

Complete Cytoreductive Surgery After Molecular Reclassification of Metastatic Desmoplastic Small Round Cell Tumor: A Care-Compliant Case Report

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Case Report

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Abstract

Background

Desmoplastic small round cell tumor (DSRCT) is an ultra-rare, highly aggressive mesenchymal malignancy that predominantly affects adolescent and young adult males. Owing to its nonspecific clinical presentation and considerable histopathological overlap with other malignant small round cell tumors, accurate diagnosis remains challenging and may substantially influence therapeutic decision-making. Despite advances in multimodal treatment strategies, long-term survival remains poor because most patients present with advanced metastatic disease.

Case presentation

We report the case of an adolescent male presenting with progressive lower abdominal pain, constipation, anorexia, and weight loss. Magnetic resonance imaging demonstrated a large pelvic mass measuring $10.3 \times 14.2 \times 14.0$ cm extending into the lower abdomen, accompanied by multiple synchronous bilobar liver metastases measuring up to 6.4 cm and a small amount of ascites. Initial histopathological evaluation of a core needle biopsy suggested a malignant germ cell tumor, and treatment was initiated according to the MAKEI V-protocol. Expert pathological review with molecular confirmation of the characteristic EWSR1::WT1 fusion established the diagnosis of desmoplastic small round cell tumor, resulting in a change of systemic treatment to a Cooperative Weichteilsarkom Studiengruppe (CWS)-based regimen. Following favorable response to neoadjuvant chemotherapy, the patient underwent complete macroscopic resection of the pelvic primary tumor together with atypical resection of all radiologically detectable bilobar liver metastases. Histopathological examination confirmed extensive chemotherapy-induced tumor regression and microscopically complete (R0) resection. Despite initially successful multimodal treatment, metastatic recurrence occurred within months, and the patient ultimately died from progressive disease.

Conclusions

This case highlights the critical role of expert reference pathology and molecular diagnostics in distinguishing DSRCT from other malignant small round cell neoplasms. It further demonstrates that complete cytoreductive surgery, including simultaneous resection of synchronous liver metastases, may be feasible in carefully selected patients after favorable response to systemic therapy. Nevertheless, early recurrence despite R0 resection underscores the aggressive systemic biology of DSRCT and emphasizes the urgent need for more effective molecularly guided treatment strategies.

Background

Desmoplastic small round cell tumor (DSRCT) is a rare, highly aggressive soft tissue sarcoma that predominantly affects adolescents and young adults, with a marked male predominance. It accounts for less than 0.5% of all soft tissue sarcomas and is associated with an unfavorable prognosis despite intensive multimodal treatment. Median overall survival remains approximately 2–3 years, and long-term survival is uncommon. (1–4)

DSRCT is defined by the pathognomonic reciprocal chromosomal translocation $t(11;22)(p13;q12)$, resulting in the *EWSR1::WT1* fusion gene. The resulting chimeric transcription factor drives tumorigenesis through widespread transcriptional dysregulation and represents the principal molecular hallmark of the disease. Recent genomic studies further demonstrate that DSRCT is a genomically quiet malignancy with relatively few recurrent secondary mutations, emphasizing the dominant oncogenic role of the fusion protein. (5–9)

Clinically, DSRCT most commonly presents as a large intra-abdominal or pelvic mass with diffuse peritoneal dissemination. At diagnosis, many patients already have advanced disease with liver, lymph node, or extra-abdominal metastases. Radiological evaluation with contrast-enhanced CT and MRI is essential for defining disease extent, assessing resectability, and planning multimodal treatment. (4, 10, 11)

Accurate diagnosis remains challenging because DSRCT shares morphological and immunophenotypic features with several malignant small round cell neoplasms, including Ewing sarcoma, CIC-rearranged sarcoma, BCOR-associated sarcoma, rhabdomyosarcoma, lymphoma, neuroblastoma, and poorly differentiated carcinoma. Current WHO recommendations therefore emphasize an integrated diagnostic approach combining morphology, immunohistochemistry, and molecular confirmation of the *EWSR1::WT1* fusion. Contemporary reviews further highlight that molecular diagnostics have become indispensable for distinguishing DSRCT from other genetically defined round cell tumors and for guiding disease-specific management. (2, 12–15)

