

# Primary abdominopelvic desmoplastic small round cell tumor: CT and correlated clinicopathologic features

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**Abstract. – OBJECTIVE:** To evaluate the CT characteristics of primary abdominopelvic desmoplastic small round cell tumor (DSCRCT) and investigate the relation between radiologic features and corresponding clinicopathologic features.

**PATIENTS AND METHODS:** A cohort study was performed on 12 abdominopelvic DSCRCT patients, the preoperative computed tomography (CT) and contrast enhancement CT scan were performed in all cases. Tumor dimension, location, calcification, organs involvement, metastasis and enhancement characteristics were retrospectively evaluated and catalogued. Histopathology and serial immunological histological chemistry (IHC) studies were as diagnostic reference standard, all clinicopathological and radiological data were analyzed with emphasis on the corresponding imaging findings.

**RESULTS:** Abdominopelvic DSCRCT mainly affects young males (male to female was 2:1), Predominantly, two individualized CT subtype patterns were noted according to its characteristic features and the most common imaging findings are extensively disseminated masses in the peritoneal cavity and/or mesentery with slight enhancement after administration of contrast (subtype 1, 9/12; 75%), the type was in correlated with the histopathologic findings of a large stromal component and scare of vessels or tumor cells. In subtype 2 (3/12; 25%), the tumor was solitary and bulky soft-tissue mass localized in retroperitoneum or retrovesical space, it manifested as heterogeneous enhancement which correlated well with the presence of abundance of microvessels and tumor cells.

**CONCLUSIONS:** Radiologically, abdominopelvic DSCRCT is lack of pathognomonic CT character, the most common CT finding is multiple soft tissue masses or solitary bulky lesion

inclined to extensively peritoneal and mesenteric spread with heterogeneous enhancement. These radiological findings are related to different histological compositions, awareness of these radiological features may facilitate the CT diagnosis.

*Key Words:*

Desmoplastic small round cell tumor, Pathology, CT.

## Introduction

Desmoplastic Small Round Cell Tumor (DSRCT) is an extremely rare, highly aggressive and malignant neoplasm initially reported by Gerald et al in 1898<sup>1</sup>. DSRCT mainly occurs in adolescents and mostly involving the abdominal and/or pelvic peritoneum and the final diagnosis was determined by histological and immunohistochemistry studies<sup>2</sup>. It has been described in a limited literature which focus on the radiologic-pathologic correlation<sup>3-9</sup>. Here, we described 12 cases of abdominopelvic DSRCT and analyzed its clinical, radiological and bio-pathological features, highlighting the pathological findings and corresponding CT features.

## Patients and Methods

This study was approved by our Institutional Review Board as a retrospective study which does not require informed consent from pa-

tients. It was performed on 12 histologically confirmed DSRCT cases between March 2003 and May 2013. The clinical data, pathological and radiological imaging studies were available for review. Helical CT scan using a slice thickness of 5 mm obtained before and after injection of iodinated contrast material was performed in all patients. Two individual radiologists reviewed the following CT image parameters: dimension, morphology, enhancement, lesion numbers and location of dominant mass, presence of metastases, distribution of intraabdominal soft tissue masses. The site of the largest mass was used to define as the primary site of the disease and tumor margins were judged by extent of tissue involvement or invasion to surrounding tissue, all cases were classified according to their type of margin, internal architecture, presence of calcification and homogeneity on enhancement scan.

## Results

### Clinical Data

In our series, the median age was  $26.4 \pm 8.4$  years with a range of 14-39 years and the ratio of male-to-female was 2:1. The most common clinical manifestations and positive physical examination findings including nausea and vomiting or distention (n=8), palpable abdominopelvic mass (n=6), and urinary disorders (n=2), superficial enlarged lymph nodes in addition to intraabdominal involvement (n=1) and no positive findings (n=1). General information, treatment modalities and follow-up data are listed in z I.

