Asian Oncology Research Journal

5(1): 7-10, 2022; Article no.AORJ.62254

Desmoplastic Small Round Cell Tumor: A Case Report

Bhuwan Kumar [a*]

[a] *Department of General Surgery, MAMC, New Delhi, India.*

Author's contribution

The sole author designed, analysed, interpreted and prepared the manuscript.

Article Information

Open Peer Review History:
This journal follows the Advanced Open Peer Review policy. Identity of the Reviewers, Editor(s) and additional Reviewers, peer review comments, different versions of the manuscript, comments of the editors, etc are available here: https://www.sdiarticle5.com/review-history/62254

Case Study

Received 01 September 2020
Accepted 04 November 2020
Published 15 January 2022

ABSTRACT

Desmoplastic small round cell tumour is a very rare, highly malignant abdominal neoplasm of mesenchymal origin. Its non-specific symptoms at presentation and poor prognosis with a five year survival rate of 15% despite treatment make it a big challenge to manage. We wish to discuss a case of a young 17-year old male with abdominal pain and a palpable lump which was eventually diagnosed with desmoplastic round cell tumour.

Keywords: Desmoplastic small round cell tumor; prognosis; survival rate; polyphenotypic differentiation.

1. INTRODUCTION

Desmoplastic small round cell tumour 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 adolescent and young adults [2]. Usually presents with widespread abdominal, serosal and mesenteric involvement with poor prognosis.

2. CASE PRESENTATION

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

**Corresponding author: E-mail: bhuwankan@yahoo.co.in, drbhuwan.uro@yahoo.com;*

A 17yr old male presented with complaints of upper abdominal pain and palpable abdominal mass of 5x5 cm in epigastric region of 1 year duration; 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 epigastric region which was well defined, mobile and non-tender. Rest of 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 heterogeneous mass lesion of size 7*6.7 cm, solid-cystic with multiple hypo echoic areas seen in retro peritoneum compressing IVC, with mild internal vascularity.
A Chest x-ray was reported normal; CECT abdomen showed lobulated well defined heterogenous enhancing intraperitoneal mass 8.6x6.7x5.9cm in size, showing calcification along the periphery with multiple non enhancing area within. It was abutting transverse colon and small bowel. A provisional diagnosis of GIST or lymphnode mass was made. A hypo attenuating lesion in left lobe of liver was also present. Multiple subcentrimetric lymph nodes were present in the mesentry.

Patient underwent exploratory laparotomy and excision of mass. It was present in greater omentum and was white in colour. There were multiple small deposits in the liver from which biopsy was taken. Multiple lymph nodes were present in the mesentry, but surrounding bowel was grossly normal.

Histopathology of 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. Tumor cells were positive for Cytokeratin, Vimentin and dot like perinuclear positivity for desmin. LCA, S100 and CD99 were negative. These finding were suggestive of Desmoplastic small cell tumour.

