International Journal of Humanities Social Science and Management (IJHSSM)
Volume 3, Issue 1, Jan.-Feb. 2023, pp: 570-571 www.ijhssm.org

Case Report: Diagnostic Challenge of Very Rare Nodal Presentation of Desmoplastic Small Cell Tumor.

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Date of Submission: 12-02-2023 Date of Acceptance: 24-02-2023

ABSTRACT :
A Desmoplastic small cell round cell tumour is a rare soft tissue sarcoma & highly aggressive tumour. Clinical diagnosis for desmoplastic small round cell tumour is challenging as patients present with vague symptoms. Correlating various investigations procedures required to confirm the diagnosis.Taking into consideration of rarity of tumour, both diagnosis and management of tumour there is no structured plan, making it difficult in diagnosis of tumour and treatment of disease. In this case report, we present a case of very rare nodal presentation of desmoplastic small round cell tumour of young female who presented with abdominal pain, low backache and loss appetiteand various investigations we have done to confirm the diagnosis.

Keywords :Desmoplastic small round cell tumour, diagnosis, nodal presentation.

CASE REPORT: A 43 year old femalecame to our institutionwith complaints of low backache and abdominal pain since 4 months, with loss of appetite since 3 months. On clinical examination patient had,palpable node present on left side in lower jugular region of app. measuring 1.5x1 cm, hard in consistency, nonmobile, other node palpable at left supraclavicular region of app. measuring 2x1cm, hard in consistency, non mobile, no other significant lymphadenopathy and with abdominal distension on palpation ill defined mass palpable over epigastric and lefthypochondrium region , of 10x 7cms, irregular in shape, firm in consistency, with ill defined margins, with no mobility. USG abdomen of patient shows a large well defined hypoechoic mass seen in tail region pf pancreas measuring 9.6x6.3 cm - s/o **malignancy, multiple pre & para aortic &peri portal lymph nodes** are seen.Later CECT abdomen done which showed a large lobulated minimally enhancing lymph nodal mass noted in gastrosplenic, hepatodudoneal , gastrohepatic,periportal , peripancreaticspaces measuring app**.11.5x7.9x7.7cm.**This **nodal mass is seen encasing the body & tail of pancreas** which on contrast administration shows hypo enhancement.Radiologically the impression given as Lymphoma .CT guided biopsy was done**.**CT guided biopsy reported as 1) small round cell tumour 2) non hodgkin lymphoma.

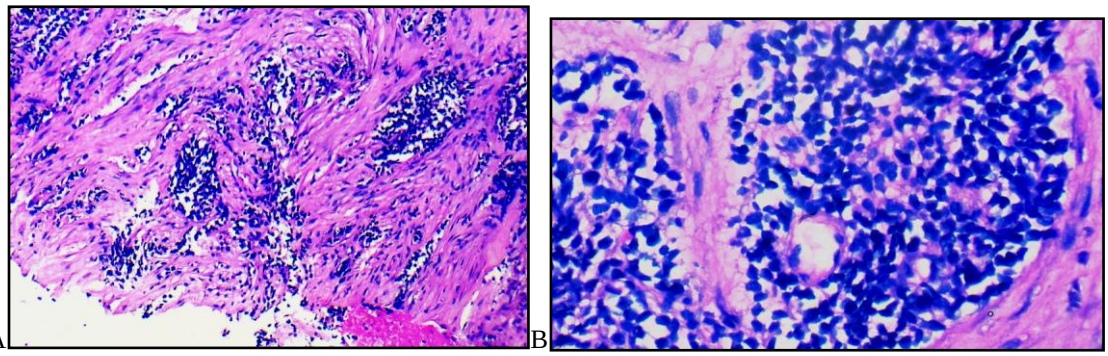

Figure 1:AExtensive fibrosis with entrapped nests and sheets of neoplastic cells, H&E, 100x

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B.Round to oval mildly pleomorphic tumor cells with hyperchromatic nuclei, increased N:C ratio and scant eosinophilic cytoplasm, H&E, 400x

Following IHC markers are done to differentiate lymphoma and small round cell tumour :
CD3, CD20,GFAP - negative,**Desmin positive KI 67 40 to 50 % Pancytokeratin positive, CD 99 showing cytoplasmic positivity**. As per test result desmin positive and cytoplasmic CD99 positivity differentiate between ewingsaroma and desmoplastic small round cell
So IHC studysays tumour as desmoplastic small round cell tumour.
PET CT scan shows intensely increased FDG concentration noted in few enlarged **left level IV** And **left supraclavicular** region measuring **24 x15 mm Suv max 11.7 .**Intensely increased FDG concentration noted in multiple discrete and conglomerate nodal massed involving p**eripancreatic ,periportal, pre paraaortic**region. The lesions is encasing the body and tail region of pancreas completely and is extending into lesser sac and gastrohepatic ligament and casing indentation over stomach and spleen. Encasement of aorta and its branches also seen, largest dimension lesion measuring **153x90x90mm with SUV max 16.3. - likley lymphoma**Intensely increased FDG concentration noted in few subcentimeter size to few centimeter size **mesenteric and b/l common iliac lymph** nodes with **SUV max 4.9 .**

Figure 2 :PET CT scan showing peripancreatic lesion encasing pancreas

PET CT scan impression suggestive of lymphoma that gain lead to consfuion in the diagnosis.
So we done cytogenetic testing in which FISH showed Positive for rearrangement of ESWR1 GENE located at 22q12.2 in 80% of cells analyzed.

Bone marrow aspiration and biopsy showed mildhypoplastic marrow for age .Flow cytometric analysis shows no overt lymphoproliferative disorder.

All the above tests and investigations lead to final diagnosis of desmoplastic small round cell tumour. Because of limited knowledge about disease there is no standardized treatment approach. As disease burden is high for this patient we planned for systemic chemotherapy 6 cycles of VAC +IE . After completion of 6 cycles reassessment followed by local therapy either by surgery or radiation therapy.

CONCLUSION :
For the final diagnosis of tumour it requires correlation of various investigations like clinical, radiological, immunohistochemical and genetical studies.