

Case report

Desmoplastic small round cell tumor involving the uterine cervix: The first reported case in the literature, and brief review of gynecologic presentations

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ARTICLE INFO

Keywords

Cervical cancer
Sarcoma

ABSTRACT

Background: Desmoplastic small round cell tumors are exceedingly rare, usually involve abdominal organs and predominantly affect male patients. We describe the first reported case arising from the uterine cervix and provide a summary of 20 previously reported cases involving gynecologic organs.

Case: A 54 year-old was diagnosed with a rapidly growing 13 cm desmoplastic small round cell tumor of the cervix. She was treated through a multimodal approach involving neoadjuvant chemotherapy and surgery. She subsequently recurred, and this was successfully treated with radiation therapy. She is well and without evidence of disease 22 months after initial diagnosis.

Conclusion: We report successful treatment through multidisciplinary and multimodal management. This can guide management of future patients as no gold-standard treatment has yet been described.

1. Case presentation

A 54-year-old G3P2 female patient presented to her family physician with a three-month history of daily vaginal bleeding. On past medical history she was healthy and was an ex-smoker. The patient was unsure whether she had undergone menopause as she was taking oral contraceptive pills in a continuous manner. The patient had previously undergone cervical cancer screening with pap smears. The 2 most recent ones, dating from 10 years and 6 years prior to this diagnosis, were normal. Her family physician performed a pap smear, and this revealed atypical glandular cells. She was referred to the Colposcopy clinic in view of this result.

She underwent her initial Colposcopy appointment approximately 5 months following symptom onset. Apart from increasing vaginal bleeding and pelvic pain, she denied other symptoms. Upon speculum examination, there was a partly necrotic tumour involving the entirety

of the cervix measuring at least 8 cm, with involvement of the vaginal fornices. On pelvic examination, there was suspected bilateral parametrial involvement, and no obvious bladder or rectal invasion. Cervical biopsies were taken, and a CT scan and MRI were arranged. On initial blood work, her hemoglobin was 81 g/L, her creatinine was 87 μmol/L, and electrolytes and liver enzymes were normal.

1.1. Pathology findings

This lesion was consistent with a desmoplastic small round blue cell tumour (DSRCT) of the cervix. Microscopic sections revealed a poorly differentiated malignancy with small round cell morphology and extensive areas of necrosis, with occasional pseudorosette-like structures (Fig. 1A). Cells were positive for vimentin with rare AE1/AE3 positive cells (Fig. 1B), desmin (Fig. 1C) and WT-1 (Fig. 1D).

Ewing sarcoma was excluded based on the absence of EWSR1 loci

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<https://doi.org/10.1016/j.gore.2022.101079>

Received 8 August 2022; Received in revised form 3 October 2022; Accepted 5 October 2022

Available online 7 October 2022

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rearrangement on the 22q12 gene, and poorly differentiated synovial sarcoma was excluded based on the absence of SS18 loci rearrangement on the 18q11.2 gene. Thus, this was classified as desmoplastic small round blue cell tumour of the cervix.

DSRCT is an aggressive soft tissue sarcoma, first described in 1989. The histological appearance consists of undifferentiated, small round blue cells surrounded by abundant desmoplastic stroma. (Ordóñez, 1998) Distinct cytogenetic feature of DSRCT can involve a chromosomal translocation involving an Ewing's sarcoma gene (EWS) and Wilms tumour gene (WT1), which result in the formation of a chimeric gene. (Gerald et al., 1995) This chimeric gene encodes for a chimeric protein with oncogenic features, including production of endogenous platelet-derived growth factor. (Gerald et al., 1995).

1.2. Radiology findings

Six days after her Colposcopy consultation, a CT of the chest, abdomen, and pelvis with IV contrast (Fig. 2) was performed and revealed an 8.9 × 8 × 8 cm necrotic cervical mass with loss of planes between the mass and the left pelvic sidewall and left iliac vessels. There was no intralesional calcification. There was suspected bilateral parametrial involvement. Enlarged obturator and bilateral inguinal lymph nodes were identified, measuring up to 1 cm. There was no metastatic disease in the chest.

