Desmoplastic Small Round Cell Tumor (DSRCT): an Unusual Intra-abdominal Tumor

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Abstract Desmoplastic small round cell tumor (DSRCT) is a rare intra-abdominal tumor commonly seen in adolescents and young adult males. It is an important differential diagnosis in these patients presenting with abdominal masses and/or GI obstruction. The management and prognosis improve if pre-operative diagnosis can be established.

Keywords Desmoplastic · Round cell tumor · Intra-abdominal tumor

Desmoplastic small round cell tumor was first reported in 1989. It is considered to be an aggressive malignancy that mainly occurs in the abdominal cavity [1]. Most patients have widespread disease at presentation, with the organ of origin difficult to ascertain. It is a malignancy with distinctive histological and immunohistochemical features [2].

Case Report

A 21-year-old male presented to us with a history of incidentally noticed mass in his right lower abdomen since about 3 months. This had gradually increased in size and was

associated with dragging type of pain since a couple of weeks. He has no history of bowel or bladder disturbances, no constitutional symptoms, and no preceding trauma. Past and family histories were not contributory.

On examination, the vitals were within normal limits. There was no pallor, edema, icterus, or lymphadenopathy. Abdominal examination revealed a 6×6-cm, nontender, hard, nodular intra-peritoneal mass in right iliac fossa (RIF) with no other pathology. Per rectal and other systemic examinations were normal.

Investigations revealed normal hematological parameters. Ultrasonography of the abdomen revealed an 8×6-cm ill-defined mass in RIF with variable echogenicity. Colonoscopy showed normal lumen and mucosal pattern of the colon and terminal ileum with no signs of external compression.

CT scan of the abdomen and pelvis showed mass lesion involving the terminal ileum and cecum, with involvement of the adjacent mesentery, with hypo- and hyper-echoic areas. Metastatic lesions are showed in the liver (as shown in Fig. 1).

In view of the above findings being nonspecific and with a possibility of a nonepithelial neoplasm, a Trucut biopsy was carried out under CT guidance. Histopathological report revealed a desmoplastic small round cell tumor (DSRCT).

Immunohistochemical (IHC) test showed that these cells have trilinear co-expression, including the epithelial marker cytokeratin, the mesenchymal markers desmin (as shown in Fig. 2) and vimentin, and the neuronal marker neuron-specific enolase. Common leucocyte antigen (CLA) was absent, thus differentiating it from lymphoma.

The final diagnosis made was stage IV DSRCT.

Treatment

A tumor board meeting was carried out after reviewing the literature on the disease, and a plan of treatment was decided as follows: induction chemotherapy → cytoreductive surgery → adjuvant chemotherapy +/- radiotherapy.

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Fig. 1 Mass lesion involving the terminal ileum and cecum, mesenteric involvement, areas of variable echogenicity, and liver metastasis

The patient was given induction chemotherapy: two cycles of vincristine of 2 mg, doxorubicin of 80 mg, and cyclophosphamide of 1,600 mg (VAC) over 2 months. The patient had partial response, with the RIF tumor reducing in size, but not the liver lesions.

He underwent laparotomy after chemotherapy, and findings on the table were as follows: large mass in the right iliac fossa involving the terminal ileal and cecal walls with diffuse peritoneal metastasis with intraparenchymal liver lesions.

The procedure was as follows: right ureteric DJ stent + right hemicolectomy (including the lesion) + total omentectomy + bilateral paracolic and pelvic peritonectomy + right diaphragmatic peritonectomy. Liver lesions were not attended as they were deep parenchymal lesions.

Procedure and post-op recovery were uneventful. Biopsy report revealed DSRCT with diffuse peritoneal lining metastasis.

After 8 weeks, he was given adjuvant chemotherapy: alternating three cycles each of ifosfamide of 7 g and etoposide of 200 mg (IE) and VAC.

At 1-year follow-up, the patient is asymptomatic, and repeat CT scan showed no evidence of intra-abdominal recurrence; however, the liver lesions persisted with the same size and number as in the previous scan. FNAC of liver lesions showed DSRCT.

The patient and his family were explained on the nature of persistent disease and associated prognosis. The choice between metastatectomy or radiofrequency ablative therapy for the liver lesions was given to them.

The patient and his family requested for time to decide. Risks involved in delaying the suggested therapy were explained to them.

Discussion

The disease is associated with unique chromosomal translocation $t(11;22)(p13;q12)$ resulting in EWS/WT1 transcript that is diagnostic of this tumor [2].

Fig. 2 IHC of tissue which shows positive staining for cytokeratin and desmin

DSRCT was first described in 1989 by Gerald and Rosai [3]. It exhibits male predominance of 90 %. Median age at diagnosis has been reported as 14, 19, and 25 years of age in different series. Eighty-five percent of patients are Caucasian [3–6].

DSRCT, a disease of the young, commonly affects the lining of the abdomen, diaphragm, spleen, liver, chest wall, skull and spinal cord, and pelvis and its organs [7]. Common presentations are loss of appetite, mass and/or distension of the abdomen, abdominal pain, GI obstruction, and symptoms related to specific organs when they are involved.

Histology of specimen will reveal well-circumscribed solid tumor nodules within a dense stroma, with areas of central necrosis. IHC shows trilinear co-expression for epithelial, mesenchymal, and neuronal markers [2].

There is no standard treatment protocol, and studies suggest multimodality approach [7–9]. Other treatment options under evaluation were hematopoietic stem cell transplantation, intensity-modulated radiotherapy, and radiofrequency ablation [10].

Radiofrequency ablation (RFA) is especially useful for patients who are not ideal surgical candidates, who have recurrent/residual tumors, or who do not benefit from conventional therapies. RFA can be effective if the tumors are small and few [11].

Overall, 1-, 3-, and 5-year survival rates are about 52.36, 27.92, and 27.92 %, respectively [1].

Conclusion

- DSRCT is an important D/D in adolescents and young adults with abdominal masses.
- Management and prognosis improve if preoperative diagnosis is established.

- Multimodality treatment involving chemotherapy + aggressive cytoreductive surgery + radiotherapy is ideal to improve prognosis and quality of life.

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