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Case Report

# Desmoplastic Small Round Cell Tumour of the Uterus: A Case Report

Nur Zaiti MA (✉), Mohd Rushdan MN, Lee SJ

Deptartment of Obstetric & Gynaecology, Hospital Sultanah Bahiyah, Km 6, Jalan Langgar, 05460 Alor Setar, Kedah, Malaysia.

**Abstract**

Desmoplastic small round cell tumour (DSRCT) is a very rare malignant tumour which commonly presented as an intraabdominal tumour. It has a distinct histological and immunophenotypic characteristic which differentiates it from other types of small blue cell tumour such as Ewing's sarcoma, primitive neuroectodermal tumour, neuroblastoma and malignant mesothelioma. Apart from the abdomen, it may also originate from other region of the body including the reproductive organs.

**Keywords:** Desmoplastic, management, prognosis, tumour, uterus

**Correspondence:**

Nur Zaiti Md Arshad. Deptartment of Obstetric & Gynaecology, Hospital Sultanah Bahiyah, Km 6, Jalan Langgar, 05460 Alor Setar, Kedah, Malaysia. Tel: +604-7406233 Fax: +604-7406851 E-mail: atiearshad@yahoo.com

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## Introduction

Desmoplastic small round cell tumour (DSRCT) is a type of small roundcell neoplasm that has features similar to Ewing or primitive neuroectodermaltumours (PNET), but less responsive to the current chemotherapeutic agents (1). This rare and highly aggressive neoplasm usually diagnosed in males during adolescence or in their early adulthood.

Desmoplastic small round cell tumour (DSRCT) was first described by Gerald and Rosai in 1989. It is a rare, highly malignanttumour with male preponderance (1). This aggressive tumour may arise from the abdomen, ovaries, liver, kidneys and brain (2). We reported a case of a 69 year old postmenopausal lady with DSCRT arising from the corpus uterine.

## Case Report

A 69-years-old lady, para 5, postmenopausal for nineteen years, was referred to Gynaeoncology Clinic complaining of postmenopausal bleeding and gradually increasing abdominal mass for 8 months. She also had intermittent abdominal pain, early satiety and abdominal fullness.

On examination, there was a huge abdominal mass equivalent to 36 weeks size gravid uterus. The mass was mobile side to side with regular surface and hard in consistency. The vagina and cervix were normal. Her CA 125 level was 65.9 (high) and CA 19-9 was 6.6 (normal). Transabdominal scan showed a large multiloculatedmass with cystic, solid areas and papillary projection. A large pelvic tumour suspicious of ovarian malignancy was seen on CT scan (Fig. 1). No ascites or pelvic lymph nodes enlargement seen.

A total abdominal hysterectomy bilateral salphingoopherectomy with pelvic lymph nodes dissection was done. Intraoperatively the uterus was enlarged at 36 weeks size with areas of necrosis over posterior serosal layer of the uterus. Both ovaries were normal, the cervix and vagina were free from tumour. Microscopic examination showed extensive necrosis (>50%). The viable cells are small and harbor moderate nuclear pleomorphism with inconspicuous nucleoli. The cytoplasm is scanty to moderate (Fig. 2). No lymphovascular invasion. The tumour infiltrated

 

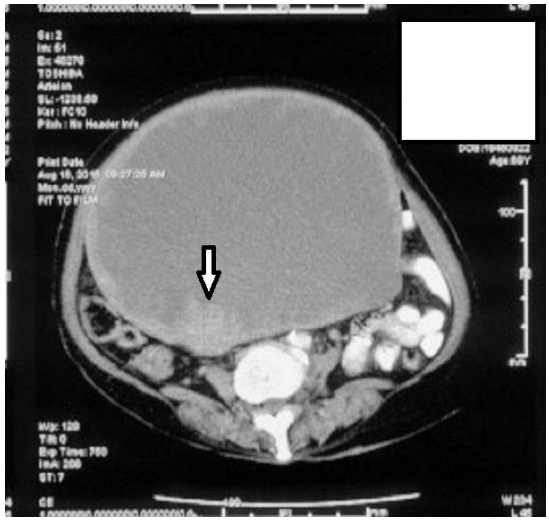

**Figure 1**: CT scan showed a large uniloculatedintraabdominal mass with cystic and small solid area (arrow).

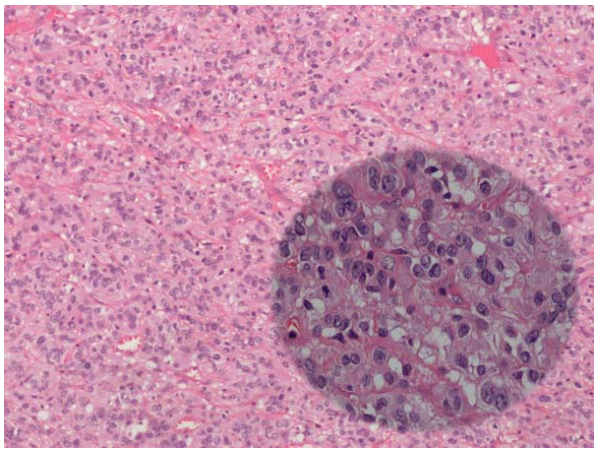

**Figure 2**: Microscopic examination showed presence of small blue cells with moderate nuclear pleomorphism with inconspicuous nucleoli and scanty to moderate cytoplasm characteristic of desmoplastic small round cell tumour.

into the myometrium from the serosal aspect with no involvement of the lower uterine segment and cervix. All lymph nodes were free from malignant cells. Immunohistochemical studies showed that the malignant cells were diffusely positive for vimentin, NSE and WT-1.