A pelvic MRI without intravenous contrast (Fig. 3) was performed 19 days after the CT scan and revealed that the mass had rapidly increased in size to 13.1 × 9.3 × 8.9 cm. The mass had heterogeneous signal which was predominantly T2 hyperintense to skeletal muscle but also had some areas which were isointense or hypointense to skeletal muscle. No hemorrhage was present. The MRI better delineated the primary location of the mass along the anterior cervix with additional involvement of the anterior vaginal wall and fornix, lower uterine segment, left adnexa and probable small areas of bladder invasion. There was extension into the left parametrium and to the left pelvic sidewall resulting in

hydroureter. The hydroureter was progressive compared to the prior CT. External iliac lymph nodes now measured 1.1 and 1.2 cm.

1.3. Multidisciplinary management

This case was presented at the Gynecologic Oncology as well as the Sarcoma multidisciplinary case conferences. The consensus was that this mass was not surgically resectable, and that radiation therapy would be reserved for symptom management or in the recurrent setting. She was offered the vincristine, doxorubicin and cyclophosphamide (VDC) protocol (Subbiah et al., 2018) every 3 weeks under the care of the Sarcoma team. After an initial 5 cycles, which she tolerated well with minimal side effects, she was found to have good radiologic response. A CT scan revealed minimal residual disease and a left external iliac lymph node measuring 0.9 × 0.5 cm.

She was examined for consideration of surgery and deemed a good candidate. Upon speculum and pelvirectal examination, there was an approximately 2 cm tumor remaining. She underwent a laparotomy, radical hysterectomy, bilateral salpingo-oophorectomy, bilateral sentinel pelvic lymph node biopsy. A full lymphadenectomy was not recommended as this was a completion surgery after chemotherapy. Radiology review had confirmed that there was no residual pathologically enlarged lymph node. At the time of surgery, there were no palpable enlarged lymph nodes. Surgical pathology confirmed the initial diagnosis, and lymph nodes were negative. The surgical margins were negative but close, and for this reason, adjuvant radiation therapy was recommended. FISH analysis on the surgical specimen confirmed a EWSR1 translocation in 12 % of cancer cells adding support to the diagnosis of DSRCT. RNA sequencing was performed on the surgical specimen and there were no reportable single nucleotide variants or gene fusions identified.

In the adjuvant setting, the patient was planned to receive 54 Gy in 25 fractions to the pelvis and vaginal vault, 45 Gy in 25 fractions to other areas of the pelvis and nodes at risk, and a single fraction of