Because of its rarity, no universally accepted treatment standard exists. Current management is based on an intensive multimodal strategy combining multi-agent chemotherapy, aggressive cytoreductive surgery whenever feasible, and selected use of radiotherapy or hyperthermic intraperitoneal chemotherapy (HIPEC). Complete macroscopic cytoreduction has consistently been associated with improved survival, although recurrence remains common even after apparently curative treatment. Advances in molecular characterization have also identified several promising therapeutic targets—including CDK4/6, androgen receptor signaling, B7-H3, IGF-1R, and WT1-directed immunotherapeutic strategies—which may improve outcomes in the future. (7–9, 16–29)

Here, we present the case of an adolescent male with metastatic DSRCT whose initial diagnosis of a malignant germ cell tumor was revised following expert pathological review and molecular confirmation of an *EWSR1::WT1* fusion. This molecular reclassification fundamentally changed the multidisciplinary treatment strategy, enabling disease-specific chemotherapy and complete cytoreductive surgery despite

synchronous bilobar liver metastases. The case highlights the critical importance of expert molecular diagnostics for therapeutic decision-making in rare sarcomas.

Case Presentation

A 16 year old male with no relevant past medical history was referred to our tertiary pediatric surgical center because of progressive lower abdominal and right flank pain, constipation, anorexia, and unintended weight loss that had developed over several weeks. Clinical examination revealed a firm palpable mass occupying the lower abdomen without signs of generalized peritonitis or bowel obstruction. Routine laboratory investigations demonstrated elevated lactate dehydrogenase (LDH) and neuron-specific enolase (NSE), whereas serum alpha-fetoprotein (AFP) and beta-human chorionic gonadotropin (β -hCG) were within the normal range.

Magnetic resonance imaging (MRI) demonstrated a large heterogeneous solid pelvic mass measuring $10.3 \times 14.2 \times 14.0$ cm, extending cranially into the lower abdomen up to the level of the fifth lumbar vertebra (Fig. 1A, C). The tumor occupied the rectovesical space and displaced adjacent pelvic organs. Although definite infiltration of surrounding structures could not be confirmed radiologically, invasion of the rectum could not be excluded. Furthermore, multiple synchronous bilobar liver metastases were identified, the largest measuring 6.4 cm in diameter (Fig. 1B). A small amount of ascites was present. Computed tomography of the chest demonstrated no pulmonary metastases.

Given the patient's age, the tumor location, and the radiological findings, the initial differential diagnosis included malignant germ cell tumor, rhabdomyosarcoma, Ewing sarcoma, lymphoma, and other malignant small round cell neoplasms.

Initial histopathological assessment

Ultrasound-guided core needle biopsy of the pelvic tumor was performed to establish the diagnosis. Initial histopathological evaluation suggested a malignant germ cell tumor. Consequently, systemic chemotherapy according to the German MAKEI V-protocol (Cisplatin, Etoposid, Ifosfamid) was initiated.

However, the histopathological findings appeared unusual in relation to the clinical presentation and imaging findings. The biopsy specimen was therefore referred to a national sarcoma reference pathology center for expert review.

Comprehensive immunohistochemical and molecular analyses demonstrated the characteristic EWSR1::WT1 fusion, establishing the definitive diagnosis of desmoplastic small round cell tumor (DSRCT).

Following interdisciplinary discussion within the institutional pediatric sarcoma tumor board, the diagnosis prompted immediate modification of systemic treatment. Chemotherapy according to the MAKEI V-protocol was discontinued, and treatment was changed to a CWS-based multimodal sarcoma

regimen: block three I3VA (Ifosfamid, Vincristin und Dactinomycin), block four CEV (Carboplatin, Epirubicin and Vincristin), Blocks five, six, eight and nine were I3VA again, while blocks seven and ten were CEV.

Response to neoadjuvant therapy

Restaging MRI performed after completion of neoadjuvant chemotherapy demonstrated a marked reduction in the size of the primary pelvic tumor together with partial regression of the hepatic metastases. Importantly, no additional metastatic lesions had developed during systemic treatment.

Following reassessment by a multidisciplinary team consisting of pediatric surgeons, hepatobiliary surgeons, pediatric oncologists, radiologists, pathologists, anesthesiologists, and radiation oncologists, complete macroscopic cytoreduction was considered technically feasible despite the presence of synchronous liver metastases.

