Original Research Paper

Volume-8 | Issue-3 | March-2018 | PRINT ISSN No 2249-555X

Oncology

DESMOPLATIC SMALL ROUND CELL TUMOR OF PARATESTICULAR ORIGIN

Deep Shankar Pruthi	Senior Resident, Department of Radiotherapy, Cancer Research Institute, SRHU, Dehradun, Uttarakhand
Saurabh Bansal*	Assistant Professor, Department of Radiotherapy, Cancer Research Institute, SRHU, Dehradun, Uttarakhand *Corresponding Author
Neena Chauhan	Professor, Department of Pathology, Himalayan Institute of Medical Sciences, SRHU, Dehradun, Uttarakhand
Meenu Gupta	Professor, Department of Radiotherapy, Cancer Research Institute, SRHU, Dehradun, Uttarakhand,
Vipul Nautiyal	Assistant Professor, Department of Radiotherapy, Cancer Research Institute, SRHU, Dehradun, Uttarakhand
Mushtaq Ahmad	Professor, Department of Radiotherapy, Cancer Research Institute, SRHU, Dehradun, Uttarakhand
Sunil Saini	Professor, Department of Surgical oncology, Cancer Research Institute, SRHU, Dehradun, Uttarakhand

ABSTRACT Desmoplastic Small Round Cell Tumor (DSRCT) of the paratesticular region is an extremely rare mesenchymal tumor occurring in adolescence with a tendency for extensive metastases. We report a rare case of DSRCT of paratesticular origin in a 24 year old male who presented with painless right testicular mass and disseminated abdominal disease. The patient underwent right inguinal orchidectomy and on Histopathology and Immunohistochemistry the diagnosis was confirmed. The patient was treated with multi agent chemotherapy with partial remission. Prognosis of such patients is generally dismal despite multimodality treatment.

KEYWORDS : Desmoplastic Small Round Cell Tumor, Paratesticular, Immunohistochemistry

Background

Desmoplastic small round cell tumor (DSRCT) was first reported by Gerald and Rosai in 1989 (1). It is an extremely rare malignancy with undetermined histogenesis. They are thought to be derived from multipotential differentiated primitive mesenchymal cells or neuroectodermal cells (2). It is prone to develop in adolescents and young adults with male : female ratio being 3 : 1. Most frequently it originates from the abdominal cavity and pelvis. Invasion of extraperitoneal sites like CNS, lung, bones, kidneys, paratesticular region, ovaries have been documented in literature (3). It has a poor 5 year survival rate.

Herein we report a case of a young male with a testicular neoplasm that was pathologically confirmed to be a paratesticular desmoplastic small round cell tumor.

Case Presentation

Clinical History

A 24 year old male patient presented in our hospital with the complaints of painless right testicular swelling and abdominal distension since the last 5 months. On Examination the patient had a right sided testicular mass of 4 x 4 cm sizewith gross ascites. The patient also had multiple right sided inguinal lymphadenopathy with largest size of 4 x 2 cm and there were multiple nodules palpable in the left iliac fossa.

The hemogram and the serum biochemistry were within normal limits. Investigative work up showed LDH levels to be 353U/L whereas other tumor markers AFP and Beta HCG were within normal limits.

Ultrasonography of the right scrotum revealed conglomerate lobulated mass of 3.3 x 2.2cm size posterior to right epididymis with raised vascularity with enlarged lymph nodes in right inguinal region 4.6 x 2.7cm. CECT of the abdomen and pelvis showed Large masses in right hepatorenal pouch and in rectovesical pouch along with multiple discrete lymph nodes in pre and para aortic region, peripancreatic and mesentry. There was presence of multiple peritoneal deposits with Ascites. Chest X Ray was normal.

Pathological Findings

The patient underwent FNAC from left iliac nodule and right testicular mass which showed features of small round cell malignant tumor. Hence a preoperative diagnosis of right testicular neoplasm with widespread abdominal dissemination was made.

The patient underwent right high inguinal orchidectomy. On histo pathological examination the testes was replaced by greyish white, homogenous tumor which has ruptured and is infiltrating the tunica. On microscopy there was presence of tumor arising from paratesticular region, Tumorwas forming solid sheets and nests which were separated by thick or thinfibrous stroma. The cells were small round cells having scanty eosinophiliccytoplasm. Mitotic figures were frequent giving the impression of small round cell tumor arising from paratesticular tissues. On Immunohistochemistry the tumor cells were positive for cytokeratin PAN, EMA, Vimentin, NSE and Desmin hence confirming the diagnosis of Desmoplastic Small Round Cell Tumor.

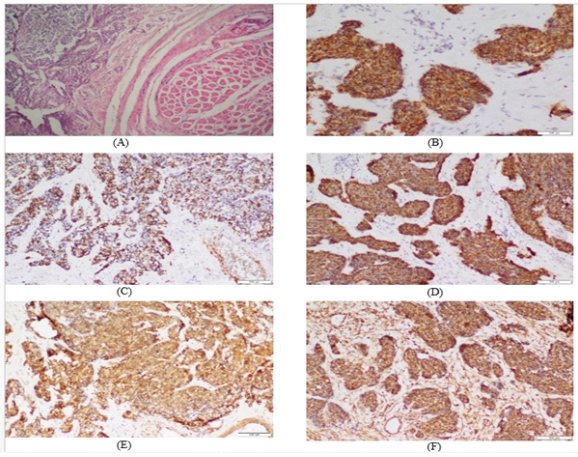

Fig 1: (A) H & E section showing desmoplastic small round cell tumor in paratesticular region. Tumor cells show positivity on immunohistochemistry for (B) CK PAN (C) Desmin (D) EMA (E) NSE (F) Vimentin.