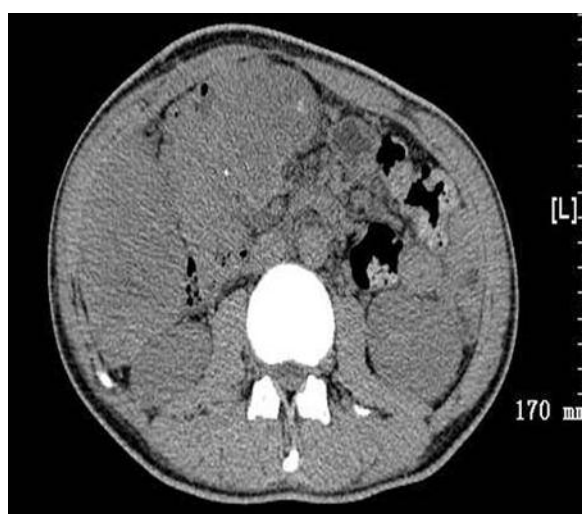
### Image Findings

In this series, all patients performed abdominopelvic CT examination and the striking character were concurrent metastases especially abdominopelvic multiple omental, serosal, or mesenteric masses. The tumors were predominantly intraperitoneal (n=7), CT also showed serosal tumor implants and intraperitoneal spread (Figures 1, 2), tumors located in the omentum and/or paravesical and pararectal region (n=5) (Figure 3). Secondary liver involvement and pleural effusion were also noted (n=2) (Figures 4, 5). Two patients had hydronephrosis (unilateral in two cases, bilateral in one) and variable bowel dilatation due to partial bowel obstruction (n=3). Areas of central low attenuation within tumors were seen in



**Figure 1.** Axial enhanced CT revealed diffuse soft-tissue nodularity masses with intraperitoneal peritoneum implants and mesenteric spread.

4 patients and scattered foci of amorphous or punctuate calcification were present in two cases (25%). On contrast enhancement CT scan, the degree of modest enhancement (n=4), obvious enhancement (n=3), slight or without enhancement (n=5) (Figure 6). According to striking CT features, the abdominal and pelvic lesions of the 12 cases were categorized into two groups, ie, the most common imaging finding of DSRCT was multiple intraperitoneal nodular soft-tissue masses in variable sizes with mesenteric and peritoneal spread (group1, n=9, mean number, 8.7; range, 3-24). Moreover, these disseminated hypoattenuating nodules can occur at other non-serosal surfaces with variable sizes

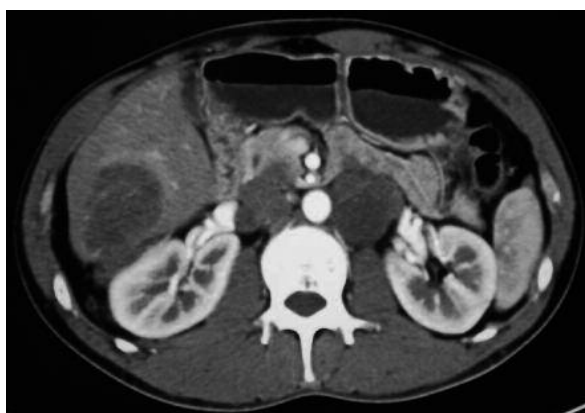


**Figure 2.** Axial unenhanced CT also showed multiple confluent solid mass inseparable from the bowel with intraperitoneal implants and lymphadenopathy



**Figure 3.** Axial unenhanced pelvic CT image showed lobulated large tumors located in paravesical and/or pararectal region displacing the rectum laterally.

and the average CT value was about 25-45HU, none of these masses had a definite organ origin. The majority of nodules are in close proximity to the mesentery or metastatic spread to the omentum. Isolated dominant mass with relatively well-defined margin (group 2, n=3; with a mean diameter of 6.8 cm, range, 4-16 cm). Punctate of calcification, central areas of low attenuation and no organ preference has been noted. Liver was one of the most common metastatic site, ascites and lymphadenopathy were also noted in three advanced cases. All dominant tumors displayed heterogeneous enhancement after IV contrast administration in group 2, the lesions demonstrate heterogeneous internal vascularity. Nevertheless, no case revealed enhancement in group 1. We noted a correlation between the



**Figure 4.** Axial enhanced abdominal CT revealed bilateral low attenuation retroperitoneal nodes with liver involvement without enhancement.



**Figure 5.** Axial enhanced abdominal CT revealed extensive peritoneum and pleural thickening with effusion and metastatic disease.

presences of different enhancement with pathological findings, areas of low attenuation and non-enhancement are often present representing necrosis or abundant of fibrous components and absent of tumor cells.

