Original Research Paper

Volume - 14 | Issue - 02 | February - 2024 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Gastroenterology

A RARE CASE OF EWINGS SARCOMA OF PANCREAS

Dr. Ubal Dhus	Medical Gastroenterology Senior Consultant.
Dr. Piramanayagam	Medical Gastroenterology Senior Consultant.
Dr. Deepak Kannan	Medical Gastroenterology Registrar, Apollo Hospitals, Greams Road, Chennai.
Dr. Megha Sharma	Medical Gastroenterology Registrar, Apollo Hospitals, Greams Road, Chennai.

KEYWORDS :

INTRODUCTION :

Small round cell tumor is a rare lethal malignant tumor seen mostly in abdominopelvic regions but extremely rare in pancreas. It can have a wide range of presentations from asymptomatic to life-threatening comorbidity, and it responds poorly to treatment despite aggressive therapy.

CASE-DETAILS:

A 24 year old male presented with epigastric discomfort along with weight loss of 10kgs over 3 months period. complete blood count, renal and liver function test were normal. Ultra sound abdomen showed a hypoechoic mass measuring 6.0 x 5.6 cms in body of pancreas. CECT abdomen showed 5.2cm smooth well circumscribed mass forming lesion in mid body of pancreas homogenously hypodense in all contrast phases with a narrowed splenic vein due to compression along with a bland thrombus distal to compression. There was a 1.5x0.7cm enchancing lymph node and a normal main pancreatic duct.EUS done showed a 4x5cm echogenic mass lesion in body of pancreas along with space-occupying lesion of 6x4mm in liver.Biopsy reported as malignant small round cell tumour with immunohistochemistry showing positive for VIMENTIN ,CD99, NKX2.2, FLI-1 and 60-70% positive for ki-67 suggestive of Ewings family of tumours. PET CT showed metabolically active uptake in primary pancreatic mass along with perigastric and peripancreatic nodal metastasis. There was also soft tissue enhancement in nasopharynx and bilateral palatine tonsils with active FDG uptake in cervical nodes Oncologist advised VAC regimen -vincristine sulfate, actinomycin-D and cyclophosphamide along with IE (Ifosfamide,etoposide) 8 cycle.Post this chemotherapy repeat PET CT was done showed a significant reduction in size of tumour and its metabolic activity.there was also a interval resolution in the activity around the nasopahrynx and palatine tonsils along with cervical nodes activity.

DISCUSSION:

Ewings sarcoma has no gender predilections and more than 70% of patients present in there teenage years.There is no specific signs and symptoms except the location of primary tumour[1].Small cell tumour of pancreas around 91% of patient present with metastasis at the time of diagnosis.[2]

There are no characteristic radiological signs , most lesions are encapsulated ,hypoechoic on USG ,hypodense on CT and iso-hypointense on T1 ,hyperintense on T2 in MRI. Finally the diagnosis is confirmed with cytogenetics and immunohistochemistry[3].

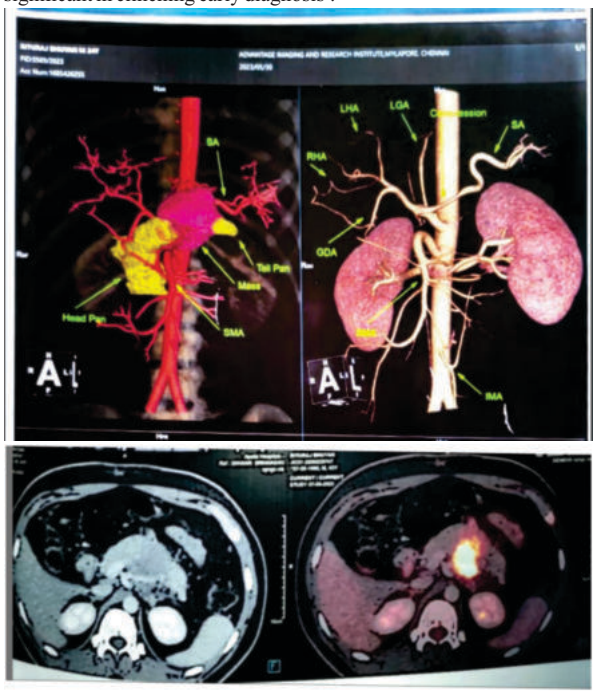
It is crucial that a patient diagnosed with this condition needs a multidisciplinary expert team management. Ewings sarcoma is a chemosensitive malignancy.The most commonly used agents are (doxorubicin,cyclophosphamide,vincristine)a three drug therapy or a five drug therapy with (combined with ifosfamide and etoposide) [3]. Gier et al showed that patients receiving a five drug vs three drug showed an improved survival rate of 72% vs 61%.[1]

