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REVIEW

Desmoplastic small round cell tumour: A review of literature and treatment options

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KEYWORDS
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Summary
Introduction: Desmoplastic Small Round Cell Tumour (DSRCT) is a rare but aggressive malignancy with poor outcome.
Aims: To review the clinico-pathological features and radiological, histological and tumour markers of the disease and to evaluate the evidence for treatment options available.
Methods: We report a clinical case from our centre and have conducted a review of the literature from Medline (Pubmed) database from 1989 to 2007.
Results: DSRCT typically presents with advanced disease and is prevalent in young males. Lack of staging criteria and small numbers of patients make comparison of evidence for its treatment difficult.
Conclusion: Surgical excision is only recommended for non-metastatic disease with combination chemo-radiotherapy as an adjunct. These modalities used in isolation may have less impact. Furthermore, the side effect profile from radiotherapy may outweigh any survival benefit.
For advanced disease, symptom control is most important as these modalities impact survival minimally and palliation of secondary symptoms is paramount. Multi-disciplinary team and specialist centre review for histology and oncology are essential in managing this disease process and will enable greater numbers of patients to be enrolled into therapeutic trials and future evolving therapies.

Contents

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Introduction

Desmoplastic Small Round Cell Tumour (DSRCT) was first described in 1989 by Gerald and Rosai [1] and Ordonez et al. [2]. DSRCT is a rare and highly aggressive tumour that usually occurs in males during adolescence and early adulthood. Only a few hundred cases have been reported in worldwide literature. The male to female ratio is 4:1 [3,4].

We will first present a brief case and then a literature review of the treatment options available for this unusual disease process.

Case information

A 28-year-old male complaining of abdominal pain, weight loss and erectile dysfunction of 2 years duration, was admitted to our surgical unit. On examination he was found to be cachectic. He had a large abdominal mass arising from the pelvis, with multiple smaller nodules palpable over the lower abdomen. Digital rectal examination was obstructed by a large rectal mass.

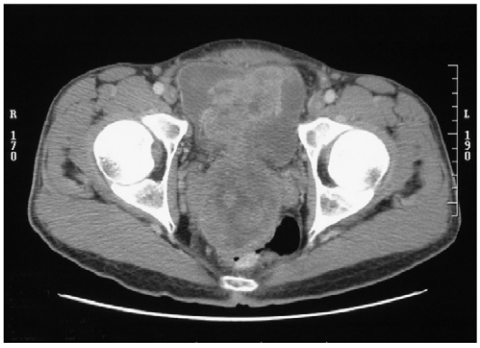

Figure 1 Abdomino-pelvic disease.

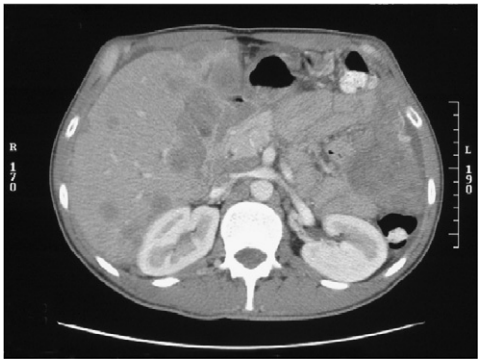

Figure 2 Liver metastases.

His blood profile revealed grossly elevated liver enzymes and a CA-125 of 328 U/ml. CT scanning suggested extensive intra-abdominal and pelvic malignancy with peritoneal deposits, liver metastases and mesenteric lymphadenopathy (see Figures 1 and 2). Histological analysis of a liver biopsy sample illustrated classical morphological features of DSRCT, and the diagnosis was confirmed by immunohistochemistry.

Clinical and pathological features

Clinical signs and symptoms of DSRCT are non-specific. DSRCT typically arises from abdominal or pelvic peritoneum as a diffuse mass, which tends to be large at presentation—up to 40 cm in some cases [5]. It is associated with abdominal distension and pain, ascites and hepatomegaly. Pressure effects of the tumour on the nearby structures have also been described, such as intestinal obstruction, hydronephrosis and urinary/erectile dysfunction (see case description above).

DSRCTs have a tendency for peritoneal and omental spread, lymph node involvement and haematogenous metastasis, especially to the liver [6]. Primary DSRCTs have been described originating outside the abdominal cavity—arising from the other mesothelial surfaces such as lung pleura and tunica vaginalis [7]. DSRCT can also arise from solid organs such as the ovaries, liver, kidneys, pancreas, bone and even the posterior cranial fossa [3,8–12].