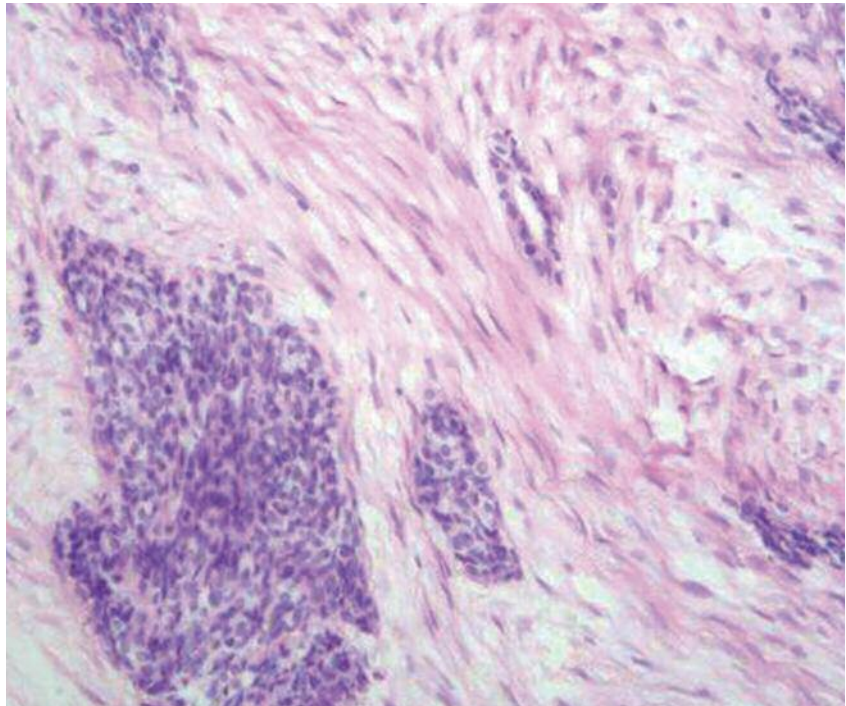
#### **Pathology Findings**

On gross inspection, the lesion revealed the presence of nonuniform white-gray multinodules. All cases were diagnosed finally by pathology which reported that tumor cells were round or oval shape with thick nuclear chromatin and few cytoplasm. It also presented distinct edges and cells like a nest and many fibrous connective tissues around the nest (Figure 7) and accompanied with necrosis and bleeding (Figure 8). Immunoperoxidase stain in these cases were posi-



**Figure 6.** Axial abdominal enhanced CT scan revealed most masses were slightly enhanced.

**Figure 7.** Nests of tumor were composed of uniform small cells with round hyperchromatic nuclei and clear cytoplasm, many fibrous connective tissues around the nest (haematoxylin-eosin stain,  $\times 200$ ).

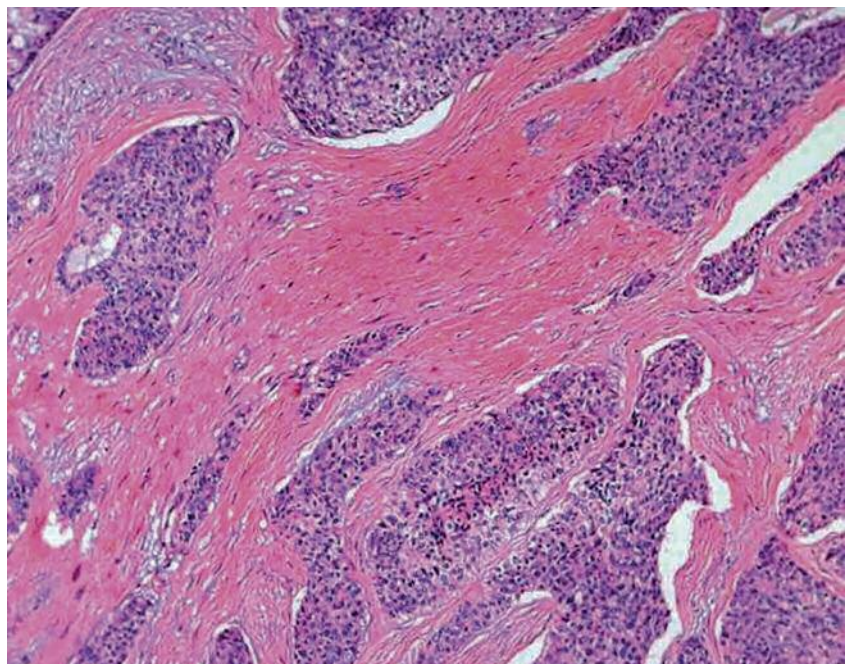


tive for EMA, keratin, desimin, vimentin, NSE, PCK, Leu-7, SMA and negative for S-100, CMA, 34BE-12.

