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A 23-Year-Old Joradanian Woman with a Desmoplastic Small Round Cell Tumor Involving the Ovary

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Study Design A
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Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Conflict of interest: None declared

Patient: Female, 23
Final Diagnosis: Desmoplastic small round cell tumor
Symptoms: Nausea • vomiting • severe abdominal pain of two weeks • weight loss
Medication: —
Clinical Procedure: —
Specialty: Obstetrics and Gynecology

Objective: Rare disease

Background: Desmoplastic small round cell tumor (DSRCT) is a rare soft tissue sarcoma that usually arises in the abdomen or pelvis in young boys and adolescents. Presenting symptoms include abdominal pain and ascites. However, DSRCT is often disseminated throughout the peritoneal cavity at diagnosis, and the prognosis is poor. This report is of a case of DSRCT in a 23-year-old Jordanian woman who presented with abdominal pain.

Case Report: An unmarried 23-year-old woman presented with abdominal pain. On examination, she was found to have ascites. A computed tomography (CT) scan of the abdomen and pelvis showed a complex cystic mass in the left ovary, multiple peritoneal deposits, a large amount of ascitic fluid, two hypodense lesions in the liver, and multiple enlarged lymph nodes. Diagnostic laparoscopy was performed, and multiple tumor biopsies were obtained. Histopathology showed a cellular tumor composed nests of small round cells embedded in desmoplastic stroma. Immunohistochemistry showed positive staining of the tumor cells for pan-cytokeratin, desmin, Wilms tumor 1 (WT1) antigen, epithelial membrane antigen (EMA), and CD56, which supported the diagnosis of DSRCT. After the second cycle of the P6 Protocol, which included seven courses of chemotherapy, the patient developed a severe and fatal infection.

Conclusions: It is important to consider the diagnosis of DSRCT that may present atypically, particularly in patients who present with abdominal and pelvic masses. DSRCT has a rapid and aggressive course that requires early and definitive diagnosis with prompt treatment that includes systemic chemotherapy.

MeSH Keywords: Antineoplastic Agents • Desmoplastic Small Round Cell Tumor • Mediterranean Region • Ovarian Cysts

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/919488

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Background

Desmoplastic small round cell tumor (DSRCT) was originally described and reported by Gerald and Rosai in 1989 [1]. DSRCT is a rare soft tissue sarcoma that usually arises in the abdomen or pelvis in young boys and adolescents, and common presenting symptoms include abdominal pain and distension [1]. The diagnosis is based on specific clinicopathological, immunohistochemical, and molecular features [1]. However, DSRCT often presents late with disseminated peritoneal tumors, and the prognosis is poor [1]. There is geographical variation in the incidence of DSRCT, which is more common in African-Americans [2].

This report is of a rare case of DSRCT that involved the left ovary in 23-year-old Jordanian woman, who presented with severe abdominal pain and who was initially diagnosed with a cystic ovary.

Case Report

A 23-year-old single Jordanian woman presented to our medical center with a two-week history of severe abdominal pain associated with nausea, anorexia, and vomiting. She had lost 8 kg in weight during the previous two months. Before the onset of these recent symptoms, she had been diagnosed in the private sector with a left ovarian cyst, which was managed conservatively. She had no family history of malignancy.

On physical examination, her abdomen was distended, and massive ascites was detected. Laboratory investigations were normal, except for an increased serum level of cancer antigen-125 (CA-125) of 91.03 U/mL (normal, <46U/mL). Contrast-enhanced computed tomography (CT) imaging of the abdomen and pelvis showed complex cystic lesions of the left ovary that was enlarged and measured 6.5×5.0 cm (Figure 1). Also, there were multiple peritoneal deposits in the abdomen and pelvis associated with thickened peritoneal reflections (Figure 2). A large amount of ascitic fluid was present in the abdomen and pelvis, and two hypodense lesions were present in the liver, the largest of which measured 1.2×0.8 cm (Figure 3). Multiple enlarged lymph nodes were seen in the mesentery, and the cardiophrenic, periaortic, para-aortic, and bilateral external iliac regions. The largest lymph node measured about 1 cm and was present in the right external iliac region.

A diagnostic laparoscopy was performed that identified multiple tumor deposits in the peritoneal wall, omentum, uterus, and bowel near the terminal ilium. There was a left ovarian mass that bled to the touch. Massive ascites was present that reached the diaphragm. During laparoscopy, ascitic fluid was aspirated, and tumor biopsies were taken.

