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Peritonectomy and hyperthermic intraperitoneal chemotherapy as treatment for desmoplastic small round cell tumour

Howard S. Fan[a], Bernie I'Ons[b], Ryan McConnell[a], Varahini Kumar[a], Saleh Alzahrani[a], David L. Morris[a,*]

[a] Department of Surgery, University of New South Wales, St George Hospital, Kogarah, NSW 2217, Australia
[b] Department of Anatomical Pathology, University of New South Wales, St George Hospital, Kogarah, NSW 2217, Australia

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ABSTRACT

INTRODUCTION: The St George Hospital specialises in peritonectomy and hyperthermic intraperitoneal chemotherapy (HIPEC) for treatment of intra-abdominal malignancies. Despite performing around 800 peritonectomy and HIPEC procedures, we have rarely encountered desmoplastic small round cell tumours (DSRCT). We present our experiences with DSRCT, and propose peritonectomy and HIPEC as a treatment option for DSRCT.

PRESENTATION OF CASE: This is a case series of 3 cases. The first case was a 26-year-old male who presented with appendicitis which we diagnosed as DSRCT and treated with peritonectomy and HIPEC. The second case was a 14-year-old male referred to our centre for peritonectomy and HIPEC after initial presentation with a pelvic mass and treatment with chemotherapy. The third case was a 21-year-old male referred to our centre for peritonectomy and HIPEC for recurrent DSRCT after previously being treated with neoadjuvant chemotherapy and surgery without HIPEC.

DISCUSSION: DSRCT is a rare, almost exclusively intra-abdominal malignancy, which predominantly affects young males. Survival prognosis remains poor in DSRCT despite conventional treatment with surgery, chemotherapy and radiotherapy; however, HIPEC has offered promising survival results. Our recurrences with peritonectomy and HIPEC at 6 months and 15 months are comparable with the literature of 8.85 months.

CONCLUSION: In our experience, patients with DSRCT who present with nodal involvement or recurrent disease tend to recur early despite treatment with peritonectomy and HIPEC. Longer term follow up of our patients and future studies involving HIPEC in DSRCT would be useful in assessing long-term clinical outcomes and survival.

1. Introduction

The St George Hospital specialises in peritonectomy and hyperthermic intraperitoneal chemotherapy (HIPEC) for treatment of intra-abdominal malignancies. Despite performing around 800 peritonectomy and HIPEC procedures, we have rarely encountered desmoplastic small round cell tumours (DSRCT). We present our experience with DSRCT and HIPEC in a series of three cases, and propose peritonectomy and HIPEC as a treatment option for DSRCT.

1.1. Case 1

A 26-year-old gentleman presented with localised right iliac fossa pain after having a colicky lower abdominal pain, which lasted 24 h associated with anorexia, and nausea but no vomiting. His bowel motions were regular, and he did not notice any blood, diarrhoea or tenesmus. He did not notice any weight loss and had been well prior to this episode. Apart from a previous cholecystectomy, his past medical and family history were unremarkable. On examination he was afebrile, tachycardic, had a soft non distended abdomen with no palpable hernias or masses. He was tender over the right iliac fossa, hypogastrium and umbilical regions with guarding, rebound tenderness, and positive Rovsing's sign.

Given the high clinical likelihood of appendicitis, the patient was taken for laparoscopic appendicectomy without imaging. Intraoperatively the appendix was found to be normal, however, a large mass was found on the right hepatic flexure with intra-abdominal pus noted. Given this finding, the procedure was converted to an open appendicectomy and right hemicolectomy with the mass sent off for histological assessment.

A 60 mm mass was resected from the colon with margins of 30 mm distally and 140 mm proximally. No masses were seen

* Corresponding author. Tel.: +61 91133070.
E-mail address: david.morris@unsw.edu.au (D.L. Morris).

