

Case of the Week

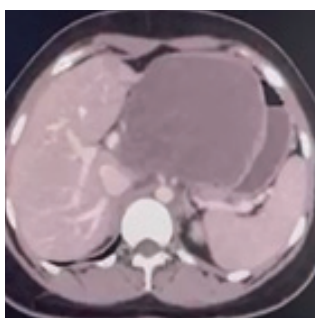
DESMOPLASTIC SMALL ROUND CELL TUMOR OF HEPATODUODENAL LIGAMENT – A RARE ENTITY

Desmoplastic small round cell tumour (DSRCT) is a rare but highly aggressive malignant tumour. It predominantly affects adolescents and young adults. It is a type of soft tissue sarcoma. Currently, there is no therapeutic pattern or standard of care for DSRCT. However, it is very important to work with a team of experts as soon as possible after diagnosis to improve the outcomes. The present case describes a rare entity of DSRCT.

An 18-year-old female was evaluated for pain/abdominal mass in Feb 2021. CECT Scan showed a large multi-cystic mass of 20x20cm in hepatoduodenal ligament infiltrating liver, tumour markers were normal. Clinico-radiological diagnosis of hydatid cyst was made and she was subjected for Exploratory laparotomy with R2 resection (partial) of the mass. Postoperative histopathology report confirmed diagnosis of DSRCT. She received 6# chemotherapy

(Ewing's protocol). Response evaluation scan showed good response with residual mass posterior to the liver, below bifurcation of middle and left hepatic veins. After a thorough discussion in multidisciplinary joint clinic, decision for surgery was done. Intraoperatively, there was a 4X3 cm-sized growth behind the caudate lobe in retro-hepatic region with frozen porta hepatis and no evidence of peritoneal metastases. Lt hepatectomy with en block excision of mass was done. Intraoperative and postoperative course was smooth and uneventful. Histopathology report revealed no residual viable tumour. She is advised to complete the adjuvant chemotherapy.

DSRCT in hepatoduodenal ligament location is very rare and to the best of our knowledge only few cases are reported. This was a unique case in terms of rare diagnosis, re-do surgery and very tricky location making surgery technically challenging and difficult.



At Presentation



Residual mass after chemotherapy

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