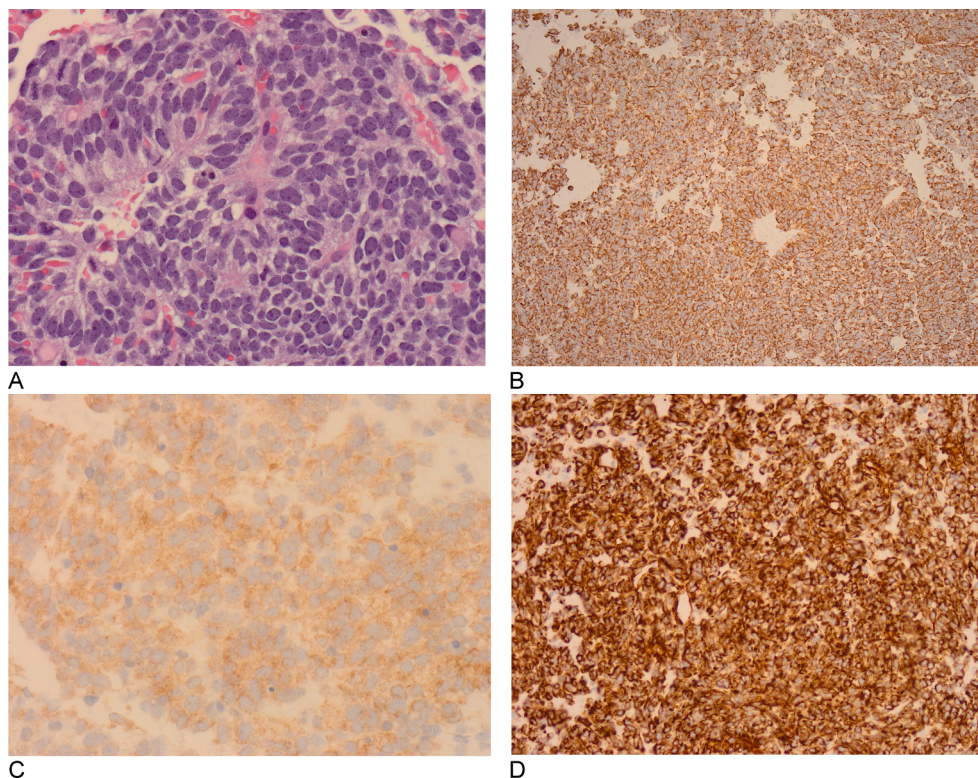


Fig. 1. (A) High power hematoxylin and eosin image showing small round blue cell morphology and extensive areas of necrosis (400x). Positive staining for (B) vimentin (40x), (C) desmin (400x) and (D) WT-1 (200x).

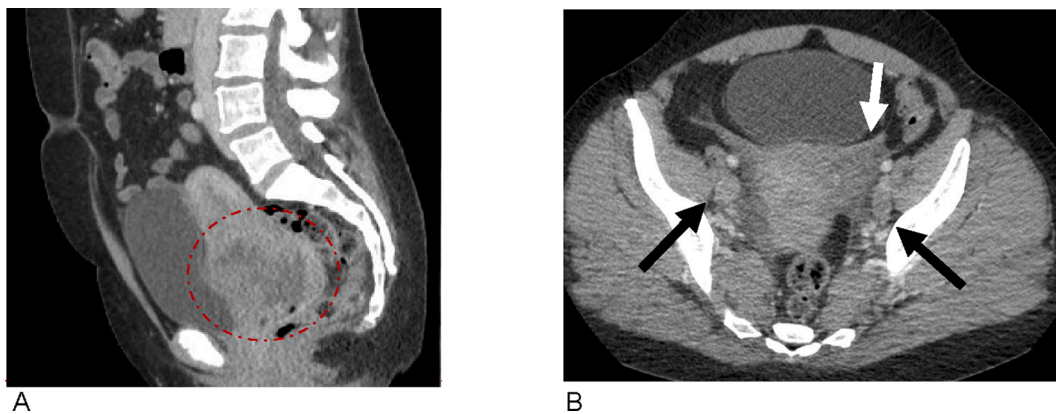


Fig. 2. Initial CT: (A) Sagittal image showing poorly delineated necrotic mass (indicated by red outline). Locule of gas inferiorly relates to recent biopsy. (B) Axial image demonstrating left parametrial/adnexal involvement extending towards the pelvic sidewall (white arrow) and bilateral external iliac chain lymphadenopathy (black arrows).