The patient received 6 cycles of chemotherapy using alternate VAC/IE protocol. The patient had good response to chemotherapy both subjective and objectively. However the patient defaulted for 5 months and again presented with pain abdomen. On Examination he had small abdominal nodules palpable on the right side of abdomen with raised LDH Levels (459 U/L). The patient was again started on chemotherapy with the same regimen and received 6 more cycles of chemotherapy. The patient had stable disease at the end of chemotherapy. The patient was lost to follow up after that and expired with a total survival time of one year and 9 months from the date of diagnosis.

DISCUSSION

Desmoplastic Small Round Cell Tumour (DSCRT) is a rare malignant tumour with a distinct histological appearance. A literature review done recently found only 13 cases (4). A pure morphological diagnosis is sometimes difficult and molecular techniques can be helpful to differentiate between other poorly differentiated tumours. The typical presentation is with a paraserosal mass and a predilection for serosal surfaces, mainly the peritoneum and the paratesticular region as seen in the present case. DSCRT shows small round cells growing in a nested pattern with abundant, desmoplastic stroma (5).This tumor was originally described as an extensive aggressive intra-abdominal mass with widespread peritoneal and lymphatic dissemination at diagnosis. The patient in our study also had disseminated disease at presentation and it clearly shows its aggressive nature (6).

The immunohistochemical profile is characterized by the co-expression of epithelial, mesenchymal and neural markers. Gerald et al suggested that DSRCT is a primitive tumour related to mesothelium, because of the prevalence in serosal cavities and its polyimmunophenotypic nature (2).DSRCT is associated with unique chromosomal translocation t (11:22) (p13:q12) resulting in an EWS/WT1 transcript. This feature is diagnostic of DSRCT. This transcript codes for a protein which is a transcriptional activator that fails to suppress tumor growth (7).

The lesion has to be differentiated from other tumors affecting the paratesticular region like rhabdomyosarcoma or malignant lymphoma (8). Due to rarity of this tumor the ideal therapeutic approach is missing and there is no standard protocol. (9). Paratesticulartumors can usually be completely resected, but this is not the case for disseminated abdominal tumors. Most authors recommend aggressive chemotherapy, however, different regimens havebeen employed (10).The diffuse nature of the tumor often makes resection with negative microscopic margin impossible and extensive aggressive surgical approach adds only to the morbidity. Hence intensive high dose chemotherapy combined with tumor debulking and whole abdomino-pelvic radiation therapy have been attempted with the hopes of prolonging disease-free survival.

CONCLUSION

DSRCTs is rare and remains a vexing disease that deserves early diagnosis and definitive treatment. Overall, the prognosis for DSRCT is poor with survival rates of <20% at 2 years. Though rare, diagnosing of this entity at the early stage remains important and should be kept in mind along with common differential diagnosis.

Multimodality treatment approach with high dose aggressive chemotherapy, optimal surgical debulking, radiotherapy and myeloablative chemotherapy with stem cell rescue have been attempted.

REFERENCES

1. Gerald W.L., Rosai J. Case 2. Desmoplastic small cell tumor with divergent differentiation. Pediatr. Pathol. 1989;9:177–183
2. Gerald WL, Miller HK, Battifora H, et al. Intra-abdominal desmoplastic small round-cell tumor. Report of 19 cases of a distinctive type of high-grade polyphenotypic malignancy affecting young individuals.Am J SurgPathol 1991;15:499 –513.
3. Crombleholme TM, Harris BH, JacirN, et al. The desmoplastic round cell tumor:a new solid tumor of childhood. J PediatrSurg1993;28:1023–5.
4. Roganovich J, Bisogno G, Cecchetto G, et al. Paratesticular desmoplasticsmall round cell tumor: case report and literature review J.SurgOncol 1999;71:269 –72.
5. Ordóñez NG, el-Naggar AK, Ro JY, Silva EG, Mackay B Hum Pathol. 1993 Aug; 24(8):850-65.
6. Roganovich J, Bisogno G, Cecchetto G, et al. Paratesticular desmoplastic small round cell tumor: case report and literature review J. SurgOncol 1999;71:269 –72.
7. Murphy AJ, Bishop K, Pereira C, Chilton-MacNeill S, Ho M, Zielenska M, et al. A new molecular variant of desmoplastic small round cell tumor: Significance of WT1 immunostaining in this entity. Hum Pathol 2008;39:1763-70.
8. Leuschner I, Newton WA Jr., Schmidt D, et al. Spindle cell variants of embryonal rhabdomyosarcoma in the paratesticular region. A report of the Intergroup Rhabdomyosarcoma Study. Am J SurgPathol 1993; 17:221–30.
9. Farhat F, Culine S, Lhommé C, et al. Desmoplastic small round cell tumors: results of a four-drug chemotherapy regimen in five adult patients. Cancer 1996;77:1363– 6.
10. Bisogno G, Roganovich J, Sotti G, et al. Desmoplastic small round cell tumor in children and adolescents. Med PediatrOncol 2000;34: 338–42.