

Desmoplastic Small Round Cell Tumour: A Rare Entity

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Abstract: *Desmoplastic small round cell tumour (DSRCT) is a rare and highly aggressive variety of sarcoma arising typically from abdominal or pelvic peritoneum. Diagnosis and treatment approaches of this entity are complex and require a skilled, experienced, multidisciplinary team. Authors report two cases:- 1) A 54 years old female presenting with complaints of abdominal pain and distension, in whom computed tomography was suggestive of Mass lesion in peritoneal cavity with liver metastasis and peritoneal deposits which was proven on histopathology as DSRCT. 2) A 22 years old male presenting with complaints of lump and severe pain in abdomen, in whom computed tomography was suggestive of Mass lesion in mesentery with extension into peritoneal cavity which was proven on histopathology as DSRCT.*

Keywords: Desmoplastic small round cell tumour (DSRCT), CT

1. Introduction

Desmoplastic small round cell tumour (DSRCT) is a rare and highly aggressive sarcoma occurring mainly in children and young adults. The first case was described in 1989 by Gerald and Rosai. A specific chromosomal translocation t(11; 22)(p13; q12) involving two genes: EWS which is characteristic of Ewing's sarcoma and WT1 which is a Wilms tumour gene, has been documented in DSRCT. This tumour has predilection for serosal surfaces such as peritoneum. Diagnosis can be made by radiological and histological features. No curative treatment has been yet documented.

2. Case Report:

Case 1

A 54-year-old female presented with a 3 months history of abdominal distention with discomfort and pain, and 20 Kg weight loss. Physical examination revealed an 8 cm sized large abdominal mass arising from right lumbar and right iliac regions. Patient's vital signs and laboratory data were normal.

Computed tomography (CT) of the abdomen showed approximately (10.3x8.7x8.5) cm³ size well defined heterogeneously enhancing soft tissue density lesion noted involving right lumbar and right iliac region along the anti-mesenteric border adjacent to caecum and ascending colon (p/o mass lesion). Lesion reaches posteriorly upto IVC and slightly compresses it and anteriorly reaches upto anterior abdominal wall. Also there were liver metastasis and peritoneal deposits in perihepatic area, in mesentery in left lumbar region and posterior to urinary bladder adjacent to sigmoid colon.

Trucut biopsy from mass showed Focal monomorphic small cell arranged in diffusely and perivascular manner. Cells are small in size with round uniform nuclei, coarse chromatin, indistinct nucleoli and moderate amount of

clear/eosinophilic cytoplasm. All findings were suggestive of desmoplastic small round cell tumour.

Case 2

A 22 years old male presented with 2 months history of lump and severe pain in abdomen. Physical examination revealed abdominal lump in hypogastric region. Patient's vital signs and laboratory data were normal.

Computed tomography (CT) of the abdomen showed diffuse plaque like heterogeneously enhancing nodular soft tissue thickening noted in mesentery extending along with the adjacent bowel loops in peritoneal cavity encases mesenteric vessels, celiac trunk, superior and inferior mesenteric artery with homogeneously enhancing perivesical and perirectal wall thickening noted (p/o mass lesion).

CT guided biopsy from mesenteric nodules – Tumour cells are seen in small lobules /nodules, are monomorphic with hyperchromatic nuclei and notable mitosis. All findings were suggestive of desmoplastic small round cell tumour.

3. Discussion

Diagnosis of intra-abdominal DSRCT is difficult and it is usually obtained at an advanced stage. It may be first suggested by the imaging findings which are useful also for staging and for a guided biopsy. However, only histological analysis led to the confirmation of diagnosis. In fact, the clinical manifestations are usually non-specific; typically, the tumour produces few symptoms until it is large enough to compress or invade surrounding structures. Symptoms include abdominal discomfort/ distension, abdominal pain, weight loss, or change in bowel habits.

Radiologically, the most characteristic imaging feature of DSRCT of the abdomen is single or multiple, lobulated peritoneal soft tissue masses without an apparent organ of origin. These abnormalities may be associated with ascites, lymph nodes, diffuse nodular peritoneal thickening or with distant metastasis. CT imaging is a useful modality for

evaluating the size and extension of desmoids and the involvement of adjacent structures before surgical resection. It is also valuable in evaluating tumour recurrence. Differential diagnosis of DSRCT are Peritoneal carcinomatosis, Non-Hodgkin's lymphoma and malignant peritoneal mesothelioma.