Radiologically DSRCT is similar to the other intra-abdominal primary tumours, and this combined with the non-specific clinical nature mandates a tissue diagnosis from biopsy samples.

The rarity of this tumour and the similarities it shares with the other small round cell tumours still make diagnosis challenging. However, distinctive cytological, histological, immunocytochemical and cytogenetic profiles have all been described for DSRCT.

The key features of DSRCT are briefly outlined:

- *Histology*: Macroscopic appearances are of a bosselated outer surface, cut surfaces are grey with areas of necrosis within [5].
  Microscopically, the tumour usually forms nests or strands of small round cells embedded in desmoplastic stroma (Figure 3). The ratio of tumour cells to stroma varies significantly across a biopsy specimen. Tumour cells tend to be small to medium in size with scanty cytoplasm, round to oval nuclei and visible mitotic figures [5,13,14].
- *Cytology*: Analysis is possible from tissue biopsy and FNA specimens, as well as ascitic and pleural tap fluid [15]. This is especially useful when biopsy of the tumour bulk is

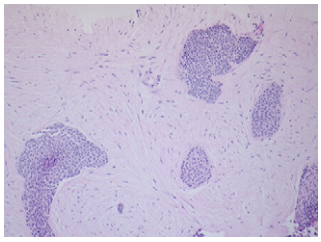

Figure 3 Small round blue tumour cells.

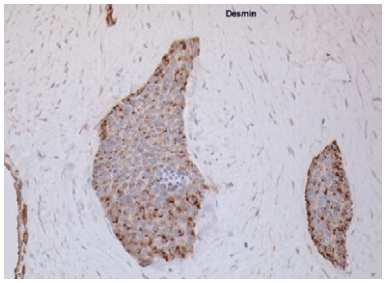

Figure 4 Staining for Desmin.

difficult. Crapanzano et al. [16] confirmed accurate diagnosis of DSRCT from cytology specimens using immunohistochemical techniques in addition to the cytomorphology.

- *Immunohistochemistry*: DSRCT cells are immunoreactive to epithelial, mesenchymal, myogenic and neural markers. They demonstrate positive staining for vimentin, desmin (Figure 4), smooth muscle actin, neuron-specific enolase, epithelial membrane antigens and cytokeratin [17,18]. This polyphenotypic antigen expression profile is unique among small round blue cell tumours.
- *Cytogenetics*: The EWS–WT1 gene fusion has more recently been discovered to be disease-specific to DSRCT [19,20]. This characteristic reciprocal translocation is demonstrated by fluorescence in situ hybridisation (FISH) or reverse transcriptase–polymerase chain reaction technique, and is specific to DSRCT at any primary location [5,17]. It is the definitive diagnostic marker.

Hybrid tumours that resemble DSRCT have been described in the literature, but these lack the characteristic EWS/WT1 gene fusion. Instead these hybrid tumours show the EWS/ERG gene fusion typical of the Ewing's sarcoma/peripheral primitive neuroectodermal tumour group [21]. Therefore, cytogenetic analysis is the most specific method of diagnosis.

Tumour markers

No blood profile abnormalities are specific to DSRCT, but as in our case, tumour markers may be elevated, especially serum CA 125 [22]. This has been reported as raised in up to 86% cases of intra-abdominal DSCRT, with a median value of 200 U/ml (range 22–735) [23]. High CA 125 levels associated with DSRCT may be related to ascites and not directly to the tumour itself [21]. Therefore, tumour markers cannot be used as diagnostic tools. Importantly, CA 125 has been shown *not* to be a good monitor of disease progression: this relies on clinical and radiological evaluation.

Treatment options

The extremely rare occurrence of DSRCT means the treatment modalities and their impact on survival have only been studied in small numbers of patients. These factors, and the young age of most patients with DSRCT, make it very difficult for clinical decisions with regard to the treatment.

In DSRCT there has not yet been a case in which treatment has led to a curative outcome. Outlined below is a summary of evidence for treatment modalities in DSRCT.

The three main treatment modalities include the following:

(1) *Surgical resection*: A complete resection is rarely possible, but excision of large peritoneal masses has been performed in groups of patients [24–31]. The complete removal of tumour deposits is usually limited by a liver metastasis, infiltration into hepatic veins or involvement of the diaphragm [24]. The impact of surgical resection upon survival remains unclear.