#### ***Therapeutic Modality***

The combination of aggressive treatments such as debulking surgery and polychemotherapy. The

complete resection is rarely possible as to extensive dissemination of most abdominopelvic DSRCT and then aggressive surgical debulking is the mainstay of the therapeutic strategy. In our series, 6 cases underwent surgical debulking and adjuvant chemotherapy (Group 1, 6/12, 50%). The other 6 cases diagnosed by fine needle aspi-



**Figure 8.** Pathology investigation showed nests of small and round undifferentiated cells separated by desmoplastic stroma with necrosis stroma and bleeding.

ration or exploratory laparotomy biopsy as to extensive distant metastasis and widespread dissemination. (Group 2, 6/12, 50%), all cases received four to six courses of multiple agents chemotherapy respectively.

### Prognosis

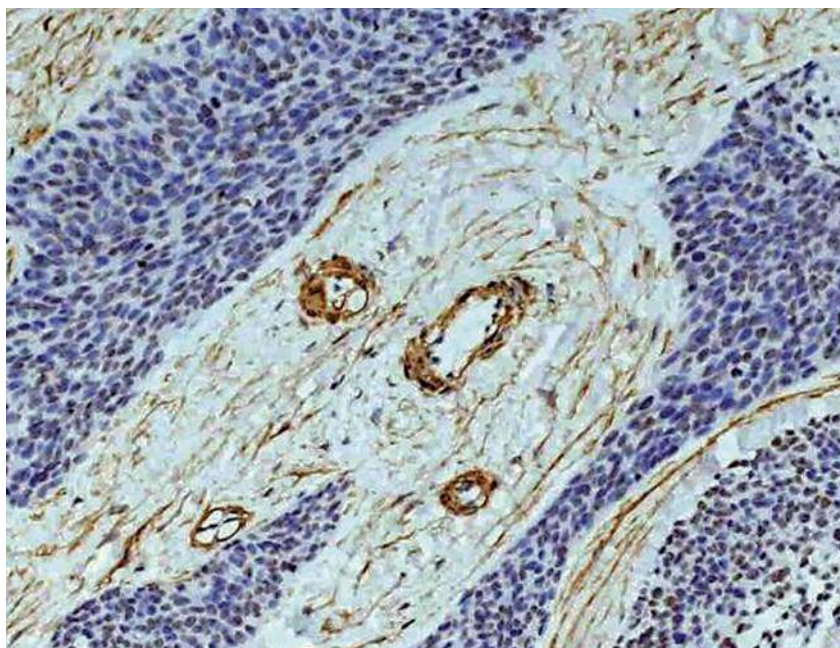
In group 1, all patients died of tumor progression or widespread metastases after diagnosis. In group 2, two patients died of tumor relapse or widespread metastases 22, 24 and 40 months after diagnosis. Only one with complete tumor resection was alive without residual tumor with a follow-up for 36 months after surgery. Complete tumor resection was an independent prognostic factor and significantly correlated with long survival. Kaplan-Meier analysis revealed the 3-year survival was 50% in group 1 vs 16.7% in group 2 ( $p < 0.05$ ) (Figure 9).

### Discussion

Due to the less knowledge of its biological behavior, it is rarity of DSRCT, meanwhile the pathogenesis of DSRCT is unclear. Histologically, the majority of DSRCT are distinguished by solid clusters of undifferentiated small round cells embedded in dense desmoplastic stroma<sup>10</sup>. These tumors are also characterized by polyphenotypic differentiation as evidenced by immuno-

histochemical staining for epithelial, mesenchymal, and neural markers including cytokeratins (EMA, AE1/3), desmin and vimentin, and neuron-specific enolase<sup>11</sup>. DSRCT belongs to the family of “small round cell tumors”; nevertheless, based on histological evaluation alone, it can be difficult to distinguish DSRCTs from other small round cell tumors. Immunocytochemical staining is useful in differentiating these malignancies. The genetic characterization of DSRCT is a chromosomal translocation of t(11;22)(p13;q12) between the Ewing’s sarcoma (EWS) gene on chromosome 22 and the Wilm’s tumor (WT1) gene on chromosome 11, leading to a EWS-WT1 fusion transcript, the characteristic translocation t(11;22)(p13;q12) is specific for DSRCT, regardless of its site. This fusion product causes a loss of the tumor suppressor function of WT1 and a putative upregulation of various families of growth factors from the EWS gene<sup>12</sup>.

