



Case Report

# Desmoplastic small round cell tumor of seminal vesicle: A case report with review of literature

Vanshika Rastogi<sup>1</sup>, MD, Sushila Jaiswal<sup>2</sup>, MD, Neeraj Rastogi<sup>1</sup>, MD, Uday Pratap Singh<sup>3</sup>, MCh

Departments of <sup>1</sup>Radiotherapy, <sup>2</sup>Pathology, <sup>3</sup>Urology, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Lucknow, Uttar Pradesh, India

## ABSTRACT

Desmoplastic small round cell tumor (DSRCT) is an aggressive, uncommon disease and difficult to diagnose and identified by translocation t(11;22) (p13;q12) leading to fusion transcript *EWSR1-WT1*. A case of DSRCT of the seminal vesicle diagnosed by histopathology, immunohistochemistry, and genetic study, and treated with chemotherapy, surgery, and radiation therapy on the lines of Ewing Sarcoma protocol is presented. Four cycles of chemotherapy was given, and he had a partial response, so underwent debulking surgery followed by radiation therapy.

**Keywords:** Chemotherapy, DSRCT, Seminal vesicle

## INTRODUCTION

Pioneer work on Desmoplastic small round cell tumor (DSRCT) was done by Gerald in 1991 and is defined by breakpoints and translocation between *EWSR1* (Chromosome 22) and *WT1* (chromosome 11), and identified by *EWSR1-WT1* fusion transcript.<sup>[1,2]</sup> This particular translocation differentiates these tumors from other small round blue cell tumors, such as Ewing.<sup>[3]</sup> Approximately 1000 cases have been documented in the literature. Due to a lack of trials, the patients are treated on the lines of Ewing Sarcoma (ES), but differ in clinical presentation and prognosis due to fundamental differences in biology. DSRCT is composed of small round cells arranged in clusters, displaying a wide range of cellular features of epithelial, mesenchymal, and neural cells surrounded by desmoplastic stroma. It commonly affects the abdominopelvic organs of adolescent and young adult males (Males:Females = 5:1). The primary tumors of the seminal vesicle are exceptionally uncommon, and adenocarcinoma being the most common. Here we present a case of DSRCT originating from the seminal vesicle.<sup>[3]</sup>

## CASE REPORT

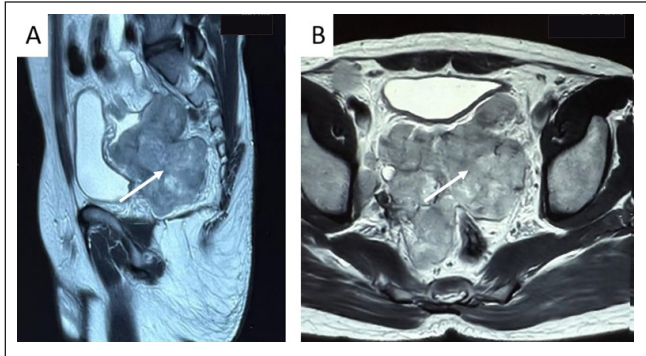
A male patient of 30 years presented with progressive abdominal pain, vomiting, weight loss, and loss of appetite for the past 6 months. He had tenderness in the right lumbar region without any palpable mass. The carbohydrate antigen (CA)19.9 marker was high at 79.8 U/ml. He was treated with two weeks of antibiotics and anthelmintics on lines of a gastrointestinal infection, but no response. Ultrasound abdomen revealed a pelvic nodal mass involving the bilateral seminal vesicles with bilateral hydronephrosis and a 5 × 5 cm lesion in segment VI of the liver. MRI abdomen revealed a conglomerate mass of 8.5 × 8 × 9 cm in the pelvic region in the rectovesical space, infiltrating bilateral seminal vesicles with multiple bilateral common, internal, and external iliac nodes with free fluid in the pelvis [Figure 1]. Fluorodeoxyglucose-Positron emission tomography-computed tomography (FDG PET-CT) showed an avid 6.5 × 4.5 cm pelvic nodal mass [standardized uptake value (SUV)=11.4] with bilateral common iliac, external, and internal iliac with left supraclavicular lymph nodes

