Intra-abdominal desmoplastic small round cell tumor
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Abstract
Intra-abdominal desmoplastic small round cell tumor (IDSRCT) is a unique, highly aggressive neoplasm that chiefly affects male adolescents and young adults and most frequently presents as a large abdominal mass with widespread peritoneal involvement at the time of diagnosis. We present two cases of IDSRCT in a young male and a female. Both typically presented with diffuse peritoneal involvement.

Introduction
Intra-abdominal desmoplastic small round cell tumor (IDRSCT) is an uncommon neoplastic condition that predominantly occurs in young adult men and usually diffusely involves the abdominal and/or pelvic peritoneum. IDSCRT has distinctive clinical, histologic and immunophenotypic features. Histologically it is typically composed of nests of small, undifferentiated round or oval hyperchromatic cells with abundant desmoplastic stroma. Immunohistochemically it is reactive for epithelial markers; keratin, epithelial membrane antigen (EMA), neural (neuron-specific enolase; NSE) and muscle markers (desmin). Molecular studies have identified a translocation t (11; 22) (p13; q12) as being unique to IDSRCT. We describe two cases of IDSRCT in a young male and a female.

Case Report

Case No. 1
A 32 year old well built male presented with complaints of pain in the left upper quadrant, weight loss and constipation for 6 months. He also noticed a mobile mass in the left upper quadrant. There was no significant past medical history and he did not have any family history of cancer. On examination there was a non tender mobile mass palpable in the left upper quadrant of the abdomen. On ultrasound examination of the abdomen, multiple deposits in the liver and spleen were observed. A barium enema revealed extrinsic compression of the colon near the splenic flexure due to a mass and a CT scan confirmed the above findings. A colonoscopic as well as needle biopsy of the mass was done which at that time revealed an undifferentiated neoplasm. He was seen in the outpatient clinic and was referred by the gastroenterologist for a surgical opinion considering this as colon cancer. A diagnostic laparoscopy was performed, multiple peritoneal deposits of tumour were found one of which was taken for biopsy. Histopathology of the specimen showed a neoplastic lesion composed of clusters of small tumor cells with scanty cytoplasm. Nuclei were of small size and pleomorphic with inconspicuous nucleoli. Marked desmoplastic response was identified around the tumor cell clusters (Figure 1). No glycogen positivity was seen on special stains. Immunohistochemical studies showed strong Cytokeratin AE1/AE3 (Figure 2), Cytokeratin Cam 5.2, CK 7, epithelial membrane antigen positivity. Globoid dot positivity of Desmin was seen along
with focal positivity of Vimentin and Neuron specific enolase. The tumour cells were negative for S-100, Mic-2, ASMA, Chromogranin and synaptophysin. Based on morphological and immunohistochemical features, a diagnosis of Intra abdominal desmoplastic small round cell tumor was made.

**Case No. 2**

A 23 year old female presented with the complaint of constipation and abdominal pain for the last two months. On examination masses were palpable in right and left pelvic regions. Per-vaginal examination showed hard irregular masses in the pouch of Douglas. Laparotomy was done; per-operatively hard irregular nodules were present in the pouch of Douglas alongwith bilateral ovarian masses. No para-aortic lymphadenopathy was present. Bowel and mesentry were normal. Total abdominal hysterectomy and bilateral salpingo-oopherectomy with omentectomy was done along with removal of masses from pouch of Douglas. Grossly right ovary was 9 x 5 cms and left ovary was 7 x 6 cm in size. Cut surface showed entire ovary replaced by grey white firm lesion. Uterus and cervix were grossly unremarkable. Multiple sections were taken from the right and left ovaries, omentum and masses from pouch of Douglas. Histological and immunohistochemical findings were consistent with intra abdominal desmoplastic small round cell tumor.

**Discussion**

IDSRCT usually involves the abdominal and/or pelvic peritoneum but extra-abdominal location has also been described. Cases are reported in the central nervous system, bone, extremity, kidney, salivary gland, paratesticular region and pleura.\(^1\) The histogenesis is uncertain.

Common clinical presentations are pain, abdominal distension and a palpable abdominal, pelvic and scrotal mass, sometimes with associated ascites. Histologically uniformly closely packed small cells are distributed in a background of desmoplastic stroma. Differential diagnosis includes lymphoma, Ewing sarcoma/PNET, neuroblastoma, alveolar rhabdomyosarcoma, malignant mesothelioma. Cytologically, the tumor cells consist of small, round to oval cells with a scant amount of light blue cytoplasm. The tumour cells were negative for S-100, Mic-2, ASMA, Chromogranin and synaptophysin. Based on morphological and immunohistochemical features, a diagnosis of Intra abdominal desmoplastic small round cell tumor was made.

**References**

3. Ordonez NG, Sahin AA. CA 125 production in desmoplastic small round cell...