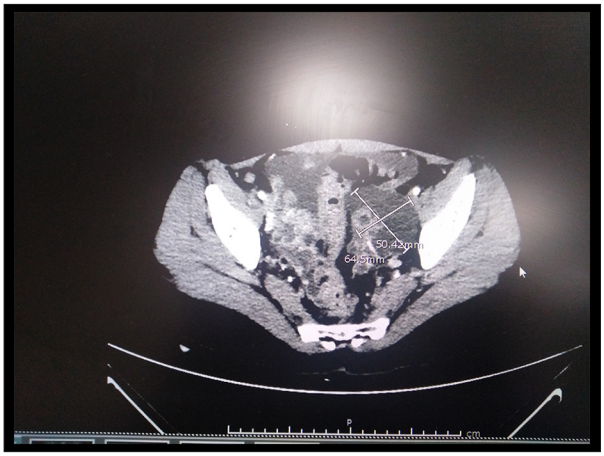

Figure 1. Abdominal computed tomography (CT) shows a complex left ovarian mass in a 23-year-old Jordanian woman who presented with abdominal pain. The axial CT scan shows the abdomen and pelvis. The ovarian mass was initially considered to be the origin of the tumor.

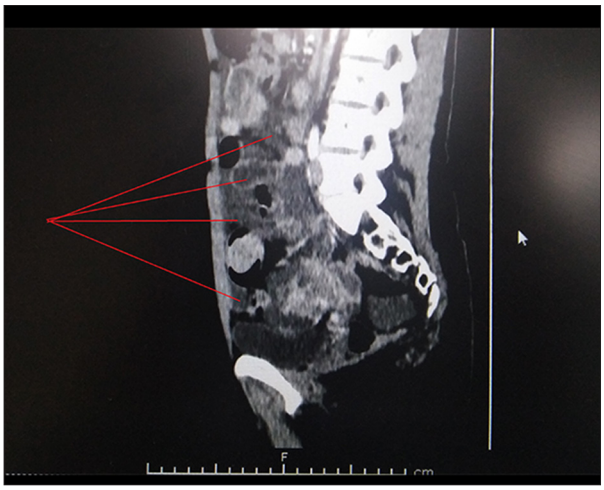

Figure 2. Abdominal and pelvic computed tomography (CT) shows multiple peritoneal deposits in a 23-year-old Jordanian woman who presented with abdominal pain. The sagittal abdominal and pelvic CT scan shows multiple peritoneal deposits indicated by red arrows.

Histopathology showed peritoneal and ovarian tissue diffusely infiltrated by irregular nests of small round cells embedded in desmoplastic stroma. The tumor cells showed hyperchromatic nuclei with some nuclear molding and scanty cytoplasm. Foci of necrosis were present, and the tumor cells had a high mitotic count with nuclear atypia. Immunohistochemistry showed that the tumor cells were positive for the expression of pancytokeratin, desmin, Wilms tumor 1 (WT1), epithelial membrane antigen (EMA), and CD56. The tumor cells showed focal immunopositivity for synaptophysin, chromogranin, inhibin, and calretinin. The ascitic fluid contained malignant cells

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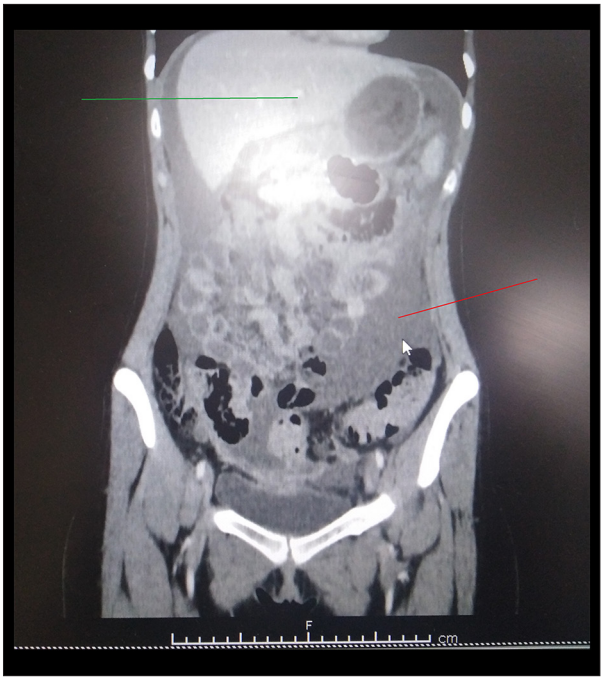

Figure 3. Abdominal and pelvic computed tomography (CT) shows ascites and a liver mass in a 23-year-old Jordanian woman who presented with abdominal pain. The sagittal abdominal and pelvic CT scan shows a large amount of ascites fluid (indicated by the red arrow) with a liver lesions (indicated by the green arrow).

with small round-cell morphology. The diagnosis was made of stage IV desmoplastic small round cell tumor (DSRCT) involving the left ovary.