\*Corresponding author: Vanshika Rastogi, Department of Radiotherapy, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Raebareli Road, Lucknow, Uttar Pradesh, 226014, India. [vanshikarastogi98@gmail.com](mailto:vanshikarastogi98@gmail.com)

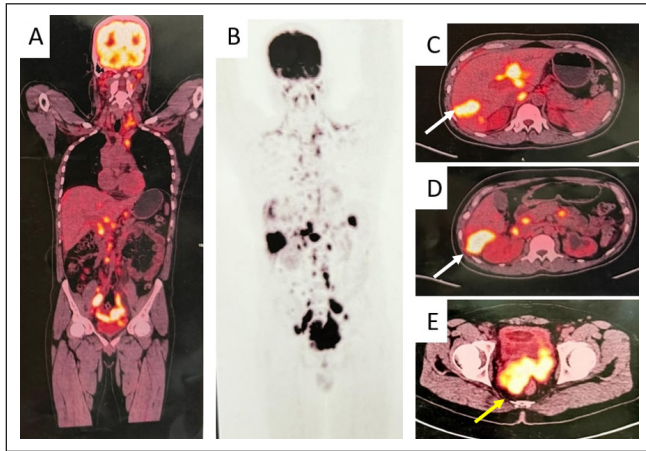
Received: 01 August 2025 Accepted: 27 December 2025 Published: 23 January 2026 DOI: 10.25259/ASJO\_58\_2025

This is an open-access article distributed under the terms of the Creative Commons Attribution-Non Commercial-Share Alike 4.0 License, which allows others to remix, transform, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms. ©2026 Published by Scientific Scholar on behalf of Asian Journal of oncology

(SUV=7.8) and a 5.8 × 5.6 cm lesion in Segment VI of liver (SUV=11.4) along with sub-diaphragmatic (SUV=6.4) peritoneal thickening [Figure 2]. PET-CT-guided biopsy of the pelvic mass and liver lesion was done.



**Figure 1:** MRI of Abdomen in (A) Sagittal and (B) Axial section shows conglomerate mass (shown by white arrows) in the pelvis in recto-vesical space infiltrating the bilateral seminal vesicles.

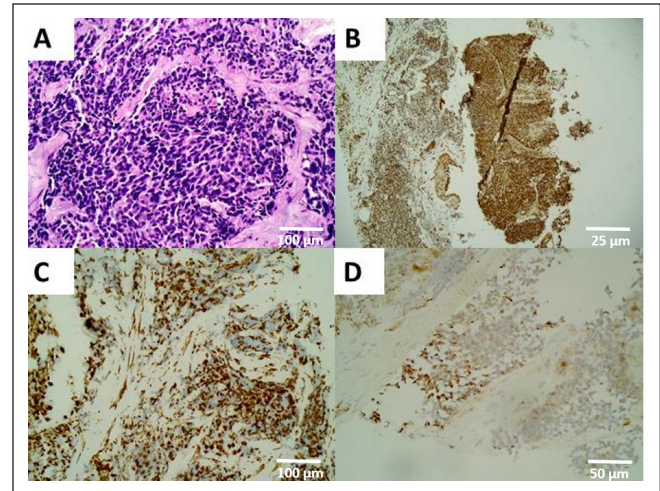


**Figure 2:** PET-CT images of whole body show widespread abnormal uptake throughout the (A) abdomen and pelvis (B) maximal intensity projection image, (C and D) FDG-avid lesion in Segment VI of the liver (shown by white arrows) with multiple retroperitoneal lymph nodes, (E) a lymph nodal mass in recto-vesical space (shown by yellow arrow). FDG: Fluorodeoxyglucose, PET-CT: Positron emission tomography-computed tomography