Regarding the aggressiveness of the disease, treatment is based on multi-modal therapy including an extensive surgery when it is feasible, systemic chemotherapy with or without abdominal radiotherapy.

It has been reported that the combination of three modalities has shown best results leading to an overall response rate of 39% and a 3-year survival rate of approximately 50% as compared to each modality used separately.

4. Conclusion:

DSRCT is a rare aggressive neoplasm characterized by a distinct radiological appearances, histological appearances and a unique cytogenetic profile, the radiologists should be familiar with this entity and help prolong survival.

5. Abbreviations

CT: Computed tomography

References

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CASE 1

CT scan images showing mass lesion (figure 1) with liver metastasis (figure 2) and peritoneal deposits (figure 3) with histological appearance of DSRCT (figure 4)

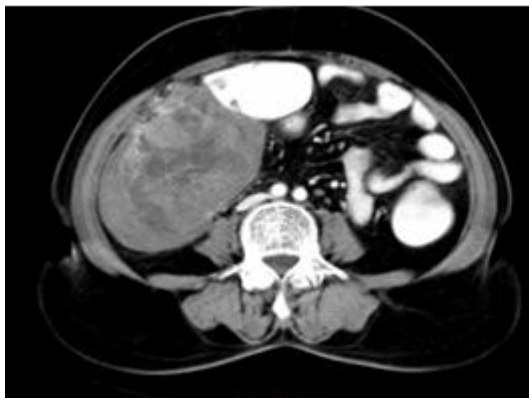


Figure 1



Figure 2

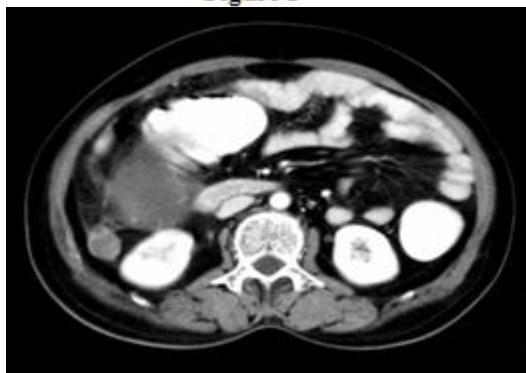


Figure 3

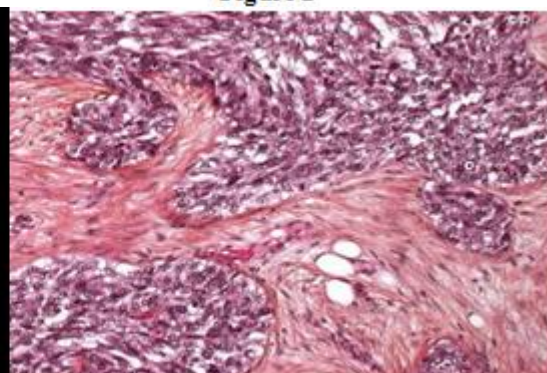


Figure 4

Case 2

CT scan images showing mass lesion in mesentery (figure 1 and 2) with histological appearance of DSRCT (figure 3).

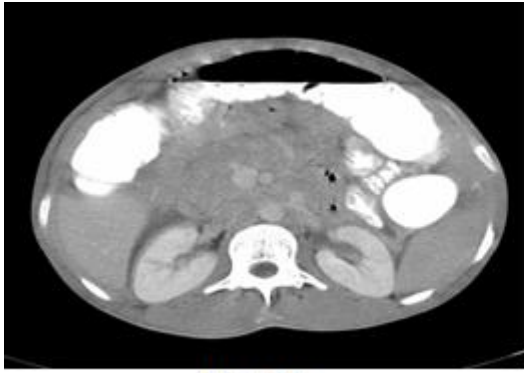


Figure 1



Figure 2

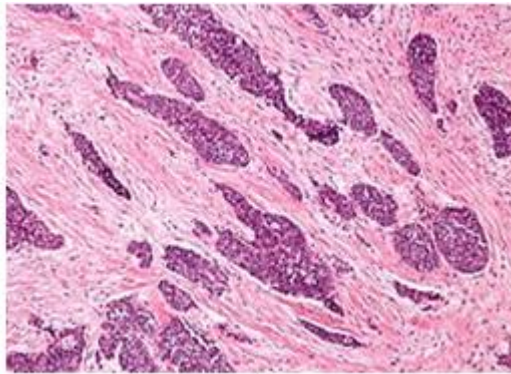


Figure 3