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

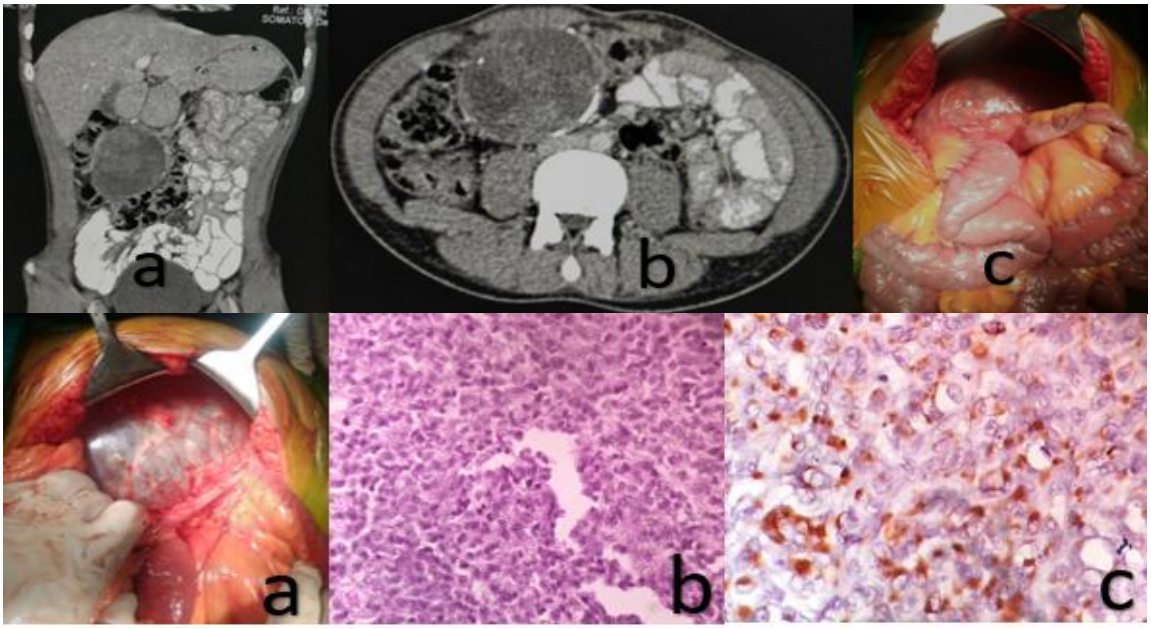

Fig. 1. Histopathology of resected specimen

Patient had normal postoperative course and was referred for chemotherapy and is on followup.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) .

3. DISCUSSION AND CONCLUSION

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 tumour (PNET) which share 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 tumour grows aggressively and can metastasize to lungs and liver. Patient can also present with multifocal disease with tumours of peritoneal cavity [5]. it usually presents as abdominal pain predominantly affecting boys and young men [6].

In 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. In addition, 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 specific identified mutation are under trial.Anti –ILG R1, temsirolimus combined with cixutumumab have reported promising results in desmoplastic round cell tumour. DSRCT is commonly fatal with a median survival of 2.5 years in spite of new advances in chemotherapy and stem cell transplantation [8].

CONSENT AND ETHICAL APPROVAL

As per university standard guideline participant consent and ethical approval has been collected and preserved by the authors.

COMPETING INTERESTS

Author has declared that no competing interests exist.

REFRENCES

1. Gerald WL, Rosai J: Case 2. Desmoplastic small cell tumor with divergent differentiation. Pediatr Pathol. 1989;9:177–183
2. Kretschmar CS, Colbach C, Bhan I, Crombleholme TM. Desmoplastic small cell tumor: A report of three cases and a review of the literature. J Pediatr Hematol Oncol. 1996;18:293-298.
3. Lal DR, Su WT, Wolden SL, Loh KC, Modak S, La Quaglia MP. Results of multimodal treatment for desmoplastic small round cell tumors. Journal of Pediatric Surgery. 2005;40(1):251-5
4. Lae ME, Roche PC, Jin L, Lloyd RV, Nascimento AG. Desmoplastic small round cell tumor: a clinicopathologic, immunohistochemical, and molecular study of 32 tumors. The American Journal of Surgical Pathology. 2002; 26(7):823-35.
5. Hayes-Jordan A, Anderson PM. The diagnosis and management of desmoplastic small round cell tumor: A review. Current Opinion in Oncology. 2011;23(4):385-9.
6. Li G, Wang HT, Gao Y, Cui XJ, Zhang GZ. Primary abdominopelvic desmoplastic small round cell tumor: CT and correlated clinicopathologic features. Eur Rev Med Pharmacol Sci. 2014;18(18): 2670-7.
7. Yang JL, Xu WP, Wang J, Zhu XZ, Zhang RY. Desmoplastic small round cell tumor: a clinicopathologic study of 15 cases. Zhonghua bing li xue za zhi Chinese Journal of Pathology. 2005; 34(10):650-5.

8. Gil A, Gomez Portilla, Brun EA et al. Clinical perspective on desmoplastic small round-cell tumor Oncology. 2004;67:231-242.

Peer-review history:
The peer review history for this paper can be accessed here:
https://www.sdiarticle5.com/review-history/62254