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DESMOPLASTIC SMALL ROUND CELL TUMOR: A CASE REPORT

General Surgery

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ABSTRACT

Desmoplastic small round cell tumor is a rare, highly malignant neoplasm originating from mesenchymal tissue which was initially described in 1991 by Gerald and Rosai.[1] It is composed of small round tumor cells of uncertain histogenesis associated with prominent stromal desmoplasia and polyphenotypic differentiation. It typically occurs in adolescents and young adults.[2] Usually presents with widespread abdominal, serosal, and mesenteric involvement with poor prognosis.

KEYWORDS

malignancy, abdominal mass, round cell tumor, desmoplastic

Case report

Here we present a case of DSRT who was managed surgically and by chemotherapy.

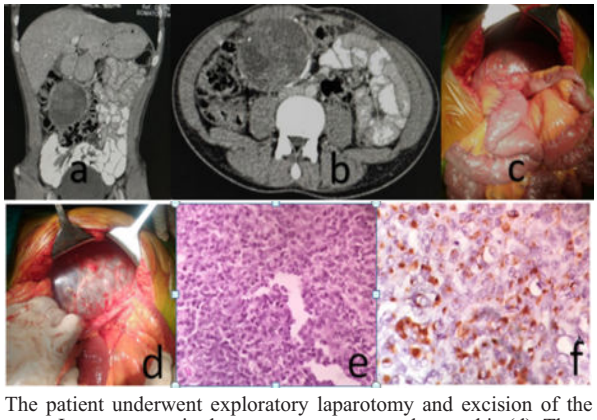
A 17yr old male presented with complaints of upper abdominal pain and palpable abdominal mass of 5x5 cm in the epigastric region of 1-year duration; the pain was dull aching and continuous which was relieved on taking analgesics. It was not associated with any nausea, vomiting, fever, or decreased appetite. He had no bladder or bowel-related complaints. There was no history of trauma or anti-tuberculosis treatment intake in past. A 5x5 cm lump was present in the epigastric region which was well defined, mobile, and non-tender. The rest of the abdomen and genitalia was grossly normal. There were multiple lymph nodes palpable in bilateral cervical and axillary and inguinal regions.

Hemogram was normal (Hb 10.8g/dl, TLC- 10200 cumm, DLC-61/28/1/9)

FNAC from cervical and inguinal lymph nodes showed reactive hyperplasia.

USG abdomen showed a heterogeneous mass lesion of size 7*6.7 cm, solid-cystic with multiple hypoechoic areas seen in retroperitoneum compressing IVC, with mild internal vascularity.

A Chest x-ray was reported normal; CECT abdomen showed lobulated well defined heterogeneous enhancing intraperitoneal mass 8.6x6.7x5.9cm in size, showing calcification along the periphery with multiple non-enhancing areas within(a,b). It was abutting the transverse colon and small bowel(c). A provisional diagnosis of GIST or lymph node mass was made. A hypoattenuating lesion in the left lobe of the liver was also present. Multiple sub-centimetric lymph nodes were present in the mesentery.



The patient underwent exploratory laparotomy and excision of the mass. It was present in the greater omentum and was white(d). There were multiple small deposits in the liver from which biopsy was taken. Multiple lymph nodes were present in the mesentery, but the surrounding bowel was grossly normal.

Histopathology of the resected specimen showed a solid cystic mass showing tumor comprising of round to ovoid cells in nests and trabeculae and focally as sheets within a desmoplastic stroma with focal cystic areas(e). Tumor cells were positive for Cytokeratin, Vimentin(f), and dot-like perinuclear positivity for desmin. LCA, S100, and CD99 were negative. These findings were suggestive of a Desmoplastic small cell tumor.

Liver nodules also showed tumor deposits. Resected margins were clear.

The patient had a normal postoperative course and was referred for chemotherapy and is on follow-up. he received 7 courses of chemotherapy with cyclophosphamide (4200 mg/m²), doxorubicin (75 mg/m²), and vincristine alternating with ifosfamide (9 to 12 mg/m²) and etoposide (500 to 1000 mg/m²).

DISCUSSION

Desmoplastic small round cell tumor is a rare aggressive malignancy classified as soft tissue sarcoma sharing some of its features with Ewing's sarcoma and primitive neuroectodermal tumor (PNET) which share the same Ewing sarcoma (EWS) fusion protein.[3]It is characterized by unique chromosomal translocation t (11; 22) (p13; q12) and fusion protein EWSR1-WT1.[4]The tumor grows aggressively and can metastasize to lungs and liver. The patient can also present with a multifocal disease with tumors of the peritoneal cavity.[5]it usually presents as abdominal pain predominantly affecting boys and young men.[6]

In the histopathological examination of Desmoplastic small round cell tumor; desmin, Cytokeratin, and vimentin are positive, of which desmin and CK being positive at the same time is considered DSRCT's most specific immunological indexes. Stromal elements are vimentin-positive, prompt from the muscle fiber mother cell. However, this tumor can express epithelial, mesenchymal, neuroendocrine, and other immunophenotypes and is cytogenetically specific. Besides, cells can generate the EWS-WT1 fusion gene.[7]

Neuroblastoma, rhabdomyosarcoma, and peritoneal carcinomatosis in young and lymphoma in older patients present with similar complaints, so these differentials need to be kept when evaluating the patient.

The prognosis is poor with 3-year survival ranging between 30-50% in those treated with complete cytoreductive surgery, chemotherapy, and radiation therapy. Lal et al reported the 3-year overall survival after this multimodality therapy to be 55% compared with 27% if anyone was missed.[3].Targeted therapies to the specifically identified mutation are under trial. Anti –ILG R1, temsirolimus combined with cixutumumab have reported promising results in desmoplastic round cell tumor DSRCT is commonly fatal with a median survival of 2.5 years despite new advances in chemotherapy and stem cell transplantation.[8]

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