Biswas et al. [30] did conclude that if complete surgical excision could be achieved, then survival improved (but excision was not curative). This, however, was mainly in peripheral and extra-abdominal disease without metastatic spread. Hassan et al. [31] found that the patients with DSRCT who underwent surgical excision had a median survival of 34 months, compared with 14 months median survival in patients undergoing biopsy only. However, of the 7 patients who underwent surgery macroscopic resection was possible in 3 patients. In the other 4 patients, only debulking was possible. All these 7 patients developed recurrent or progressive disease, and a second operation was needed in 6 patients.

Similarly, Lal et al. [29] studied a cohort of 66 patients, and found gross tumour resection to be highly significant in prolonging overall survival. The 3-year survival was 58% in patients who had undergone surgical resection, as compared to a 0% 3-year survival rate in non-resectable patients. This almost certainly represents a reflection on the differing stages of disease rather than the true impact of surgery.

Conversely, various other authors have concluded that the surgical excision does *not* significantly improve survival: Livaditi et al. [27] found that among those who underwent radical tumour excision with adjuvant chemotherapy, all had tumour recurrence within 2–6 months. Again, Gil et al. [28] found no correlation between the surgical excision and improved survival rates. Other studies found a complete surgical excision not to be possible, even after chemotherapy [25,26].

However, palliative surgical debulking does have a role in symptom relief, especially when mass effects from the tumour are apparent.

(2) *Combination chemotherapy*: The response of DSRCTs to conventional chemotherapy is poor, as exemplified by early case reports in the literature. Subsequently, many aggressive combination chemotherapy regimes have been trialled in DSRCT [24–34]. Unfortunately, no curative outcome has yet been achieved, and there has been no significant impact on long-term survival.

Bertuzzi et al. [32] studied high-dose chemotherapy (ifosfamide, epirubicin and vincristine) in patients with various types of small round cell tumours. The subset with DSRCT had a very poor response to treatment (a 43% histological response versus the 85% response in the other small round cell tumour types).

Many other centres have used aggressive combination chemotherapy (alkylating chemotherapy/P6 protocol), and some tumour response to chemotherapy is usually demonstrated. The P6 protocol involves 7 courses of chemotherapy, courses 1–3 and 6 use cyclophosphamide, doxorubicin and vincristine. Courses 4, 5 and 7 are infusions of ifosfamide and etoposide.

A 39% response rate to multi-agent chemotherapy was observed in a series of 18 patients [30]. However, this again is a subset of patients with atypically early and/or peripheral disease, where surgical excision was possible also. Disease recurrence is the rule, often within 6 months of the chemotherapy [27]. There are two exceptions to this in patients who had systemic chemotherapy with good response and survived until 55 and 101 months [28]. These are the longest survivors in the literature.

(3) *Local radiotherapy*: Local radiotherapy in DSRCT has not been used as extensively as surgery or chemotherapy. Goodman et al. [35] studied 21 patients treated with combination chemotherapy and whole abdominopelvic irradiation (WAPI) with external beam radiation to the entire abdomen and pelvis at a dose of 30 Gy. Unfortunately, acute haematological and gastrointestinal toxicities occurred in $>75\%$ patients. Response to WAPI was disappointing, with a median survival of 32 months and a median time to relapse of only 19 months.

However, combination triple modality therapy has shown the best results with Quaglia et al. [29] demonstrating a 3-year survival of 55% in patients receiving chemotherapy, radiotherapy and surgery versus 27% when all the three modalities were separately examined.

Heterogeneity of the data and lack of staging criteria, together with small numbers of patients in single-centre series mean comparison of all the techniques is difficult. This coupled with lack of multi-centre trials and locally prescribed chemotherapeutic protocols means, the true outcome is difficult to measure.

Also DSRCT spans the paediatric/adult divide of medicine, thus prohibiting larger numbers of patients being reviewed in centres and there may be a true difference in outcome or response to treatment modalities in differing age groups.

Prognosis

As exemplified above, the prognosis in DSRCT is poor. Patients universally die of the disease, most often within 3 years of diagnosis. This prognosis is despite the best attempts at medical and surgical treatment.

Future therapies

Several other modalities of treatment have been used in treating DSRCTs. Mazuryk et al. [36] described the benefit of autologous stem cell support in this group of patients. The isolated case report had no control and the patient did receive aggressive multi-modality treatment with chemoradiation also. However, after the follow-up of 26 months the patient remained alive. Bertuzzi et al. [32,37] did not show a good response to the high-dose chemotherapy in DSRCTs and in 10 patients could not demonstrate a complete response conversion with EWS-WT1 positivity in all the follow-up patients having undergone a high-dose chemotherapy and stem cell support.