Surgical treatment

Exploratory laparotomy confirmed a large pelvic tumor originating within the rectovesical space. Despite the close anatomical relationship to the rectum, diffuse peritoneal carcinomatosis was not present.

Complete en bloc resection of the pelvic tumor with partial resection of the omentum majus and appendectomy was successfully achieved while preserving both the urinary bladder and rectum. Following resection of the primary tumor, all radiologically detectable liver metastases were removed by multiple atypical wedge resections in the segments II/III, Iva, V, VI, VII and VIII, resulting in complete macroscopic cytoreduction. (Fig. 2A-D)

No major intraoperative complications occurred.

Histopathological examination of the resection specimen

Macroscopic examination confirmed complete removal of both the pelvic tumor and all resected liver metastases.

Microscopic evaluation demonstrated extensive chemotherapy-induced tumor regression consistent with a favorable pathological response.

All surgical margins were microscopically tumor-free (R0 resection).

The diagnosis of desmoplastic small round cell tumor was confirmed by histopathological examination.

Follow-up

The postoperative course was uneventful, and no clinically relevant surgical complications occurred. The patient recovered well and subsequently continued multimodal oncological treatment according to institutional recommendations. The patient received maintenance chemotherapy with Trofosfamid and Idarubicin, as well as Trophosphamid and Etoposid in alternating cycles.

Despite the initially favorable postoperative findings, follow-up imaging performed several months after surgery demonstrated recurrent metastatic disease. Salvage molecular therapy was evaluated but was not possible. A relapse chemotherapy was started according to the CSW-Guidance with TECC (Topotecan, Etoposid, Carboplatin, Cyclophosphamid). The first and second relapse chemotherapy block were with Topotecan, and Carboplatin, the third with Topotecan and Cyclophosphamid. The patient suffered from therapy-associated complications, such as tumor cachexia, fungal pneumonia and meningitis with postmenigitic hydrocephalus. The implantation of a VP-shunt, the change to a VA-shunt due to pleural effusions, the externalization of it and the relocation of the VA-shunt were also part of the complications.

However, progressive systemic disease developed despite treatment.

Following repeated multidisciplinary discussion and consultation with the patient and his family, further curative treatment was considered no longer feasible, and management was transitioned to best supportive and palliative care.

The patient ultimately died from progressive metastatic disease.

Discussion

Diagnostic implications of molecular pathology

DSRCT remains one of the most diagnostically challenging malignant small round cell tumors because of its overlapping morphological and immunophenotypic features with Ewing sarcoma, CIC-rearranged sarcoma, BCOR-associated sarcoma, rhabdomyosarcoma, lymphoma, neuroblastoma, and poorly differentiated carcinoma. Contemporary WHO recommendations therefore emphasize an integrated diagnostic approach combining histomorphology, immunohistochemistry, and molecular confirmation of the EWSR1::WT1 fusion. Recent reviews further highlight that molecular diagnostics have become indispensable for distinguishing genetically defined round cell sarcomas and for guiding disease-specific management. (2, 12–15)

Our patient illustrates the profound clinical impact of expert pathological review. The initial diagnosis of a malignant germ cell tumor resulted in treatment according to a germ cell tumor protocol. Only after reference pathology and molecular confirmation of the EWSR1::WT1 fusion was the diagnosis revised to DSRCT, leading to a complete change in therapeutic strategy. This case underscores that molecular confirmation is not merely diagnostic but directly determines multidisciplinary treatment decisions in rare sarcomas. (2, 5–6, 12–15)

Multimodal treatment and the role of surgery

Because prospective randomized trials are lacking, current management of DSRCT relies on multimodal treatment consisting of intensive multi-agent chemotherapy, aggressive cytoreductive surgery whenever feasible, and selected use of radiotherapy and HIPEC. (31) Retrospective series consistently demonstrate that complete macroscopic cytoreduction is associated with prolonged survival, although patient selection and retrospective study design must be considered when interpreting these findings. (3, 4, 11, 16–19, 32)