Based on the microscopic examination and immunohistochemical studies, the final diagnosis was Desmoplastic small round cell tumour Stage 1 based on FIGO Classification for Corpus Uteri Cancer 2009. The patient was discharged well post operatively. No further adjuvant chemotherapy or radiotherapy was given. Her CA 125 normalized at three weeks after the surgery. She recovered well with good performance status (ECOG 1). Further assessment at 3 and 6 months post-surgery showed that patient was well with no evidence of recurrence and her biochemical markers (CA125) were also normal.

**Discussion**

Desmoplastic small round cell tumour is a rare, highly malignant and aggressive tumour. Commonly affecting males with ratio of 4:1(3) in early 20s (4), only few reported cases of DSRCT in females were published. Less than 30 cases of intraabdominal DSRCT in adult women have been reported. Most of the patients with DSRCT presented at advanced stage hence the survival were poor.

Like the carcinoma of the ovary, most patients with DSRCT presented late. The common clinical presentation includes abdominal mass, pain, compressive symptoms such as constipation and incontinence and constitutional symptoms. Clinical examination may reveal a palpable huge abdominal mass with irregular border and surface and intraoperatively the tumour might be located either in the abdominal or pelvic cavity. Extensive peritoneal involvement with diffuse tumour is also a common finding during the surgery (2). DSRCTs may spread locally to the peritoneum or omentum and distally to the liver, lungs and bone marrow (5). The clinical manifestation of DSRCT can mimicked ovarian malignancy making the diagnosis difficult without surgical evaluation and histopathologic confirmation. This patient presented with postmenopausal bleeding as the tumour has infiltrated the myometrium.

Desmoplastic small round cell tumour is diagnosed based on its distinct characteristic on HPE and IHC. The characteristic appearance of undifferentiated small blue cells with nuclear pleomorphismand scanty cytoplasm is seen microscopically. Immunohistochemistry is important in differentiating DSRCT from other small round cell tumour. This study may reveal the coexpression of epithelial (cytokeratin), mesenchymal (vimentin and desmin) and neural markers (NSE and neurofilament) in the same tumour cell which is diagnostic of DSRCT (6). The other marker includes WT1 and CD99 (7).

The management of DSRCT and the impact of the treatment modality to the survival rate has only been studied in a small number of patient. Surgery, chemotherapy,and radiotherapy have been used in the treatment of DSRCT (7).

DSRCT with extensive peritoneal involvement makes complete surgical resection rarely possible, especially

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in those with distant metastasis to the liver, hepatic veins or involvement of the diaphragm (5). The impact of surgery upon survival remains unclear however if a complete resection is achieved, the survival rate improved especially without metastatic spread (8). Hassan et al. found that the patients with DSRCT who underwent surgical excision has 34 months survival rate as compare to 14 months in those who had undergone biopsy only (9). It was also found that patients who undergone gross tumour resection has improved survival (10). Debulking surgery has a role in symptomatic relief from the compressive effect of the tumour.

The response of DSRCT to conventional chemotherapy regime is poor (7). Multidrugs chemotherapyhas been used in DSRCT however none have shown curative outcomes (8). Bertuzzi et al. found that those with DSRCT had a very poor response to multidrug therapy (11). The overall response rate from DSRCT group was 43% as compared to overall response of 85% in other small round cell tumour groups. Kushner et al. used neoadjuvantmultidrug chemotherapy followed by debulking surgery (12). All of the patients showed some response however no complete pathologic responses seen. Thus, debulking surgery to remove the bulky tumour and multidrug therapy offers a better prognosis longer progression-free interval (12).

Radiotherapy has not been used as a primary therapy in DSRCT (7). Radiotherapy has been used in combination with surgery and chemotherapy but not alone. A combination of chemotherapy, surgical debulking, and external beam radiation used in a study found an overall survival of 48% (13). The median time to relapse was 19 months and the most common site of recurrence is intraperitoneal and/or hepatic recurrence.

## Conclusion

Desmoplastic small round cell tumour is a rare aggressive tumour which may also arise from the reproductive organs. Though it is rare, and clinical presentation may mimic other types of uterine malignancy, DSRCT should be considered in patients presented with advanced malignancy as it demonstrates distinct histological features and unique cytogenetic profile as compared to other types of small cell tumour. There were limited reports on best treatment option for DSRCT due to small number of reported cases and most patients presented at late stage, hence had a poor outcome. However complete surgical resection in early disease appears promising for a better survival. As for this patient, she presented at early stage with complete surgical resection and she is diseased free for almost one year up to date, we are optimistic for a longer survival rate though the long term sequelae of the disease is yet to be evaluated.

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