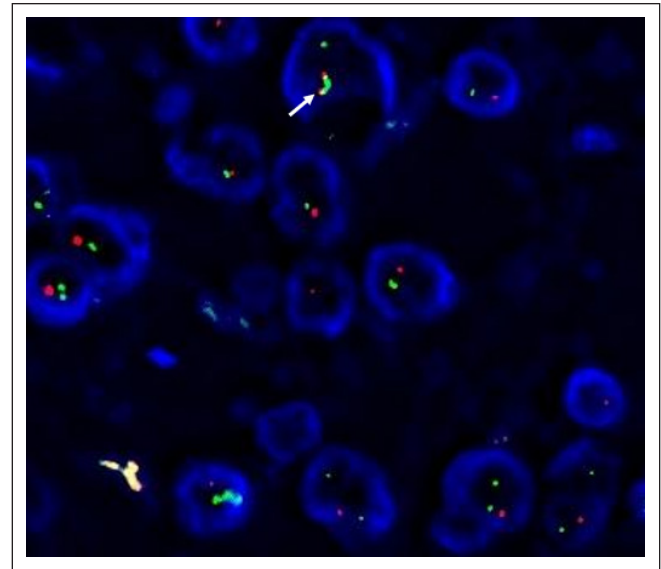
### Pathology

The tumor tissue had a soft texture and a greyish-yellow color. Slides were stained with hematoxylin and eosin, and they revealed that the tumor cells were in sheets and clusters enclosed by thick, dense fibrous tissue. The tumor cells were small, round, and had small foci of rosette-like areas, hyperchromatic nuclei, inconspicuous nucleoli with mild pleomorphism, and varying amounts of cytoplasm [Figure 3A]. Immunohistochemistry [Figure 3B, C, and D] showed focal dot-like cytoplasmic positivity for cytokeratin, vimentin, desmin, and CD99 in tumor cells and negative

for synaptophysin, chromogranin, CD45, NKX2.2, and BCL2. No SS18 (18q11.2) gene rearrangement was seen on FISH testing, and on genetic testing, *EWSR1-WT1* fusion transcript [Figure 4] was detected, diagnostic of DSRCT.



**Figure 3:** H and E-stained section show (A) a small round cell tumor separated by desmoplasia (100X). Tumor cells are positive for (B) Vimentin (400X), (C) Cytokeratin (100X) and (D) Desmin (200X) on immunohistochemistry. H and E: Hematoxylin and eosin



**Figure 4:** FISH showing *EWSR1-WT1* fusion gene. *WT1* and *EWSR1* gene loci are represented by red and green signals respectively, and fusion is represented by red-green fused signals (yellow) (shown by white arrow). FISH: Fluorescence in-situ hybridization

The patient was given 4 cycles of Vincristine, Adriamycin, and Cyclophosphamide (VAC) alternating with Ifosfamide and Etoposide (IE) chemotherapy as per EFT-2001 protocol. He had a partial response, so, underwent debulking surgery and post-op radiation therapy. The patient was then continued further 6 cycles of chemotherapy and has shown

a good response at 6 months post radiation therapy, and has now been planned for an interim PET scan.

## DISCUSSION

The incidence of DSRCT is 0.2-0.7/million<sup>[1]</sup> with an age-standardized incidence rate of 0.3/million in adults of 20-24 years.<sup>[3]</sup> Earlier, it was called as "Sarcomatosis" (a large abdominal tumor) and now identified as DSRCT.<sup>[1]</sup> The surveillance, epidemiology, and end results (SEER) database reported 192 cases of DSRCT between 1973-2007. It arises from mesothelial cells and can spread to the abdominal peritoneum and often involves pleura/lung and is cytogenetically diagnosed by the presence of a specific reciprocal translocation t(11;22)(p13;q12), seen in >90% of patients regardless of origin of primary.<sup>[4]</sup> The EWS gene is located on chromosome 22q12, and the WT1 gene on 11p13. *WT1* is a tumor suppressor gene encoding a transcription factor which enhances the expression of platelet-derived growth factor and inhibits p53. This fusion leads to loss of suppressor function of *WT1* and control of growth factors like PDGF $\alpha$ , FGFR4, VEGFR2, and IGF2 from the EWS gene. This fusion transcript is detected by RT-PCR or fluorescence in-situ hybridization (FISH). The ES family tumors also carry EWS gene translocation, and DSRCT is distinguished from Ewing by amplified Androgen Receptor (AR) expression.<sup>[5]</sup> Microscopically larger cells with clear cytoplasm display anisonucleosis, dispersed chromatin palisading at the periphery resembling rosettes and pseudo-papillae. The epithelial markers cytokeratin and epithelial membrane antigen (EMA) are positive. Cytoplasmic immunoreactivity has a dot-like pattern seen in cytokeratin-positive cells, such as CK20 in Merkel cell carcinoma, CK5/6 in malignant mesothelioma, and CD20 in Lymphoma. Blastemal predominant Wilms tumor, one of the differential diagnoses, is characterized by poorly differentiated cells with desmoplastic stroma and immunoreactivity for desmin and cytokeratin exhibiting focal triphasic elements as glomeruloid bodies and *WT1* immunoreactivity at both the amino and carboxy terminuses but no *EWSR1-WT1* rearrangement. Early start of chemotherapy is recommended as per the Chicago Consensus and MD Anderson Cancer Centre guidelines. There is a partial response to chemotherapy and frequent distant relapses after radical surgery, and most of patients die due to the disease within 3 years of diagnosis.<sup>[6,7]</sup> Average disease-free interval is 10-15 months, and median survival is 28-60 months.<sup>[8]</sup>

