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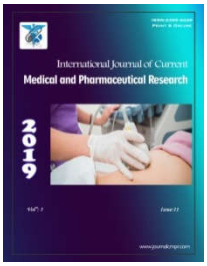

Research Article

DESMOPLASTIC SMALL ROUND CELL TUMOUR OF THE PERITONEUM (A CASE REPORT) AND LITERATURE REVIEW

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ABSTRACT

The peritoneum is a serious lining of mesothelial cells with a rich vascular and lymphatic capillary network that covers the abdominal and pelvic walls and organs. Peritoneal neoplasm can originate de novo from the peritoneal tissues and defined then as primary peritoneal carcinomatosis (PPC), invade or metastasize into the peritoneum from adjacent or remote organs and will be known as secondary peritoneal carcinomatosis (SPC), No ethnic predisposition or other known risk factors have been identified as specific for the disease. Highly aggressive tumour usually occurs in males during adolescence and early adulthood. Desmoplastic small round cell tumor (DSRCT) is a distinctive clinicopathologic entity and immunophenotypic feature with an aggressive clinical course that typically involves the abdominal and/or pelvic peritoneum of young males. It behaves aggressively and only 29% of patients survive up to 3 years (1).

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INTRODUCTION

Desmoplastic small round cell tumour

Primary peritoneal carcinomatosis (PPC) usually diagnosed in their third or fourth stage. In the third stage, cancer is present in the lining of the abdomen (peritoneum) whereas at the stage 4, it will expand to places such as the liver, lungs, or distant lymph nodes (1). Because of its rarity, patients with DSRCT have not been included in clinical trials therefore severely limiting our understanding of the disease and our access to patient outcome data., less than 200 cases has been reported in the literature and since then multimodality treatment including chemotherapy, radiation and aggressive surgery, has not improved outcome(1).

Despite multimodal treatment, including aggressive surgical excision, chemotherapy, and radiotherapy, multiple series have shown that approximately 60–70% of patients succumb to their disease within 3 years, and most experience resistant and recurrent disease before the end of life(2)

The neoplasm usually appears as an extensive intra-abdominal or less often endo-pelvic mass with widespread peritoneal and lymphatic dissemination, without an apparent organ of origin. Patients may present with dozens to hundreds of tumors studding the peritoneal cavity. Despite this presentation, it is not primarily considered metastatic but multifocal (2). Other areas less often affected may include lymph nodes, the lining of the abdomen, the diaphragm, hollow viscous, splenic hilum, mesentery of small and large bowel, and at the pelvis peritoneum between the bladder and rectum. The most common sites of metastatic spread are the liver, lungs and bones.

DSRCT typically arises from abdominal or pelvic peritoneum as a diffuse mass, which tends to be large at presentation-up to 40 cm in some cases. It is associated with abdominal distension and pain, ascites and hepatomegaly. Pressure effects of the tumour on the nearby structures had been described, such as intestinal obstruction, hydronephrosis and urinary/erectile dysfunction (3).

Despite the enormous size the tumors can grow to, there are few early warning signs. The majority of patients are young and healthy as the tumor grows and spreads uninhibited within the abdomen or pelvis. The rarity of this tumour and the similarities it shares with the other small round cell tumours still make the diagnosis challenging.

CASE REPORT

17 year-old young female presented with 6 months history of increasing fatigue, pallor and dyspnea at rest associated with epigastric and right upper quadrant abdominal pain, several

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episodes of melena decreased appetite, early satiety and unintentional weight loss (15Kgs).

Physical examination showed mild tachycardia with a heart rate of 106 bpm, Blood pressure 100/65mmHg.Cardiac examination was free of murmurs. Her abdominal examination was positive for a palpable hard, tender RUQ masses, measuring 7*5 cm. Rectal examination showed no fissures, tags, or fistulae; the rectal vault contained Melenic stool residues that was strongly positive for occult blood.

Her labs. studies : WBC=7500 NEU 79% LYMPH 9.7% ,HB=**4.5** g/dl HCT=**13.6**% MCV=63.2 fl PLAT= 325000,BUN,Cr,Elect,Ca,Ph,Mg: Were NL,PT,PTT: Were NL,LDH: 514 IU/L, Antibody anti-LCA: negative, Antibody anti-CD-99: negative, tumor marker CA 125:neg.

The patient was empirically placed on a proton-pump inhibitor, and an upper GI endoscopy was performed the following day showed deformed bulb with bleeding ulcer treated by 1/10000 epinephrine injection and two retention clips.(Fig. 1).

CT scan of abdomen: large peritoneal mass occupying the right upper quadrant, CT scan brain, thoracic and pelvic did not show any primary tumor. (Fig.2).

Laparoscopic exploration and excisional biopsy was taken from the huge unresected abdominal mass. Pathology and immune-histochemical profile confirmed the diagnosis of ***Desmoplastic small round cell tumour of the peritoneum (DSRCT)***.