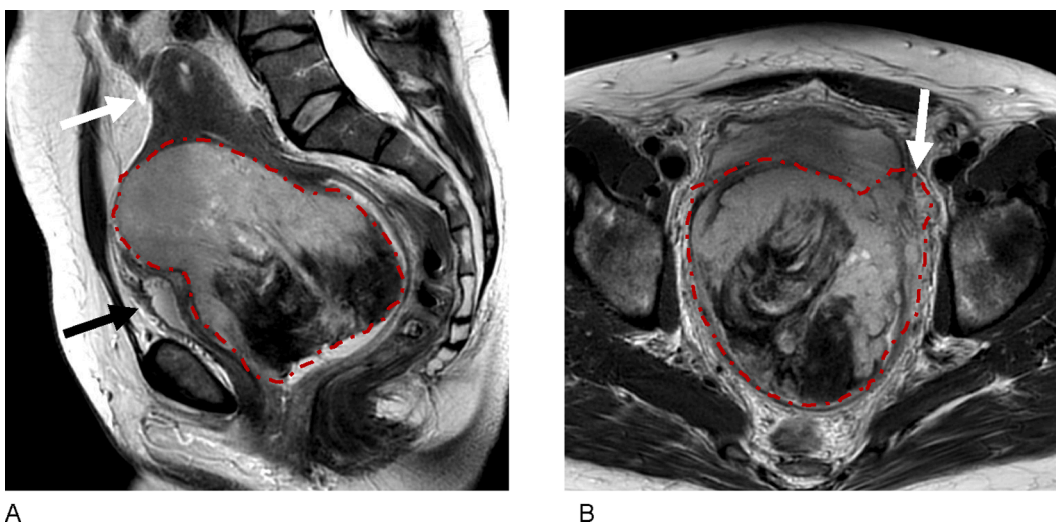


Fig. 3. Initial MRI: (A) Sagittal T2-weighted image showing heterogeneous anterior cervical mass (outlined in red) with involvement of the lower uterine segment, anterior vaginal wall/fornix, and probable small areas of posterior bladder wall involvement. The uterine body is indicated by the white arrow and the partially distended bladder by the black arrow. (B) Axial T2-weighted image showing heterogeneous mass (outlined in red) and left parametrial invasion (white arrow). Hydroureter is not shown.

brachytherapy of 7 Gy to the upper 3 cm of the vagina. However, upon CT and MRI planning prior to radiation treatment, she was found to have asymptomatic recurrent disease at the left pelvic sidewall resulting in hydroureter. The recurrent mass measured $3.1 \times 2.6 \times 2$ cm on MRI (Fig. 4). The radiation plans were thus adapted. This recurrence was successfully treated with 45 Gy to the larger nodal volume, 54 Gy to the area of close margins and 63 Gy to gross disease (Fig. 5), resulting in complete radiologic response.

She then received 1 additional cycle of VDC, followed by 3 cycles of vincristine and cyclophosphamide without doxorubicin, as the maximum cumulative dose of doxorubicin had been reached. CT scan and PET scan 1 month following this treatment revealed complete resolution of disease (1 year after initial treatment).

The patient is now undergoing observation under the care of her multidisciplinary team. She was most recently seen 22 months following initial presentation, and 9 months since completion of her last chemotherapy cycle, with no evidence of residual disease on physical examination and CT scan. She is not reporting any long-term side effects from this multimodal treatment.

2. Discussion

DSRCT is exceedingly rare with an annual incidence of approximately 0.1 case/million. It often presents in young adults in their early 20s, with the majority of cases affecting males. (Ordóñez, 1998; Morani et al., 2019 Mar) It is estimated that there are 200 to 450 cases reported in the literature. (Morani et al., 2019 Mar) Patients tend to present with nonspecific symptoms such as abdominal pain and distention, constitutional symptoms, and/or a palpable mass. DSRCTs most often originate in the abdomen, with approximately half of patients presenting with metastatic disease, often involving the liver, lungs, pleura, and mediastinum. DSRCTs of gynecologic origin are extremely rare, and we described, to our knowledge, the first reported case involving the uterine cervix. The overall prognosis is poor, with a 5-year overall survival rate reported at approximately 15–30%. (Subbiah et al., 2018; Morani et al., 2019 Mar) The rarity of DSRCTs, especially of gynecologic origin, along with current poor prognosis of the clinical course highlights the importance of reporting patient clinical presentation and evaluating current treatment regimens.

Histologically, DSRCTs consist of nests of tumour cells encompassed by dense desmoplastic stroma. (Ordóñez, 1998) However, DSRCT tumour cells co-express a variety of markers, including mesenchymal

