Desmoplastic small round cell tumor - abdomen

Sir,

Desmoplastic small round cell tumor (DSRCT) is a very rare malignancy of childhood and young adults, with male preponderance presenting as intra-abdominal or pelvic tumor. This aggressive malignant neoplasm tends to present with vague abdominal discomfort/distension, abdominal pain, weight loss, or change in bowel habits.

A 24-year-old male presented to a hospital with history of pain in the abdomen and significant weight loss since 2 months. The ultrasonography revealed large multiple solid masses in the left hypochondrium, bilateral lumbar and pelvic regions, the largest measuring 11cm in the greatest dimension. A differential diagnosis of abdominal tuberculosis and lymphoma were offered. At our center, on examination, he was detected to have a solid mass in the left iliac fossa and an enlarged left supra-clavicular lymph node. CT scan revealed extensive serosal, peritoneal and mesenteric soft tissue masses with retroperitoneal lymph nodes. Possibility of GIT malignancy with secondaries and soft tissue sarcoma were added. FNAC of the left iliac fossa revealed sheets and nests of small round cells with round-to-oval nucleus, which were hyperchromatic, regular nuclear margins, scanty cytoplasm and a moderate degree of pleomorphism. A differential diagnosis of round cell sarcoma and NHL were offered. It was advised to get bone scan, screening for testis and serum markers - CEA, AFP, CA19.9 to rule out any occult carcinoma. The biopsy of the supraclavicular mass showed effaced architecture, nests and islands of round blue cells small in size with high mitotic activity, hyperchromatic nuclei and overcrowding. Dense fibrous bands separating the metastatic tumor were also seen [Figure 1]. Immunohistochemistry revealed that the tumor was positive for pan-cytokeratin, desmin; and negative for CD45,

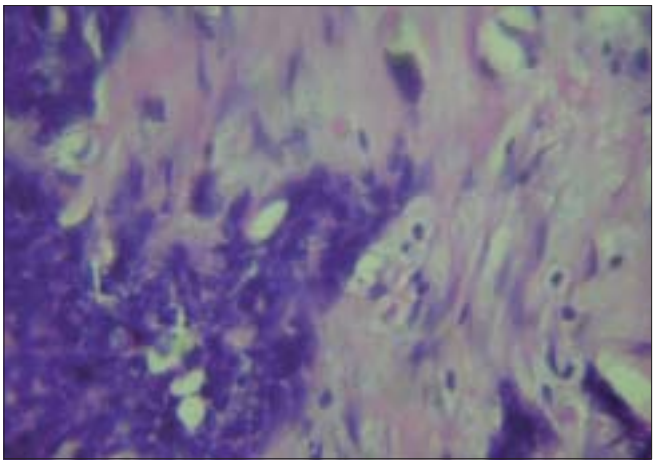

Figure 1: Supraclavicular lymph node - DSRCT (H&E, ×400)

CD117 and AFP. A final diagnosis of DSRCT was made. The patient was started on chemotherapy of Adriamycin, Cyclophosphamide and Vincristine. The patient after five cycles of chemotherapy was subjectively better, asymptomatic and the CT scan revealed the greatest dimension of tumor as 6cm.

The histological differential of round cell tumor includes Ewing's sarcoma, neuroblastoma, Wilm's tumor, rhabdomyosarcoma and primitive neuroectodermal tumor.[1] DSRCT has a characteristic IHC described above and balanced translocation involving chromosomes 11 and 22 (t (11:22) (p13: q12)), similar to PNET and Ewing's sarcomas.[2,3] Metastasis to liver, bone marrow and lymph nodes has only been documented in around 33% of DSRCT cases.[3]

Most investigators believe that the tumor originates from the mesothelium (or from submesothelial or subserosal mesenchyme), which is most extensive in the peritoneum. DSRCT has also been described at other sites, including the paratesticular region, pleural region, lung, ovary and sinus cavity.[4]

The cytological aspirate of DSRCT shows moderate-to-high cellularity, with tumor cells arranged singly and in clusters. The cells demonstrate high nuclear/cytoplasmic ratios, granular chromatin reminiscent of small-cell carcinoma, usually inconspicuous nucleoli, smooth-to-irregular nuclear membranes and frequent nuclear molding. The cytoplasm is scant-to-moderate, pale blue and occasionally vacuolated. The diagnosis only based on cytology at times is very difficult as the differential diagnosis is very vast. Apart from clinical feedback, immunocytochemistry and cytogenetics become essential to make the diagnosis only on aspirate.[5]

Radiological differential diagnoses of this entity include peritoneal carcinomatosis from primary malignancies, malignant mesothelioma, gastrointestinal carcinoid, malignant melanoma, soft-tissue sarcomas (MFH, desmoid fibromatosis), peritoneal tuberculosis, fibrosing mesenteritis, splenosis and amyloidosis. Median survival is approximately 17 months. However, aggressive multimodality therapy consisting of high-dose chemotherapy, surgical resection (debulking) and radiotherapy (external beam 30Gy) may prolong the survival.[3,6]

Prateek Kinra, A. K. Pujahari[1]

Department of Pathology, Institute of Aerospace Medicine, Bangalore - 17, [1]Command Hospital Air Force, Bangalore, India

Address for correspondence:
Prateek Kinra, Department of Pathology, Institute of Aerospace Medicine, Vimanpura, Bangalore - 560 017, India.
E-mail: pkinra_in@yahoo.com

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