Chemotherapy treatment began with the P6 Protocol, and after the second cycle of cyclophosphamide, doxorubicin, and vincristine, she suffered severe lower abdominal pain associated with nausea and vomiting, neutropenic fever, and progressive dysphagia. On examination, abdominal distension was present. The abdomen was tense and tender with positive transmitted thrill on percussion. Her temperature was 39°C. A contrast-enhanced CT scan showed a pleural effusion, ascites, and a distended loaded colon with the maximum diameter of the transverse colon of 7–8 cm. The CT scan also showed that there was progression of liver metastasis and lymphadenopathy. She was managed with an enema and laxatives and removal of an indwelling central catheter. Blood culture identified *Staphylococcus epidermidis* and *Staphylococcus warneri* infection. She was treated with granulocyte colony-stimulating factor (GCSF), vancomycin, meropenem, micafungin, and levofloxacin. However, septic shock progressed to multiorgan failure. The patient died after her second dose of chemotherapy four months since she was initially diagnosed with stage IV DSRCT.

Discussion

To the best of our knowledge, this is the first reported case of desmoplastic small round cell tumor (DSRCT) in Jordan, the fourth within the region of Jordan and the Mediterranean region. Koniaria et al. reported a DSRCT in a 29-year-old man from the Middle East living in Greece [3]. Also, Idris reported a case of gastric DSRCT in a 28-year-old Syrian man [4]. In 2013, Abu-Zaid et al. described a case of DSRCT in a 26-year-old woman from Saudi Arabia [5]. Recently, Nacef et al. reported a case in a 46-year-old Tunisian woman [6]. Lettieri et al. reported the findings from an epidemiological study that was based on the Surveillance, Epidemiology, and End Results (SEER) registry data from the National Cancer Institute (NCI) and identified that 134 out of 192 case of DSRCT occurred in Caucasians, and 46 were diagnosed in African-Americans [7]. Hassan et al. and Schwarz et al. reported a higher incidence of DSRCT in Caucasians and no cases in Americans of Mediterranean descent [8,9]. Recently, Stiles et al., in a nationwide study, reported similar results [10]. However, Worch et al. studied racial differences in the incidence of mesenchymal tumors associated with EWSR1 gene translocations identified as DSRCT [2]. DSRCT was three-times more likely to occur in African-Americans compared with Caucasians [2]. Until the current case report, there have been no previous reports of cases of DSRCT from the Mediterranean region and Jordan.

In the US, DSRCT has gained increasing recognition since the clinicopathological description of 19 cases by Gerald et al. in 1991, and the identification of diagnostic chromosomal and molecular abnormalities [11]. Additional large clinicopathologic series confirmed that DSRCT predominantly affects young males, usually arises infra-diaphragmatically, and expresses a heterogeneous phenotype [11]. DSRCT is classified as an undifferentiated soft tissue tumor with small cell histological features in which the tumor cells are closely arranged in nests with epithelioid cells of different sizes and shapes [3]. DSRCT also includes a desmoplastic reaction with the formation of fibrous tissue around and among tumor cell nests [3]. In most cases of DSRCT, desmin and cytokeratin expression are found, which are the most specific immunohistochemical markers for DSCRT, with the stromal elements being vimentin-positive [3]. The clinical manifestations of DSCRT depend on the site of involvement. Intra-abdominal and intra-pelvic DSCRT are associated with abdominal distension, abdominal discomfort, and an abdominal mass that may be associated with constipation, dysuria, and intestinal obstruction [3]. Most patients develop significant weight loss [3]. The patient described in this case report complained of abdominal and pelvic pain, and an initial diagnosis was made in the private sector of ovarian cyst, which was managed conservatively.

Because of the rarity of the tumor, the optimal treatment for patients with DSRCT has not yet been determined as the small

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number of patients has not supported large multi-center clinical trials. The management of patients with DSRCT remains challenging, and the prognosis is poor despite the use of aggressive chemotherapy regimens, such as Protocol P6, or whole abdominal radiation therapy and debulking surgery [12]. Currently, other approaches to the treatment of DSRCT are being evaluated, including the use of targeted therapies [12]. Also, the use of complete surgical resection combined with hyperthermic intraperitoneal chemotherapy and postoperative intensity-modulated radiation therapy (IMRT) are being investigated [12].

Conclusions

A rare case of desmoplastic small round cell tumor (DSRCT) in a 23-year-old Jordanian woman is reported. This case has highlighted that this tumor may present atypically, particularly in patients who present with abdominal and pelvic masses. DSRCT has a rapid and aggressive course that requires early and definitive diagnosis with prompt treatment that includes systemic chemotherapy. In this case, the left ovarian mass was initially diagnosed as an ovarian cyst, which delayed definitive diagnosis and treatment, resulting in symptomatic presentation with advanced-stage malignancy. It is hoped that the presentation of this case will highlight the occurrence of this rare malignancy, especially in young adults, women, and all racial groups.

Conflict of interest

None.

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