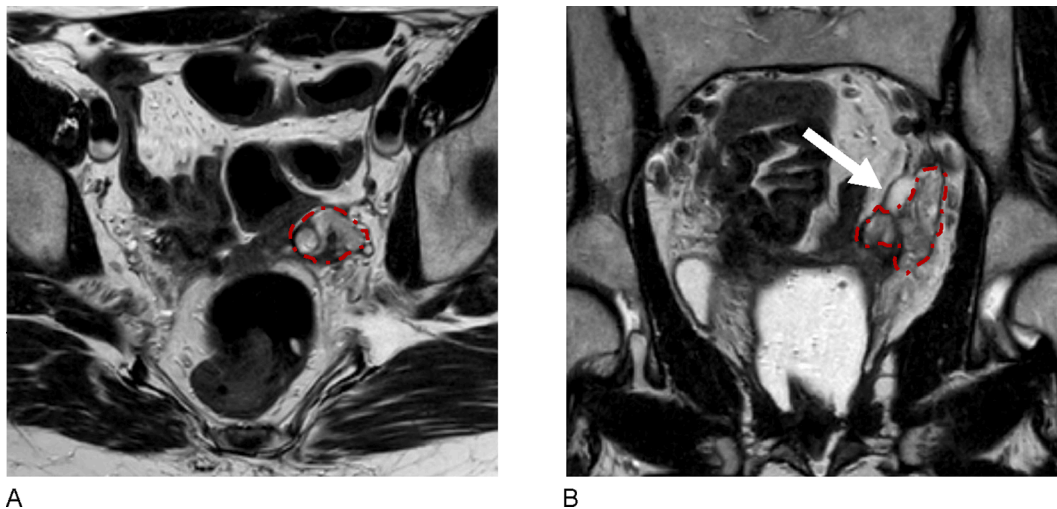


Fig. 4. Follow-up MRI 7 months after initial MRI: (A) Axial oblique T2-weighted image and (B) Coronal oblique T2-weighted showing intermediate signal serpiginous recurrent tumor along the left vaginal cuff near the pelvic sidewall measuring $3.1 \times 2.6 \times 2$ cm. Its location resulted in moderate left hydronephrosis (white arrow). The vagina is distended by gel on image B.

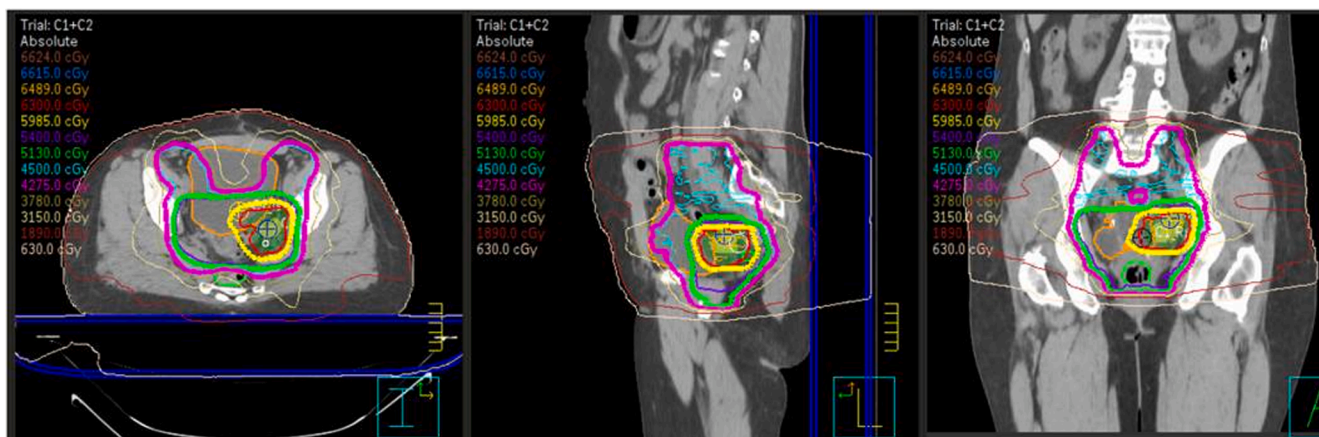


Fig. 5. Radiation plan: 45 Gy were given to the area delineated in pink, 54 Gy to the area of close margins delineated in green, and 63 Gy to the gross recurrent tumor delineated in yellow.

(vimentin, desmin), epithelial (epithelial membrane antigen, cytokeratin), and neural (neurone specific enolase, CD56) cell markers. (Ordóñez, 1998) The co-expression of these markers by the tumour cells differentiates DSRCTs from other small round cell tumours. Along with this distinguishing characteristic, the appearance is of high cellularity, with a multitude of undifferentiated uniform round cells of small to medium size, with dispersed cytoplasm. The nuclei are generally round shaped, arranged in either a nest of spindle cell formation, with granular chromatin that resemble small cell carcinoma.

Due to the rarity of this disease, there is currently no standardized treatment for DSRCTs. In patients presenting with resectable disease, complete and aggressive surgical resection is the cornerstone of treatment. (Young et al., 1992) However, as DSRCT commonly presents as metastatic disease, complete resection is often not feasible. A multimodal therapeutic strategy is often recommended, and a combination of surgery, chemotherapy and radiation therapy has been reported to lead to improved prognosis.

