

Intra-abdominal desmoplastic small round cell tumor

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A 30-year-old Syrian male presented with complaints of dull aching abdominal pain for 2 weeks associated with progressive abdominal distension. On examination he had abdominal shifting dullness. Routine investigations were normal, ultrasound scan revealed moderate ascites. Ascitic tap showed viscous straw-colored fluid with high protein (4.2 g/dL) and albumin (3.9 g/dL) levels, low serum ascites albumin gradient (SAAG) (1.0 g/dL) and high total leukocyte count (8950/mm³, predominantly lymphocytic). Evaluation for tuberculosis and tumor markers (carcinoembryonic antigen, CA19-9, α -fetoprotein) were non-contributory. His ascitic fluid cytology showed abnormal cells suspicious of malignancy.

Computed tomography (CT) of abdomen showed ascites, omental cakes and retroperitoneal lymph nodes (hypodense nodes measuring up to 3.0 cm) (Fig. 1). CT-guided biopsy of retroperitoneal lymph nodes and omental cakes showed dense infiltration of small round blue cells (Fig. 2) in clusters and stained positive for desmin, AE1/AE3 and EMA with Ki-67 of 60%. A diagnosis of a rare neoplasm, intra-abdominal desmoplastic small round cell tumor (IDSRCT) was established. With diffuse intra-abdominal spread and progressive disease, he was started on IE/VAC (ifosfamide, etoposide)/(vincristine, doxorubicin and cyclophosphamide) chemotherapy (alternating cycles three weekly). He received 4 cycles of chemotherapy with initial clinical and radiological response. Unfortunately, the patient started to deteriorate thereafter with symptomatic right pleural effusion. A repeat positron emission tomography scan showed right-sided pleural effusion, reappearance of ascites, peritoneal caking and increase in size of mesenteric and retroperitoneal lymph nodes. He was started on second-line chemotherapy with docetaxel and gemcitabine. However, he developed refractory sepsis and multiple organ dysfunction after the 1st cycle of second line chemotherapy and succumbed to illness.

IDSRCT is a rare neoplasm with mean age of diagnosis 22 years (range 6-49), and male to female ratio 4:1 [1]. The tumor typically develops in the abdominal cavity, invading the omentum with multiple nodular deposits in peritoneum, diaphragm, splenic hilum, mesentery of small and large bowel. Clinically, patients remain asymptomatic for a variable period and then present with symptoms of abdominal pain and/or distension, constipation, vomiting, and weight loss. Ultrasound, CT scan or magnetic resonance imaging demonstrate lesions of varying size (from millimeter sized lesions to large confluent nodules). Histopathologically it is characterized by small round cells that have a positive reaction to immunostaining for epithelium, myogenic cells and neurogenic cells. Chromosomal translocation t11,22 is identified in this tumor. In most cases, they present with metastatic disease, resection is incomplete

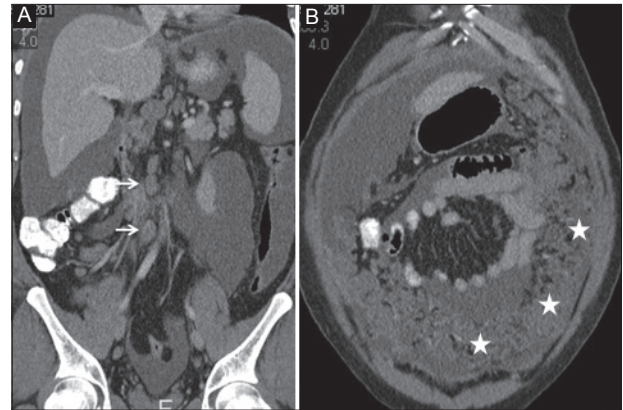


Figure 1 Computed tomography (CT) scan of abdomen. Coronal CT contrast images (A, B) show massive ascites, diffuse soft tissue thickening of omental (star), and retroperitoneal and mesenteric lymphadenopathy (white arrow)

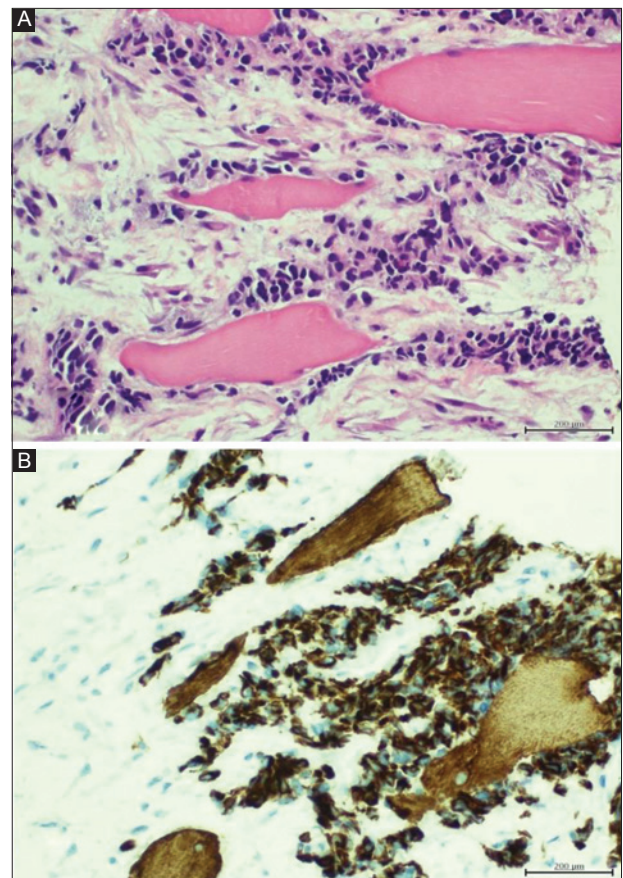


Figure 2 Immunohistopathological appearance of omental deposits. (A) Microscopic examination revealed undifferentiated small cell tumor dissecting between muscle bundles (Hematoxylin and Eosin \times 400). (B) Immunohistochemical stain using antibody against Desmin shows characteristic perinuclear staining of the tumor cells (Desmin \times 400)

and chemotherapy is only temporarily effective, median survival ranging between 17-25 months [1]. Other small round cell tumors include embryonal rhabdomyosarcoma, small cell carcinoma, mesothelioma and Ewing's sarcoma.

Despite polychemotherapy, whole abdominal radiation and debulking surgery results are mostly suboptimal. Hyperthermic intraperitoneal chemotherapy after tumor debulking, postoperative intensity modulated radiotherapy, treatment of hepatic metastases with [90Y] Yttrium microsphere liver embolization have been tried in adjuvant settings [2]. There are rare reports of IDSRCT and only one report of the same presenting with ascites [3]. We wish to emphasize that while evaluating low SAAG ascites, rare malignancies like IDSRCT need to be considered.

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Conflict of Interest: None

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