Fig 1

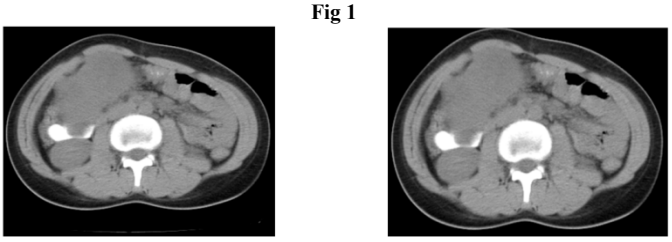

Fig 2

DISCUSSION

The DSRCT was until recently classified as a soft tissue undifferentiated sarcoma with Clusters of small round blue cells nests of unequal size and hyperchromatic nuclei with increased nuclear/cytoplasm ratio and a high level of karyokinesis surrounded by a dense fibro-sclerotic stroma. In a clinicopathologic investigation of four cases of desmoplastic small round cell tumor.(4), this tumor is characterized by a typical polyphenotypic profile with expression of epithelial, mesenchymal and Trilinear expression:

1. Epithelial marker: keratin and epithelial membrane antigen
2. Mesenchymal and myogenic markers: vimentin/desmin
3. Neuronal marker: neuron-specific enolase and CD56

The reciprocal chromosomal translocation, t(11;22)(p13;q11 or q12) that results in fusion of exon 7 of the Ewing's sarcoma gene EWS on chromosome 22 with exon 8 of the Wilms' tumor suppressor gene WT1 on chromosome 11.

EWS/WTI transcript is diagnostic of this tumor and codes for a protein that acts as a transcriptional activator that fails to suppress tumor growth.

Imaging plays a vital role in the evaluation of patients with suspected or proven peritoneal malignancy. Nevertheless, despite significant advances in imaging technology and protocols, assessment of peritoneal pathology remains challenging. The combination of complex peritoneal anatomy, an extensive surface area that may host tumour deposits and the considerable overlap of imaging appearances of various peritoneal diseases often makes interpretation difficult. Contrast-enhanced multi-detector computed tomography (MDCT) remains the most versatile tool in the imaging of peritoneal malignancy. However, conventional and emerging magnetic resonance imaging (MRI) and positron emission tomography (PET)/CT techniques offer significant advantages over MDCT in detection and surveillance(5).. CT manifestations of the DSRCT occurring in the abdomen and pelvis are variable and indistinguishable from other diseases. In addition, the most common imaging finding of patients with intra-abdominal DSRCT was multiple bulky heterogeneous peritoneal soft tissue masses without an apparent organ-based primary and mesenteric lymphadenopathy. These findings are thus characteristic of DSRCT

Diagnosis can only be made after removal of the suspicious mass and histological analysis with immunohistochemically demonstration of the trilinear nature of the tumor (6). The CT, MRI, and FDG-PET/CT imaging findings of abdominopelvic desmoplastic small round cell tumors need correlation with histopathologic findings (7,8).

Zhang et al compared different imaging modalities such as CT scan, MRI and PET scan in 7 cases of abdominopelvic DSRCT confirmed by histopathologic analysis. Results showedthat radiological findings of DSRCT include multiple masses with heterogeneous density/intensity and without an organ origin. FDG-PET/CT provided additional information on the stage of the tumor. Peritoneal seeding; peritoneal effusion, hepatic metastasis, and retroperitoneal lymphadenopathy were also found (9). No blood profile abnormalities are specific to DSRCT, tumour markers may be elevated, especially serum CA 125. This has been reported as raised in up to 86% cases of intra-abdominal DSCRT, with a median value of 200 U/ml (range 22–735) [2,3]. High CA 125 levels associated with DSRCT may be related to ascites and not directly to the tumour itself (7, 8). Therefore, tumour markers cannot be used as diagnostic tools. Importantly, CA 125 has been shown also not to be a good monitor of disease progression which relies on clinical and radiological evaluation.

The extremely rare occurrence of DSRCT means that the treatment modalities and their impact on survival has only been studied in small numbers of patients. In DSRCT there has not yet been a case in which treatment has led to a curative outcome.

A complete resection is rarely possible, but excision of large peritoneal masses have been performed in groups of patients. The complete removal of tumour deposits is usually limited by a liver metastasis, infiltration into hepatic veins or involvement of the diaphragm (9).The impact of surgical resection upon

survival remains unclear. Complete surgical resection, including 1–2 mm implants, is necessary to achieve long term disease control and is rarely possible.

The prognosis remains poor with median survival of approximately 17 months, 3-year survival rate of 29% and 5-year survival rate of 18%(10).The monoclonal antibody therapies used to target novel cell surface antigens expressed in human solid tumours are well described. In DSRCTs two antigens have been studied, G(D2) and the antigen to antibody 8H9.In a series of 46 DSRCT samples the antigen expression was 70% for G(D2) and 96% for 8H9 (11).These may represent novel approaches to diagnosis and treatment; especially with anti- G (D2), showing some efficacy in treating minimal residual disease in neuroblastoma.

The most current novel therapy is the use of continuous hyper thermic peritoneal perfusion (CHPP) therapy at 40°C with cisplatinum for 90 minutes, but outcomes are yet to be assessed

Yttrium microspheres has been used successfully to treat liver metastasis from DSRCT is a novel treatment for liver metastasis in DSRCT

Heterogeneity of the data and lack of staging criteria, together with small numbers of patients in single-center series mean comparison of all the techniques is difficult. This coupled with lack of multi-center trials and locally prescribed chemotherapeutic protocols means, the true outcome is difficult to measure. In addition, DSRCT spans the pediatric/adult divide of medicine, thus prohibiting larger numbers of patients been reviewed in centers and there may be a true difference in outcome or response to treatment modalities in differing age groups (11).

A high degree of suspicion, a thorough physical examination, a full imaging check and an aggressive therapeutic approach are required in order to identify this disease and fight for a better quality of life for these young patients. Future genetic therapies focused on developing targeted immunotherapy, might promise more optimistic results. Until then more than one medical specialties should collaborate in order to face the challenge and treat such patients.

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