DSRCTs mainly affect young adolescents with a male-to-female ratio about 4:1 and symptomatic presentation depending on tumors location. The tumor has a predilection for the surface of the omentum, mesentery of bowel or pelvis peritoneum, the disease can also occur at other non-serosal surfaces. In the early stage of DSRCT, the tumor appeared as single or multiple nodules. Concurrent metastases, particularly involving pleura and lung are common at the time of diag-



**Figure 9.** Immunoperoxidase stain revealed that the tumor cells were positive for SMA.

nosis. Digestive organs involvement is secondary, liver is one of the most common sites for metastatic disease beyond the peritoneum. Clinically, patients may be asymptomatic for long time and clinical presentation comes when the tumor burden is large. Some abdominopelvic DSRCTs present with persistent, nonspecific abdominal symptoms such as discomfort or distention, constipation, bowel obstruction, nausea or emesis, weight loss, ascites, increasing abdominal girth and palpable masses, infiltration of urinary organs may cause urinary disorders such as hydronephrosis or dysuria. Occasionally, incidental palpable abdominal masses may be the major presentation. Clinical presentation may be related to tumor size, distant metastasis and infiltration of the surrounding structures. Although it can develop at various sites, most cases usually present with widespread abdominopelvic serosal involvement which growth pattern closely mimics that of mesothelioma. It is speculated that the cell origin of DSRCT may be a primitive mesothelial cell. Gerald et al<sup>13</sup> statistic 109 DSRCT patients and mostly occurred in intra-abdominal cavity, occasionally isolated case in perididymis, pleura, posterior fossa and hands. Typical cases of DSRCT in the intra-abdominal cavity or gastrointestinal tract are accompanied by abdominal mass and/or pain which are similar to other gastrointestinal tumors. Ascites and solitary to multiple metastases nodules can also present in some patients. Urinary tract symptoms were often caused by ureteral or bladder involvement, occasionally, DSRCT can originate from the genitourinary tract system<sup>14</sup>.

It is the most widely used that the CT scan and it is preferred diagnostic modality for initial diagnosis and for follow-up assessment, hallmark features are a spectrum of multiple nodules or lobulated confluent peritoneal masses with irregular contour located within mesentery, omentum and paracolic gutter or along peritoneal surfaces which lack of organ base, and this appearance is attributable to widely spread of DSRCT. Although these findings are nonspecific, tumors without an apparent primary organ-based distribution can be suspicious for DSRCT especially in young adults.

In our series, abdominopelvic space was the commonest site at presentation, most patients frequently presented with extensive and multiple local disease and these radiographic findings must be distinguished from peritoneal carcinomatosis. Occasionally, DSRCT showed isolated and bulky heterogeneous soft tissue mass oc-

curred at non-serosal surfaces which preoperative imaging had a low diagnostic utility. Bellah et al<sup>7</sup> analyzed CT characteristics of 11 patients with DSRCT and found that most characteristic features include bulky intraabdominal soft-tissue masses that involve omental and serosal surfaces, without a distinct organ of origin and widespread implant of the tumor. In the early stage of abdominopelvic DSRCT, the tumor appeared as single or multiple nodules, nevertheless, advanced DSRCT often develop into bulky and multiple masses, particularly involving and displacing the neighboring organs with concurrent metastases, liver and pleura are commonly affected at the time of diagnosis. In our series, the most common character was 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 intraabdominal DSRCT tumor<sup>15</sup>. The most useful radiographic method is CT scan especially with intravenous contrast. In our initial investigation results, the imaging characteristics of DSRCTs depend on their contents, a striking feature of our study which not emphasized in previous reports was the presence of abundant fibrous connective tissues or stromal composition in relationship with slight or no enhancement on CT scan; we speculate it might relate to minimal vessel supply compared with abundant tumor cells type. Non-enhancement areas often represent necrosis, hemorrhage or fibrous components. The solid components may demonstrate mild enhancement on arterial phase images and is related to the densely desmoplastic stroma tissue. In enhancement cases, we speculated that this may be caused by densely packed tumor cells and vascularity.

