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From Histopathology to Therapeutic Strategies: Perspectives in Desmoplastic Small Round Cell Tumor Management

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Abstract:

- ***Background:***

Desmoplastic small round cell tumor (DSRCT) is a rare and highly aggressive soft tissue sarcoma, typically affecting young males and characterized by the EWSR1–WT1 fusion gene. Its diagnosis is challenging due to non-specific clinical features and overlapping histological patterns, making immunohistochemistry and molecular testing essential.

- ***Case Presentation:***

We report the case of a young patient presenting with an abdominal mass. Histopathological analysis revealed a small round cell tumor with desmoplastic stroma. Immunohistochemistry was consistent with DSRCT, showing co-expression of epithelial, mesenchymal, and neural markers. Molecular confirmation of the EWSR1–WT1 fusion established the diagnosis. The patient underwent multimodal treatment including high-dose chemotherapy and cytoreductive surgery.

- ***Discussion:***

This case illustrates the critical role of immunohistochemistry in diagnosing DSRCT and the importance of an aggressive multimodal approach combining chemotherapy, surgery, and, in selected cases, locoregional strategies such as HIPEC. Nevertheless, relapses remain frequent and prognosis is dismal, highlighting the urgent need for novel therapeutic strategies, including targeted agents and immunotherapy.

- ***Conclusion:***

DSRCT is a rare but distinct entity requiring comprehensive pathological evaluation and aggressive multimodal management. Early recognition is crucial, although outcomes remain poor despite intensive therapy.

Keywords: *Desmoplastic Small Round Cell Tumor; EWSR1–WT1 Fusion; Immunohisto Chemistry; HIPEC; Targeted Therapy.*

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I. INTRODUCTION

Desmoplastic small round cell tumors (DSRCTs) are exceptionally rare and highly aggressive malignancies that mainly develop in the abdominal and pelvic regions [1]. Classified as a subtype of soft tissue sarcoma, DSRCTs were officially recognized as a distinct pathological entity in 1991, following their initial description by Gerald and Rosai [2]. Since then, the number of reported cases has been steadily increasing, with approximately 1000 to 1200 cases documented in the literature to date [3]. DSRCT is most frequently diagnosed during adolescence or early adulthood and disproportionately affects males, with a reported sex ratio of nearly four males for every female(4:1) [4,5]. From a clinical standpoint, patients typically present with presence of multiple intra-abdominal masses, often without a clearly identifiable organ of origin. These tumors frequently involve the peritoneal and serosal surfaces, leading to widespread peritoneal dissemination. The clinical symptoms are usually non-specific, including abdominal distension, discomfort, a sensation of heaviness, pain and nausea. These vague manifestations and the insidious progression often lead to a delayed diagnosis with extensive peritoneal involvement and metastatic spread. The prognosis of DSRCT remains poor,

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despite aggressive multimodal treatment that may include intensive chemothérapy, surgery, radiotherapy, the five-year overall survival rate is estimated to range between 15% and 30% [5,6]. We report a clinical case of advanced peritoneal DSRCT managed in our department — the first of its kind to be encountered in our institution. This case represented a significant diagnostic and therapeutic challenge due to the rarity and complexity of the tumor.

II. CASE REPORT

A 28-year-old man presented with abdominal pain and progressive abdominal distension that had been evolving over the past six months. The patient had no prior medical history and no family history of cancer. Physical examination revealed a distended and firm abdomen, with pain elicited on palpation. A contrast-enhanced abdominopelvic CT scan was performed, revealing findings consistent with diffuse peritoneal carcinomatosis associated with ascitic fluid. A total colonoscopy and an upper gastrointestinal endoscopy were performed, both of which revealed no abnormalities.

The patient was referred for an exploratory laparoscopy, which revealed multiple diffuse intra-abdominal masses. Several biopsies were taken during the procedure.

Histopathological examination of the biopsy specimens revealed a malignant tumor proliferation with high cellular density. The tumor was composed of sheets and clusters of small round cells with scant, weakly eosinophilic cytoplasm and round, hyperchromatic nuclei. A high mitotic activity was also noted. A broad immunohistochemical panel was performed. The tumor cells showed strong positivity for AE1/AE3 and EMA, while CD99, desmin, myogenin, CD45, S100, HMB45, melan-A, and WT1 were negative. Focal positivity was observed for CD56, synaptophysin, and neuron-specific enolase (NSE), whereas chromogranin was negative. The Ki-67 proliferation index was approximately 80%, indicating a highly proliferative tumor. A targeted transcriptomic analysis revealed an EWSR1/WT1 fusion transcript, as identified by the BED Fusion v2.2 panel.

The patient recieved 1st line of chemotherapy with alternating regimens of Doxorubicin, Cyclophosphamide, Vincristine and Ifosfamide/ Etoposide for six cycles, achieving a satisfactory clinical response. Following the recurrence of ascites, a second-line chemotherapy with the FOLFOX regimen (5-fluorouracil and oxaliplatin) was administered for 12 cycles, without evidence of response. A third-line regimen consisting of doxorubicin, cyclophosphamide, and cisplatin was then initiated, with no significant clinical benefit. The patient subsequently underwent a first session of pressurized intraperitoneal aerosol chemotherapy (PIPAC), repeated one month later, which provided only short-term efficacy, followed by a rapid reaccumulation of ascites. It was then decided to discontinue all disease-directed treatments and refer the patient to best supportive care. The patient died nine months later, resulting in a total survival time of 26 months.