Radiation therapy has a role particularly in patients after residual disease after surgery and chemotherapy.Proton beam radiation therapy and intensity modulated radiation therapy (IMRT) showed promising results but lack randomization.

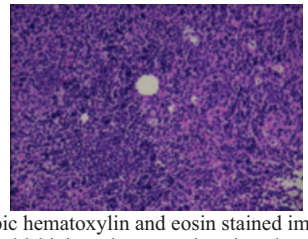
Here we treated with 5 drug regimen and patient has been able to tolerate it without significant side effects.

CONCLUSION:

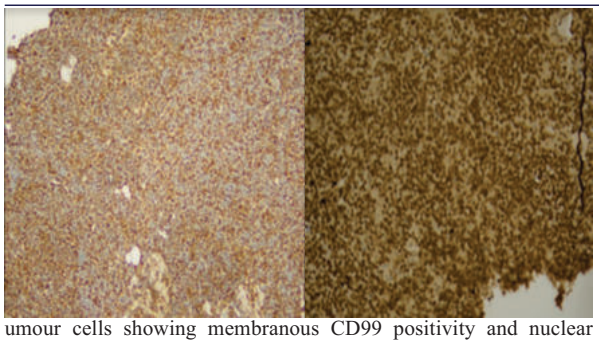
Extraosseous Ewings sarcoma is a rare entity with a diagnostic dilemma. Early recognition provides the best chance of survival with current treatment modalities. Extraosseous Ewing Sarcoma should be considered in the differential diagnosis of intraabdominal and extraintestinal masses, especially in patients with no specific signs and symptoms.The importance of histology and immunohistochemistry is significant in clinching early diagnosis[4].



CECT AND PET CT showing mass enchancing lesion In the body of pancreas.



Light microscopic hematoxylin and eosin stained images show small round blue cells with high nuclear cytoplasmic ratio.



umour cells showing membranous CD99 positivity and nuclear positivity for FLI-1

REFERENCES:

1. Grier HE, Krailo MD, Tarbell NJ, et al.: Addition of ifosfamide and etoposide to standard chemotherapy for Ewing's sarcoma and primitive neuroectodermal tumor of bone. N Engl J Med. 2003, 348:694-7
2. Wang D, Rong Y, Wu W, Jin D. Primary small cell carcinoma of the pancreas: rare type of pancreatic cancer and review of the literatures. World J Surg Oncol. 2012 Feb 8;10:32. doi: 10.1186/1477-7819-10-32
3. Bose P, Murugan P, Gillies E, et al.: Extraosseous Ewing's sarcoma of the pancreas. Int J Clin Onc. 2012, 17:399-406.
4. Yachida S, Zhong Y, Patrascu R, Davis MB, Morsberger LA, Griffin CA, Hruban RH, Laheru D, Iacobuzio-Donahue CA. Establishment and characterization of a new cell line, A99, from a primary small cell carcinoma of the pancreas. *Pancreas.* 2011;**40**(6):905–910.
5. Winter JM, Narang AK, Mansfield AS, Herman JM, Cameron JL, Laheru D, Eckhauser FE, Olson MT, Hruban RH, Miller RC, Andersen DK. Resectable pancreatic small cell carcinoma. *Rare Tumors.* 2011; ;**3**(1):e5.