Newer chemotherapy regimes used effectively in rhabdomyosarcoma have had limited success in to small series of paediatric patients with DSRCTs, namely vinorelbine combined with low-dose cyclophosphamide and iritonecan as a sole agent. Their role has not been fully proven yet [38,39].

The monoclonal antibody therapies used to target novel cell surface antigens expressed in human solid tumours are well described [40]. In DSRCTs two antigens have been studied, G(D2) and the antigen to antibody 8H9. In a series of 46 DSRCT samples the antigen expression was 70% for G(D2) and 96% for 8H9 [41]. These may represent novel approaches to diagnosis and treatment, especially with anti-G(D2) showing some efficacy in treating minimal residual disease in neuroblastoma [42].

The propensity of DSRCTs in young adolescent males has provoked the study of combined androgen blockade in patients with refractory and multi-modality failure disease. Fine et al. [43] analysed the role of androgen receptors (ARs) and c-Kit in this group of patients. Immunohistochemical analysis showed positivity of 37% for AR and 35% for c-Kit and showed these receptors to be functional in DSRCT cells. Of six patients treated with anti-androgen therapy three had a clinical benefit in the post-multi-modality setting.

The most current novel therapy is the use of continuous hyperthermic peritoneal perfusion (CHPP) therapy. As part of an ongoing phase I study into the use of hyperthermic cisplatin in the treatment of refractory solid childhood tumours (ClinicalTrials.gov Identifier: N CT00436657) Hayes-Jordan et al. [44] describe two patients treated in this way. CHPP at 40 °C with cisplatinum for 90 min is the treatment protocol, but outcomes are yet to be assessed.

Conclusion

DSRCT is a rare and aggressive tumour that affects young males. It is usually an abdominopelvic malignancy that demonstrates distinct histological appearances and a unique cytogenetic profile. There have been many different approaches to the treatment of DSRCT, but unfortunately it remains incurable and has poor long-term survival rates.

However, with an aggressive approach to the treatment using multiple modalities, some temporary benefit to survival can be achieved. Patients who have combination chemotherapy (P6 protocol), tumour-shrinking radiotherapy and surgical tumour excision have the longest short-term survival rates. The role of autologous stem cell transplantation and high-dose chemotherapy remains unclear.

When a less-aggressive approach is taken with surgical intervention alone, no real long-term survival benefits have been identified except in atypical case series of extra-abdominal and peripheral disease states. Therefore, surgery should only be considered in these cases; but for more typically advanced abdomino-pelvic disease or metastases,

it should be regarded as a means of palliation or to relieve concomitant symptoms. The role of surgery may develop with changing techniques, e.g. CHPP, to become more influential in determining the outcome.

Similarly, DSRCT is generally chemo-sensitive to combination regimes, but response is short lived and there is poor survival gain.

Patient involvement in management options is essential in deciding which modality will be the most suitable and referral to a tertiary centre is advised.

In view of the evidence above, these are the following author's advises:

1. In a patient who has presented with early or peripheral disease with favourable radiological appearances and absence of metastases, it is appropriate to embark on an aggressive treatment course with a combination chemotherapy and surgical excision and local radiotherapy, so as to achieve the best possible survival benefits. The specific chemotherapeutic options are outlined above, but remain a locally determined entity.
2. Alternatively, in a patient with non-resectable disease the palliative measures need to be taken, with surgical intervention only if necessary for symptom relief (bowel obstruction, for example). Palliative chemotherapy remains controversial in this group, as the side effects of the treatment may outweigh the minimal gain in survival. Patient preference becomes of utmost importance in this situation. Certainly, radiotherapy when used as an isolated treatment may impart more toxicity than benefit.

To conclude, the current treatments for desmoplastic DSRCTs are not curative and do not impart long-term survival benefits. However, since DSRCT was identified, there have been many advances in treatment as management options are continually being refined and improved. There is still no clear evidence base for the treatment, and this is due to both the rarity and aggressive nature of this tumour and its lack of staging criteria and paediatric/adult case mix.

Thus, future patients with DSCRT need to be enrolled, wherever possible, in multi-centre trials so as to continue to advance the treatment boundaries of this rare cancer.

Conflict of Interest Statement

None.

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