III. DISCUSSION

DSRCT are particularly rare, and highly aggressive malignant neoplasms,That typically occur in adolescents and young adults [7], with a median age at diagnosis of approximately 22 years , and reported extremes ranging from 5 to 60 years. A strong male predominance is well documented, with a sex ratio of about 4:1 [5,8]. The incidence is extremely low, and due to its rarity, most data are derived from small retrospective series or case reports.

DSRCT most often arises from the serosal surfaces of the abdominal cavity, with the peritoneum being the predominant primary site [9]. Nonetheless, occurrences have also been reported in other locations, such as the thoracic cavity, central nervous system (and orbit), facial, ovary, prostate and extremities [3,5,9,10, 11].

Most patients present with an advanced stage of disease, and metastatic spread is common at diagnosis, with frequent involvement of the peritoneum, liver, spleen, and supradiaphragmatic lymph nodes. Several staging systems have been proposed; notably, Saltsman and colleagues proposed an imaging-based risk stratification system dividing patients into three categories: intermediate risk in the absence of hepatic involvement or ascites, high risk when either liver involvement or ascites is present, and very high risk when both features coexist. [12]. In our case, the patient presented with an advanced stage of disease, classified as high risk according to Saltman’s staging system. This category is associated with a significantly poorer prognosis, reflecting the aggressive nature of DSRCT and its tendency for extensive peritoneal dissemination and early visceral involvement.

On histologic examination, DSRCT demonstrates a heterogeneous differentiation pattern with epithelial, myogenic, stromal, and neural features [13]. The tumor is composed of nests of malignant cells surrounded by a prominent desmoplastic stroma. These cellular aggregates vary in morphology, ranging from rounded to elongated configurations. Neoplastic cells are generally small to intermediate in size, with round-to-oval, hyperchromatic nuclei and poorly visible nucleoli. Mitotic activity and areas of necrosis are commonly observed. The cytoplasm is generally scant with poorly defined cell borders. In larger, more pleomorphic cells, intracytoplasmic eosinophilic rhabdoid inclusions may also be observed [14].

Immunohistochemistry of DSRCT consistently demonstrates a polyphenotypic profile, with co-expression of epithelial, mesenchymal, and neural markers. Desmin (often with a dot-like perinuclear staining pattern that is considered highly suggestive of DSRCT), epithelial markers (AE1/AE3, CK7, EMA), and nuclear WT1 are the most reliable and recurrent findings, while CD99 is frequently positive. Other markers such as vimentin, NSE, or chromogranin A are inconsistently expressed. This characteristic immunoprofile is crucial for distinguishing DSRCT from other small round cell tumors (table 1).

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A hallmark of DSRCT is the presence of the EWSR1-WT1 gene fusion, resulting from the translocation t(11;22)(p13;q12) [14]. This chimeric transcript encodes an aberrant transcription factor that combines the N-terminal transactivation domain of EWSR1 with the DNA-binding domain of WT1, leading to deregulated expression of genes involved in cell proliferation, differentiation, and survival. Detection of this translocation is considered a molecular gold standard for diagnosis, as it distinguishes DSRCT from other small round cell tumors with overlapping morphology and immunophenotype [2,21]. Beyond its diagnostic relevance, the EWSR1-WT1 fusion also represents a potential therapeutic target, although effective strategies directly targeting this fusion remain lacking.

To date, there is no established standard of care for DSRCT, and treatment strategies are extrapolated from sarcoma protocols. A multimodal approach combining intensive multi-agent chemotherapy, cytoreductive surgery, and locoregional treatments such as hyperthermic intraperitoneal chemotherapy (HIPEC) or abdominal radiotherapy has been associated with improved outcomes in selected patients [22].

Although most patients with DSRCT present with advanced disease, aggressive surgical resection continues to represent a critical determinant of survival. Given the typically large tumor burden, surgery is generally preceded by high-dose neoadjuvant chemotherapy to increase the likelihood of complete cytoreduction [23] . The surgical aim is to achieve resection of more than 90% of the tumor mass , ideally leaving residual disease of less than 1 cm, as this has been correlated with improved outcomes[24]. However, this approach is best suited for patients without liver or extraperitoneal metastases, in whom debulking surgery has demonstrated survival benefits [25]. In contrast, its application in patients with extensive metastatic involvement may not provide the same advantage and can be associated with increased morbidity and mortality [26].