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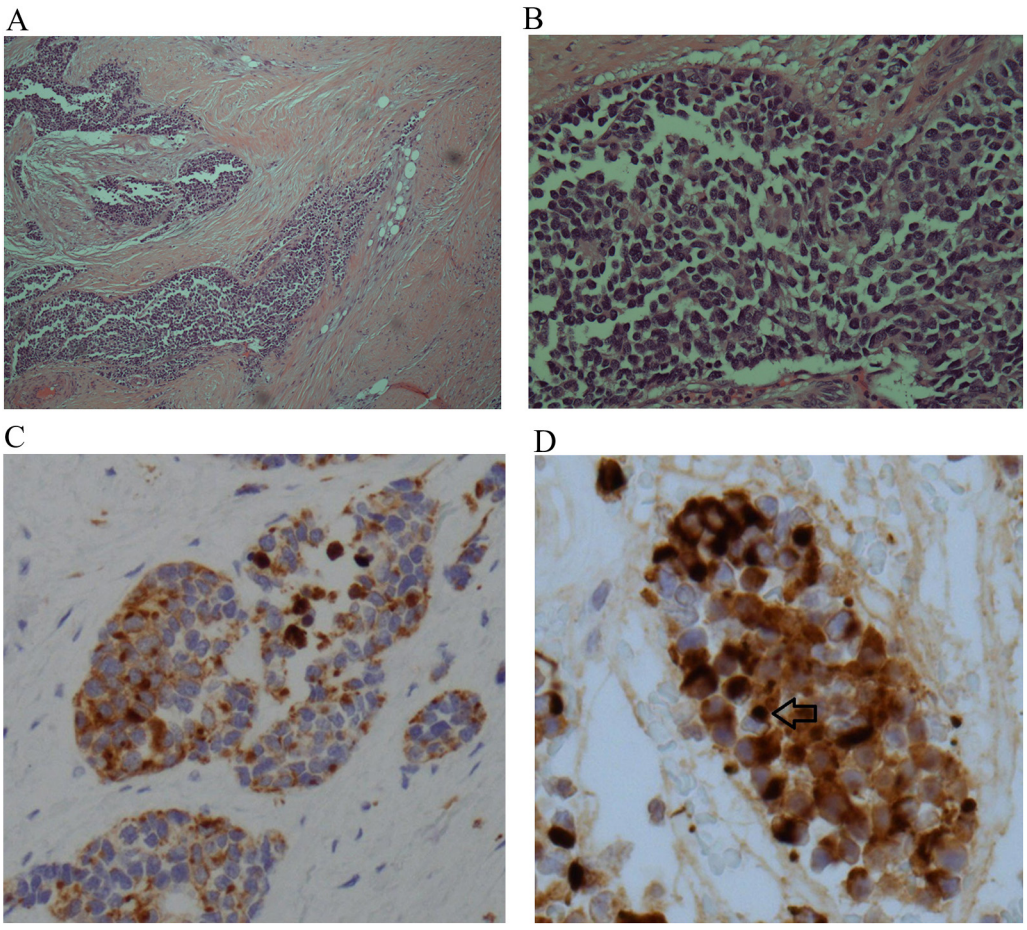

Fig. 1. (A) Nests of tumour cells surrounded by desmoplastic stroma (magnification: 100×). (B) Cells with hyperchromic nuclei and scant cytoplasm (magnification: 400×). (C) Immunohistochemical stain positive for cytokeratin (CAM 5.2). (D) Immunohistochemical stain positive for desmin, with immunoreactivity present in a typical perinuclear dot-like Golgi pattern (large arrow).

on the serosal surfaces. The tumour involved the full thickness of the colon wall invading through the serosal fat, just reaching the serosal surface. No malignancy was identified within fifteen pericolic lymph nodes. Histology and immunohistochemistry confirmed a high grade undifferentiated DSRCT with a Ki67 of 30%. The diagnosis of DSRCT was made on the basis of the tumour morphology showing nests of tumour cells with hyperchromic nuclei and scant cytoplasm surrounded by desmoplastic stroma, and positive immunohistochemistry staining for desmin and cytokeratin (Fig. 1). Furthermore, fluorescence in situ hybridisation (FISH) techniques revealed positive EWSR1 arrangements, which further supported our diagnosis. Interestingly, the tumour did not stain positive with the WT1 antibody which is present in the majority of DSRCT.

Staging workup with chest and abdomen CT, hepatic angiogram CT and PET scans revealed a stage 1 DSRCT. Given the aggressive nature of DSRCT, the patient was treated with a potentially curative peritonectomy two weeks later with the aim of removing microscopic intraperitoneal disease and improving long term survival. The intraoperative assessment revealed a peritoneal cancer index (PCI) of 4, with a small volume of disease in the pelvis. A revision of the ileocolic anastamosis, omentectomy, pelvic peritonectomy, excision of the umbilicus and intraoperative ultrasound for hepatic lesions was subsequently performed. The procedure achieved a complete cytoreduction of the tumour with a completeness of cytoreduction (CC) score of 0. Our procedure involved HIPEC with cisplatin at 41.5 °C for 90 min. Postoperatively, urine output was maintained at 100 ml/h for 24 h given the nephrotoxicity of cisplatin. The repeat histopathology showed peritoneal deposits of DSRCT in the ileocolic anastamosis, and pelvic peritoneum. The surgical margins of resection were clear. No tumour was detected in the lymph nodes examined.

The patient's recovery was complicated by a pulmonary embolus on postoperative day 10, for which heparin infusion was initiated and subsequently switched to therapeutic enoxaparin. The patient was discharged from hospital on postoperative day 19 and commenced on systemic chemotherapy using cyclophosphamide, doxorubicin and vincristine. The patient had one episode of febrile neutropenia after his first cycle of chemotherapy and was admitted to hospital for intravenous antibiotics. He has since tolerated his cycles well and was on his fourth of nine cycles at the time of submission and would continue with ongoing oncologist follow up. The patient had repeat colonoscopies and PET scans with no evidence of recurrence at 6 months post peritonectomy. He remained asymptomatic with ECOG performance status grade 0, and was continuing with gym training.

1.2. Case 2

A 14-year-old male was referred to our unit with stage 2 DSRCT after being treated with 11 weeks of vincristine, doxorubicin, ifosfamide, and etoposide. He originally presented with a large pelvic mass which was compressing against his right ureter, bladder and prostate. He was referred to our centre after his initial chemotherapy and had an intraoperative PCI of 12. We performed a laparotomy for right diaphragm strip, cholecystectomy, right