## CONCLUSION

Early diagnosis of DSRCT is difficult due to a lack of specific signs and symptoms; hence, early diagnosis by histology, immunohistochemistry, and gene studies and treatment is crucial for improved outcome. The survival is poor despite of multimodality treatment as patients relapse at distant sites and are unresponsive to second-line therapy. The standard treatment includes surgery, chemotherapy, and adjuvant radiation therapy. *EWSR1-WT1*

fusion gene may be a promising target for future therapy and research, and targeting downstream genes and signaling pathways activation may help in developing immunotherapy.

**Author contributions:** VR and NR: Conceptualization and design, writing and review; SJ: Data Acquisition and curation; UPS: Supervision and resources.

**Ethical approval:** Institutional Review Board approval is not required.

**Declaration of patient consent:** The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**Financial support and sponsorship:** Nil

**Conflicts of interest:** There are no conflicts of interest.

**Use of artificial intelligence (AI)-assisted technology for manuscript preparation:** The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

## REFERENCES

- Gerald WL, Ladanyi M, de Alava E, Cuatrecasas M, Kushner B H, LaQuaglia M P, *et al.* 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;16:3028-36.
- Tsoukalas N, Kiakou M, Nakos G, Tolia M, Galanopoulos M, Tsapakidis K, *et al.* Desmoplastic small round-cell tumour of the peritoneal cavity: Case report and literature review. *Ann R Coll Surg Engl* 2020;102:e77-e81.
- Lettieri CK, Garcia-Filion P, Hingorani P. Incidence and outcomes of desmoplastic small round cell tumor: Results from the surveillance, epidemiology, and end results database. *J Cancer Epidemiol* 2014;2014:680126.
- Liang L, Tatevian N, Bhattacharjee M, Tsao K, Hicks J. Desmoplastic small round cell tumor with atypical immunohistochemical profile and rhabdoid-like differentiation. *World J Clin Cases* 2014;2:367-72.
- Bulbul A, Shen JP, Xiu J, Tamayo P, Husain H. Genomic and proteomic alterations in desmoplastic small round blue-cell tumors. *JCO Precis Oncol* 2018;2:PO1700170.
- Scheer M, Vokuhl C, Blank B, Hallmen E, von Kalle T, Münter M. Desmoplastic small round cell tumors: Multimodality treatment and new risk factors. *Cancer Med* 2019;8:527-42.
- Honoré C, Delhorme JB, Nassif E, Faron M., Ferron G., Bompas, *et al.* Can we cure patients with abdominal desmoplastic small round cell tumor? Results of a retrospective multicentric study on 100 patients. *Surg Oncol* 2019;29:107-12.
- Chen W, Chen H, Zhao C, Xie S, Qin J, Liu W. Desmoplastic small round cell tumor: Report of two cases and literature analysis of radiological findings. *Quant Imaging Med Surg* 2023;13:4762-69.

**How to cite this article:** Rastogi V, Jaiswal S, Rastogi N, Singh UP. Desmoplastic small round cell tumor of seminal vesicle: A case report with review of literature. *Asian J Oncol.* 2026;12:1. doi: 10.25259/ASJO\_58\_2025