Table 1 : Overview of Immunohistochemical Profiles Reported in DSRCT Cases from Literature

		AE1 AE3	CK 7	CK20	Desmin	EMA	Cg A	NSE	Vimentin	CD99	WT1
Puri, Nikhil et al. (2024) [3]		+	-	-	+	ND	-	ND	ND	ND	-
Chen, Guoyong & al. (2024) [13]		+	+	-	+	ND	-	+ (partially)	+	ND	-
Granja, NM & al. (2005) [14]	***Case1***	+	-	-	+	+	+	+	+	ND	+
	Case2	+	-	-	-	+	+	+	+	ND	+
	Case3	+	ND	ND	ND	ND	ND	+	+	ND	+
Taei, Tareq Hamed Al et al. [15]		+	ND	ND	+	ND	ND	ND	ND	+ (partially)	+
Aguilera, Dolly et al. (2008) [16]		ND	ND	-	+	+	-	ND	+ (partially)	+	ND
Li, Runze & al. (2023) [17]		+	ND	ND	+	+	-	+	ND	+	+
Wang, L & al. (2021) [18]	***Case1***	+	ND	ND	+	-	-	ND	-	+	+
	Case2	+ (partially)	ND	ND	+	+ (partially)	-	ND	+	+	+
	Case3	+	ND	ND	+ (partially)	+ (partially)	-	ND	+ (partially)	+	+
	Case4	+	ND	ND	+	+	-	ND	+ (partially)	+	+
	Case5	-	ND	ND	+	+	+	ND	+	+	+
	Case6	-	ND	ND	-	+	+	ND	+	ND	+
	Case7	+	ND	ND	+	+	-	ND	+	ND	+
	Case8	+	ND	ND	+	+	-	ND	+	-	+
Tsoukalas, N & al. (2020) [19]		+	ND	ND	+	ND	ND	+	ND		+
Jin, D & al. (2020) [20]		+	ND	ND	+	+	-	-	+	+	-

CK7: Cytokeratin; EMA: Epithelial Membrane Antigen; CgA: Chromogranin A; NSE: Neuron-specific Enolase; WT1: Wilms Tumor protein 1; ND: not done.

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Several authors have investigated the role of hyperthermic intraperitoneal chemotherapy (HIPEC), most commonly using cisplatin (100 mg/m² for 90 minutes at 41 °C), as an adjunct following optimal cytoreductive surgery in DSRCT [27]. While some studies have suggested potential survival benefits and improved local control, results remain inconsistent, and the approach is associated with significant toxicity. Consequently, despite its promising rationale, the effectiveness of HIPEC in this setting remains controversial, and its use is best considered within clinical trials or highly selected patients [28].

Whole abdominopelvic radiotherapy (WAP-RT) has been explored as an adjuvant option after surgery in DSRCT. A reported experience by Goodman & al. showed that, although feasible, WAP-RT led to significant gastrointestinal and hematologic toxicity, and most patients ultimately relapsed [29]. Only a small minority achieved durable remission. More recent use of intensity-modulated radiotherapy (IMRT) appears less toxic, but relapse rates remain high. Overall, WAP-RT may offer temporary local control, but its benefit on survival is limited and it should be considered only in selected cases or clinical trials [8,30].

Given these limitations of locoregional approaches, systemic strategies have gained increasing interest. High-intensity polychemotherapy regimens, such as the P6 protocol, have long represented the backbone of treatment, while immunotherapy has also been explored as a potential strategy in DSRCT. However, the generally low mutational burden of sarcomas significantly limits the efficacy of immune checkpoint inhibitors, resulting in disappointing clinical outcomes.

Targeted therapies have also been investigated in advanced or progressive DSRCT, mainly focusing on antiangiogenic and tyrosine kinase inhibitors. Pazopanib has shown modest but clinically meaningful activity, with multi-institutional series reporting disease stabilization in a subset of heavily pretreated patients [31,32]. Additional data from the French national OUTC registry support the antiangiogenic effect of pazopanib and other targeted agents in this setting [33]. Sunitinib has likewise demonstrated limited yet notable activity in small case series, suggesting potential benefit in selected patients with refractory disease [34]. Imatinib has been evaluated in pediatric solid tumors, including DSRCT, but its efficacy appears minimal due to the lack of KIT or PDGFRA activating mutations in this tumor [35]. Overall, while these agents may offer temporary disease control, their impact remains modest, underscoring the need for more effective molecularly guided strategies.

In summary, outcomes after relapse remain poor, highlighting the urgent need for more effective therapeutic approaches.

Overall survival in DSRCT remains poor. Reported median survival ranges between 17 and 36 months, with most patients experiencing relapse even after aggressive multimodal therapy. Five-year survival rates rarely exceed 15–25%, and long-term survivors remain exceptional. These poor outcomes emphasize the urgent need for innovative therapeutic strategies, including targeted therapies and immunotherapy, to improve patient survival.

IV. CONCLUSION

This case underscores the diagnostic and therapeutic challenges associated with DSRCT. Despite aggressive multimodal strategies combining chemotherapy, surgery, and radiotherapy, the prognosis in advanced stages remains dismal, with survival rates still very low. The limitations observed in this case, consistent with reports in the literature, highlight the urgent need for more effective therapeutic strategies. In this context, further exploration of targeted therapies and immunotherapy holds promise and may pave the way toward improved outcomes in this rare and aggressive disease.

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