There has been evidence of chemosensitivity of DSRCTs reported in the literature. Neoadjuvant chemotherapy in combination with surgical cytoreduction has been shown to improve patient outcomes. (Subbiah et al., 2018; Young et al., 1992) Adjuvant chemotherapy may also be provided after incomplete surgical resection to hinder further disease progression. There is no standardized chemotherapy regimen.

Commonly prescribed regimens include combinations of doxorubicin, vincristine, dactinomycin, cyclophosphamide, ifosfamide, and etoposide, some of which were administered to the presenting case as per the VDC protocol. (Subbiah et al., 2018; Young et al., 1992) Second line agents include temozolomide/irinotecan, cyclophosphamide/topotecan, and high dose ifosfamide, all of which have shown clinical benefit in chemotherapy resistant Ewing sarcoma. Patients typically receive several cycles prescribed at 3 week intervals, although some have been reported to receive dose-dense therapy biweekly. (Subbiah et al., 2018; Young et al., 1992).

Similarly, DSRCT radiosensitivity has been reported. Although radiation therapy alone may not significantly improve patient outcomes, it has been reported that adjuvant radiation therapy in combination with surgery has a survival advantage for patients in comparison to patients who did not receive radiation therapy. Furthermore, adjuvant radiation therapy may be used as consolidation chemotherapy, which has been demonstrated to lead to improved outcomes in comparison to chemotherapy and surgery alone (Subbiah et al., 2018). In this case report, we described excellent response to the VDC chemotherapy regimen, as well as excellent radiosensitivity at the time of tumor recurrence.

2.1. Review of gynecologic DSRCTs

cases of gynecologic DSRCT, 16 of which were of primary ovarian origin.

There is currently limited literature regarding DSRCTs in female patients. In Table 1, we provide a summary of 20 previously reported

Table 1
Overview of Reported Gynecological Cases of Desmoplastic Small Round Cell Tumour.