The differential diagnosis of DSRCT faces a dilemma and encompasses a wide range of neoplastic and inflammatory conditions, especially diffusely spreading entities such as desmoid tumor, malignant peritoneal mesothelioma, lymphoma, peritoneal sarcomatosis, tuberculosis, gastrointestinal stromal tumor and Castleman disease and so on<sup>16</sup>. Radiographic appearance of most primary and metastatic abdominal tumors are similar to DSRCT and should be considered

in the differential diagnosis, close scrutiny for tumor location especially metastatic spread along the serosal coverings, multiple peritoneal nodular with the primary origin of mesentery, omentum or retroperitoneum and scattered throughout the peritoneal cavity may help to differentiate it<sup>17</sup>.

A proper consensus about treatment has not yet been established. The treatment of DSRCT remains a clinical challenge and lack of standard treatment modalities, despite multiple treatment strategies including high-dose 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<sup>18</sup>.

Aggressiveness of DSRCT may add a surgical burden and the impact of surgical resection upon survival remains unclear owing to intra-abdominal localization, frequent multiple peritoneal implants or multifocal lesions, complete resection is usually impossible. Patients had improved survival in those who underwent gross tumor resection. Cytoreductive or debulking surgery has been performed before chemotherapy is used for symptomatic relief. However, its impact on survival is uncertain. Although aggressive surgical intervention may result in better survival, adjuvant therapy remains unclear due to the high recurrence and rarity of the tumor. Complete surgical excision could improve survival; however, a complete resection is very difficult sometimes and only debulking was possible. Debulking surgery in our investigation is defined as definitive removal of at least 90% of the tumor burden. Given the frequent peritoneal involvement, the patient should be performed peritonectomy. In our surgical group series, only 3 cases were totally excised, and surgical debulking of large masses in the other 3 cases had been performed. Surgical debulking has a role in symptom relief, especially those intestinal obstruction cases. Among those, 4 cases developed local recurrence or progressive disease. Many chemotherapy combinations have been tried, but the optimal scheme and generally accepted chemotherapy option have not determined at present. In previous investigation, DSRCT has been confirmed to be moderately sensitive to intensive chemotherapy, unfortunately, response duration was extremely poor<sup>19</sup>. Many aggressive combination chemotherapy regimens have been trialed in DSRCT but none have shown curative outcome.

DSRCT is too rare to establish chemotherapy guidelines on the basis of published medical literature and our initial experience. Moreover, randomized trials comparing high-dose chemotherapy or chemotherapy plus surgery to chemotherapy alone are impossible to carry out. More efforts to prolong survival and produce a symptomatic benefit are justified. In our initial case load, if the tumor is too extended to be radically excised, the patient should start chemotherapy. Unfortunately, the response of DSRCTs to conventional chemotherapy is poor or temporarily effective, its impact on overall survival remains to be determined, meanwhile the optimal chemotherapy modalities remain to be determined. The survival benefit from chemotherapy may outweigh its side effect profile.

Radiotherapy especially whole abdominopelvic external beam radiation to the entire abdomen and pelvis in DSRCT has not been used as extensively owing to its acute toxicities and low response rate. However, combination triple modality therapy has reported by Lal et al<sup>20</sup>. A total of 66 cases with DSRCT, including 29 patients (44%) underwent chemotherapy, surgery and radiotherapy and three-year survival was 55% in those receiving comprehensive treatments compared to 27% in which all three methods were not performed.

## Conclusions

No pathognomonic radiological character was noted in abdominal DSRCT, radiologically, it was difficult to differentiate from other soft-tissue tumors. CT findings are variable, the most common imaging finding is multiple soft tissue masses inclined to extensively peritoneal and mesenteric spread. Occasionally, isolated bulky mass situated in peritoneal or pelvic space with obviously heterogeneous enhancement, these features may reflect the different histological composition of the tumors, and familiarity with these different radiological features may help improve the diagnosis. Owing to a retrospective study, limited cases, nonuniform imaging protocols and absence of more histological data to confirm the conclusion is the demerits of this analysis.

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## Conflict of interest

The authors declare that they have no conflict of interest.