In the present case, induction chemotherapy produced substantial tumor regression, allowing complete resection of the primary pelvic tumor together with atypical resections of synchronous bilobar liver metastases. Histopathological examination demonstrated a marked treatment response and complete (R0) resection. Although evidence regarding hepatic metastasectomy in DSRCT remains limited, available surgical series support resection of all technically resectable disease as part of maximal cytoreduction whenever feasible. (3, 18, 19, 32)

Tumor biology outweighs local disease control

Despite complete cytoreductive surgery and favorable histopathological response, our patient developed early metastatic recurrence and ultimately died of progressive disease. This clinical course illustrates the fundamental biological aggressiveness of DSRCT. While surgery can achieve excellent local disease control, recurrence remains frequent, suggesting that systemic tumor biology rather than local control ultimately determines long-term outcome in many patients. (3, 4, 7–9, 11, 20)

The central role of the EWSR1::WT1 fusion oncogene is further supported by experimental studies demonstrating persistent dependence of DSRCT cells on fusion-driven transcriptional programs despite conventional chemotherapy. These observations provide a biological explanation for the high recurrence rates observed even after apparently complete surgical resection. (7–9, 20–22)

Emerging molecular and targeted therapeutic strategies

The identification of the EWSR1::WT1 fusion has not only improved diagnostic precision but has also established a biological framework for the development of targeted therapies. Functional studies demonstrate that DSRCT remains highly dependent on fusion-driven transcriptional programs, supporting the concept of directly or indirectly targeting this oncogenic dependency. (7–9, 20)

Among the most promising approaches, inhibition of EWSR1::WT1-dependent transcription by lurbinectedin and CDK4/6 inhibition have shown encouraging preclinical activity, providing proof-of-concept that the molecular drivers of DSRCT can be therapeutically exploited. (8, 9, 20)

In parallel, immunotherapeutic strategies are rapidly evolving. Because WT1-derived peptides are constitutively expressed as part of the fusion program, WT1-directed vaccines and T-cell receptor (TCR)-based therapies represent biologically attractive investigational approaches. In addition, B7-H3 has

emerged as a highly expressed surface antigen in DSRCT, supporting the development of antibody-based and radioimmunotherapeutic strategies. Although these approaches remain investigational, they illustrate the transition from empiric chemotherapy toward biologically informed treatment strategies. (23, 25, 26, 28, 30)

Taken together, these advances suggest that future treatment of DSRCT will increasingly combine multimodal local therapy with molecularly targeted and immune-based approaches directed against the unique biology of the EWSR1::WT1 fusion. (7–9, 20) (23, 25, 26)

Clinical implications

This case highlights several clinically relevant aspects of DSRCT management. First, expert pathological review with molecular confirmation should be considered mandatory whenever DSRCT is suspected because diagnostic reclassification may fundamentally alter treatment strategy. Second, multidisciplinary evaluation at experienced sarcoma centers is essential to identify patients who may benefit from aggressive multimodal treatment including complete cytoreductive surgery. Finally, despite optimal local treatment, the poor long-term prognosis underscores the urgent need for molecularly targeted and immunotherapeutic approaches capable of improving systemic disease control. (2–4, 11–19) (7–9, 20–29, 30, 32)

Conclusion

Desmoplastic small round cell tumor remains one of the most aggressive malignancies affecting adolescents and young adults, with prognosis largely determined by early metastatic dissemination and limited efficacy of currently available systemic therapies.

This case demonstrates that expert molecular pathology is not merely confirmatory but can fundamentally alter clinical management. In our patient, molecular reclassification from an initially presumed malignant germ cell tumor to DSRCT resulted in immediate modification of systemic therapy and enabled individualized multidisciplinary treatment planning. Following favorable response to neoadjuvant chemotherapy, complete macroscopic cytoreductive surgery including simultaneous resection of synchronous bilobar liver metastases was successfully achieved with microscopically negative margins.

Despite optimal local treatment and a favorable pathological response, early metastatic recurrence occurred, highlighting that aggressive tumor biology rather than insufficient local control remains the principal obstacle to long-term survival in DSRCT.

This case supports early referral of diagnostically challenging malignant small round cell tumors to specialized sarcoma centers with expertise in molecular pathology and multidisciplinary management. Furthermore, it underscores the urgent need for prospective collaborative studies and biologically targeted therapeutic approaches capable of improving systemic disease control in this rare and highly aggressive malignancy.