| Author and year | Age of patient at diagnosis | Organ involved | Primary Treatment | Outcome | First recurrence | Treatment of recurrence | Survival |
|-----------------------------------------------------------|-----------------------------|--------------------------|---------------------------------------------------------------------------------------------------------|--------------------------------------------------------------|--------------------------------------------|------------------------------------------------------------|------------------------|
| (Young et al., 1992) | 15 | Ovarian | Surgery, multi-agent systemic chemotherapy including carboplatin | N/A | N/A | N/A | 4 months |
| | 15 | Ovarian | Surgery | N/A | 7 months | Surgery | Unknown |
| | 14 | Ovarian, Uterine | Surgery | N/A | N/A | N/A | Unknown |
| Zadoulek et al., 1995(Zaloudek et al., 1995) | 22 | Ovarian | Surgery, 6 cycles BEP | Suboptimal resection, partial response to chemotherapy | 12 months | N/A | 18 months |
| Slomovitz et al., 2000(Slomovitz et al., 2000) | 11 | Ovarian | Cycles 1–3, 6: VDC; Cycles 4–5: IE | Very good radiologic response | 9 months | Topotecan | 11 months |
| Elhajj et al., 2002 (Elhajj et al., 2002) | 27 | Ovarian | Surgery | Suboptimal resection | 11 months | 3 cycles etoposide, cisplatin, then 6 cycles high-dose VDC | 42 months |
| Church et al., 2006 (Church et al., 2006 Sep 1) | 23 | Vaginal, abdomino-pelvic | 6 cycles alternating cisplatin, doxorubicin vs TIP, cisplatin and mesna uroprophylaxis | Minimal response to chemotherapy | 20 months | 6 cycles palliative carboplatin, etoposide | 27 months |
| | 29 | Uterine | 6 cycles alternating BEP and TIP | No response (recurrence 6 weeks post-chemotherapy) | 7 months | 1 cycle CAP | 12 months |
| Engohan-Aloghe et al., 2009 (Engohan-Aloghe et al., 2009) | 21 | Ovarian | Surgery, chemotherapy | Partial response to chemotherapy | N/A | N/A | 7 months ⁺ |
| Ota et al., 2010(Ota et al., 2010) | 26 | Ovarian | Surgery, chemotherapy involving VDC/IE as per P6 protocol | Partial response | N/A | N/A | 23 months |
| | 19 | Ovarian, Uterine | Surgery, 5 cycles BEP | Partial response | 5 months after secondary debulking surgery | N/A | 11 months |
| D'Ippolito et al., 2012(D'Ippolito et al., 2012) | 29 | Ovarian | Surgery | No evidence of disease recurrence | N/A | N/A | 25 months ⁺ |
| Monappa et al., 2013(Monappa et al., 2013 Dec) | 32 | Pelvic mass | 3 cycles cisplatin and paclitaxel | N/A | N/A | N/A | 36 months* |
| Nakayama et al., 2014(Nakayama et al., 2014) | 6 | Ovarian | Surgery, 7 cycles IE/VDC | Suboptimal resection | 3 months | RT | 28 months |
| | 28 | Ovarian | 14 cycles neoadjuvant IE/VDC, surgery, 2 cycles adjuvant IE/VDC. Removal of doxorubicin after 8th cycle | Incomplete response to chemotherapy | 6 months post chemotherapy | cyclophosphamide/topotecan | 40 months |
| | 17 | Ovarian | Surgery, laser ablation, 9 cycles IE/VDC, whole abdominal radiation | Optimal resection but visible sub-centimeter disease present | N/A | N/A | 11 months ⁺ |
| Xie and Shen, 2016 (Xie and Shen, 2016 Feb 1) | 30 | Ovarian | Surgery, chemotherapy with VDC | Partial response to chemotherapy | N/A | N/A | 15 months ⁺ |
| Fagouri et al., 2018 (Fagouri et al., 2018) | 16 | Uterine | 3 cycles doxorubicin, ifosfamide, G-CSF | N/A | Not indicated | Surgery | 6 months |
| Altal et al., 2019 (Altal et al., 2019) | 23 | Ovarian | 2 cycles of VDC as per P6 protocol | Poorly tolerated | N/A | N/A | 4 months |
| Vujic et al., 2020 (Vujic et al., 2020 May) | 19 | Ovarian | Surgery, 3 cycles IVA, 3 cycles CEV, 3 cycles IVE | Optimal resection, disease remission | N/A | N/A | 40 months ⁺ |
| Present case | 54 | Uterine cervix | Neoadjuvant 5 cycles VDC, surgery. | Good response to treatment (minimal residual disease) | 11 months | RT, consolidation with VDC | 22 months ⁺ |

RT: radiation therapy; BEP: bleomycin, etoposide, cisplatin; VDC: vincristine, doxorubicin, cyclophosphamide; IE: ifosfamide, etoposide; TIP: paclitaxel, ifosfamide, cisplatin; CAP: cyclophosphamide, doxorubicin, cisplatin; IVA: ifosfamide, vincristine, actinomycin; CEV: carboplatin, epirubicin, vincristine; IVE: ifosfamide, vincristine, etoposide.

* Lost to follow up, + Alive when reported.

3. Conclusion

In conclusion, DSRCT is a rare mesenchymal tumour more commonly affecting young male patients. DSRCTs involving gynecologic organs are very rare. We described, to our knowledge, the first reported case originating in the uterine cervix. We report a multimodal treatment approach, with excellent response to the VDC regimen as well as radio-sensitivity at the time of recurrence.

Funding

The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Patient consent for publication

Written informed consent was obtained directly from the patient.

CRedit authorship contribution statement

Raveena Kapoor: Conceptualization, Data curation, Formal analysis, Investigation, Methodology, Writing – original draft. **Monalisa Sur:** Data curation, Formal analysis, Investigation, Writing – review & editing. **Yoan K. Kagoma:** Data curation, Formal analysis, Investigation, Writing – review & editing. **Kara Schnarr:** Data curation, Formal analysis, Investigation, Writing – review & editing. **Kevin Zbuk:** Data curation, Formal analysis, Investigation, Writing – review & editing. **Julie My Van Nguyen:** Conceptualization, Data curation, Formal analysis, Investigation, Methodology, Project administration, Supervision, Visualization, Writing – original draft, Writing – review & editing.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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