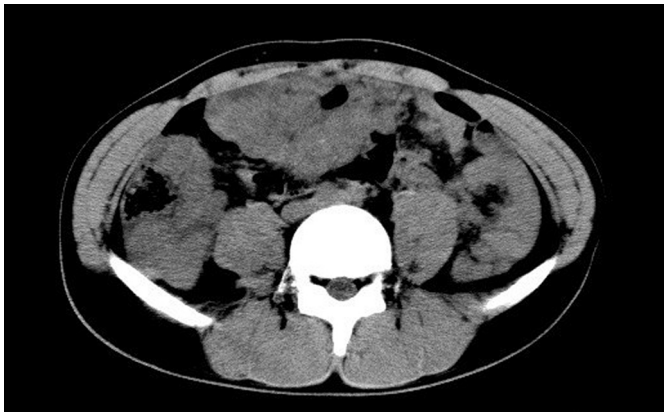

Fig. 1. Abdominopelvic CT scan revealed diffuse multiple tumor nodules in peritoneal and mesenteric surfaces.

Table 1
Clinical and CT characteristics of abdominal and pelvic DSRCT.

	Sex/age (years)	Symptoms/Signs	Tumor localization	Treatment	Survival (months)
1	M/27	Abdominal pain, nausea, palpable mass	Omentum, mesentery pelvic	Debulking surgery, chemotherapy	22
2	M/14	Distention, vomiting	Intestine, abdomen	Debulking surgery, chemotherapy	19
3	F/24	Bosom frowsty, chest pain	Abdomen, subxyphoid	Debulking surgery, chemotherapy	7
4	M/38	Abdominal pain	Retroperitoneum, abdomen	Cytoreductive surgery, chemotherapy	36
5	M/49	No symptom	Abdomen	En bloc resection, chemotherapy	Follow up 36, alive
6	F/16	Nausea, distention, abdominal pain	Omental and serosal surfaces	Cytoreductive surgery, chemotherapy	13
7	M/54	Distention, palpable mass	Intestine, abdomen	Debulking surgery, chemotherapy	40
8	M/30	Abdominal pain, hydronephrosis	Mesentery, pelvic concurrent metastasis	Cytoreductive surgery, chemotherapy	24
9	M/18	Vomiting	Intestine, mesentery	En bloc resection, chemotherapy	Follow up 44, alive
10	M/29	Abdominal pain, urinary retention	Omentum, mesentery pelvic, liver	Debulking surgery, chemotherapy	18
11	M/36	Bosom frowsty, chest pain, cough	Abdomen, involving mediastinum and pleura	Radiochemotherapy, chemotherapy	10
12	F/27	Nausea, vomiting, palpable mass, hydronephrosis	Intestines, mesentery, paravesical	Radiochemotherapy, chemotherapy	6
13	M/6	Abdominal pain	Abdomen	Chemotherapy	12
14	F/38	Distention	Abdomen, retroperitoneum	Chemotherapy	5
15	M/16	no symptom	Omentum	Chemotherapy	25
16	M/40	Abdominal pain	Intestine, paravesical	Chemotherapy	9
17	M/31	Abdominal pain, palpable mass	Mesentery, intestines	Radiochemotherapy, chemotherapy	9
18	M/34	Chest pain	Mesentery, involving pleura, liver	Chemotherapy	8

M: male, F: female.