Abbreviations

BMI: Body mass index

CEV: Carboplatin, Epirubicin, and Vincristine

CWS: Cooperative Weichteilsarkom Studiengruppe

DSRCT: Desmoplastic small round cell tumor

EWSR1: Ewing sarcoma breakpoint region 1

HIPEC: Hyperthermic intraperitoneal chemotherapy

I3VA: Ifosfamide, Vincristine, and Dactinomycin

MAKEI: Maligne Keimzelltumoren Study Group

MRI: Magnetic resonance imaging

PEI: Cisplatin, Etoposide, and Ifosfamide

R0: Microscopically margin-negative resection

TECC: Topotecan, Etoposide, Carboplatin, and Cyclophosphamide

WT1: Wilms tumor 1

Declarations

Ethics approval and consent to participate

Ethics approval was not required for publication of this single case report according to institutional regulations.

Consent for publication

Written informed consent for publication of this case report and the accompanying images was obtained from the patient and his legal guardians. A copy of the signed consent form is available for review by the Editor-in-Chief of this journal.

Availability of data and materials

All data generated or analyzed during this study are included in this published article. Additional anonymized clinical information is available from the corresponding author upon reasonable request.

Competing interests

The authors declare that they have no competing interests.

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Use of AI

The authors used ChatGPT (OpenAI, San Francisco, CA, USA; GPT-5.5) exclusively to improve the language and readability of the manuscript. The authors reviewed and edited all AI-generated text and take full responsibility for the accuracy and integrity of the final manuscript.

Authors' contributions

SR: Data collection. conceptualization, Writing first draft.

MR: Writing – review.

xZ: Data collection

AN: Supervision, Writing – review.

MCS: Supervision, Writing – draft.

FM: Supervision. Conceptualization, Writing – review.

ST: Writing – review & editing.

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Tables

Tables 1 and 2 are available in the supplementary files section

Figures



Figure 1

Initial magnetic resonance imaging: (A) Contrast-enhanced MRI demonstrating a large heterogeneous pelvic mass measuring **10.3 × 14.2 × 14.0 cm**, extending cranially into the lower abdomen to the level of the fifth lumbar vertebra. (B) Synchronous bilobar liver metastases, with the largest lesion measuring **6.4 cm**. (C) Contrast-enhanced MRI demonstrating the pelvic mass in sagittal sclice.

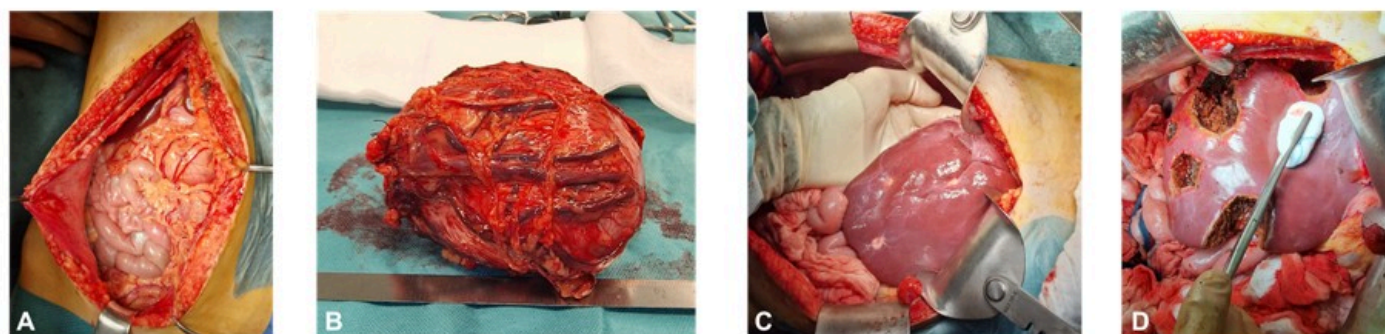


Figure 2

Macroscopic appearance of the resected pelvic tumor and representative liver metastases following complete cytoreductive surgery. A: Laparotomy overview. B: in toto resected DRSCT. C: Liver metastases.

D: Atypical resection of bilobular liver metastases.

Supplementary Files

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