## References

- 1) GERALD WL, ROSAI J. Desmoplastic small cell tumor with divergent differentiation. *Pediatr Pathol* 1989; 9: 177-183.
- 2) 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. *Am J Surg Pathol* 1991; 15: 499-513.
- 3) THOMAS R, RAJESWARAN G, THWAY K, BENSON C, SHAHABUDDIN K, MOSKOVIC E. Desmoplastic small round cell tumour: the radiological, pathological and clinical features. *Insights Imaging* 2013; 4: 111-118.
- 4) ARORA VC, PRICE AP, FLEMING S, SOHN MJ, MAGNAN H, LAQUAGLIA MP, ABRAMSON S. Characteristic imaging features of desmoplastic small round cell tumor. *Pediatr Radiol* 2013; 43: 93-102.
- 5) ZHANG WD, LI CX, LIU QY, HU YY, CAO Y, HUANG JH. CT, MRI, and FDG-PET/CT imaging findings of abdominopelvic desmoplastic small round cell-tumors: correlation with histopathologic findings. *Eur J Radiol* 2011; 80: 269-273.
- 6) EIRIZ MARTÍNEZ S, CONCEIÇÃO E SILVA JP. Desmoplastic small round cell tumor of the abdomen: CT findings and radiologic-pathologic correlation in 3 cases. *Radiologia* 2009; 51: 313-317.
- 7) BELLAH R, SUZUKI-BORDALO L, BRECHER E, GINSBERG JP, MARIS J, PAWEL BR. Desmoplastic small round cell tumor in the abdomen and pelvis: report of CT findings in 11 affected children and young adults. *AJR Am J Roentgenol* 2005; 184: 1910-1914.
- 8) CHOULI M, VIALA J, DROMAIN C, FIZAZI K, DUVILLARD P, VANEL D. Intra-abdominal desmoplastic small round cell tumors: CT findings and clinicopathological correlations in 13 cases. *Eur J Radiol* 2005; 54: 438-442.
- 9) TATEISHI U, HASEGAWA T, KUSUMOTO M, OYAMA T, ISHIKAWA H, MORIYAMA N. Desmoplastic small round cell tumor: imaging findings associated with clinicopathologic features. *J Comput Assist Tomogr* 2002; 26: 579-583.
- 10) SANDBERG A, BRIDGE J. Updates on the cytogenetics and molecular genetics of tissue tumors: desmoplastic small round-cell tumors. *Cancer Gene. Cytogene* 2002; 138: 1-10.
- 11) ORDONEZ NG. Desmoplastic small round cell tumor: a histopathological study of 39 cases with emphasis on unusual histological patterns. *Am Surg Pathol* 1998; 22: 1303-1313.
- 12) KIM J, LEE J, BRANTON P, PELLETIER J. Modulation of EWS/WT1 activity by the v-Src protein tyrosine kinase. *FEBS Lett* 2000; 474: 121-128.
- 13) GERALD WL, LADANYI M, ALAVA E. Clinical pathologic and molecular spectrum of tumors associated with t(11;22)(p13;q12): desmoplastic small round cell tumor and its variants. *J Clin Oncol* 1998; 16: 3028-3036.
- 14) SU MC, JENG YM, CHU YC. Desmoplastic small round cell tumor of the kidney. *Am J Surg Pathol* 2004; 28: 1379-1383.
- 15) PICKHARDT PJ, FISHER AJ, BALFE MD. Desmoplastic small round cell tumor of the abdomen: radiologic-histopathologic correlation. *Radiology* 1999; 210: 633-638.
- 16) LEVY AD, ARNAIZ J, SHAW JC, SOBIN LH. From the archives of the AFIP: Primary peritoneal tumors: Imaging features with pathologic correlation. *Radiographics* 2008; 28: 583-607.
- 17) LYER RS, SCHAUNAMAN G, PRUTHI S, FINN LS. Imaging of pediatric desmoplastic small-round-cell tumor with pathologic correlation. *Curr Probl Diagn Radiol* 2013; 42: 26-32.
- 18) STUART-BUTTLE CE, SMART CJ, PRITCHARD S, MARTIN D, WELCH IM. Desmoplastic small round cell tumour: a review of literature and treatment options. *Surg Oncol* 2008; 17: 107-112.
- 19) LIPPE P, BERARDI R, CAPPELLETTI C, MASSACESI C, MATTIOLI R, LATINI L, CELLERINO R. Desmoplastic small round cell tumour: a description of two cases and review of the literature. *Oncology* 2003; 64: 14-17.
- 20) LAL DR, SU WT, WOLDEN SL, LOH KC, MODAK S, LAQUAGLIA MP. Results of multimodal treatment for desmoplastic small round cell tumors. *J Pediatr Surg* 2005; 40: 251-255.