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A CASE REPORT OF PRIMARY HEPATIC NEUROENDOCRINE TUMOUR

Pathology

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ABSTRACT

INTRODUCTION: Primary hepatic neuroendocrine tumours are rare. Excluding extrahepatic origin of the tumour with liver metastasis is important.

CASE REPORT: A 55year old male complaining of weight loss, intermittent fever, anorexia for 2 months .No abnormality detected in routine investigations. On whole body CT-scan a rounded solitary space occupying hepatic lesion measuring 3.5x 3.4 was seen. USG-guided FNAC was done. MGG stain smear shows small round cells with high N-C ratio, stippled chromatin and indistinct nucleoli. Further, IC staining shows positivity with synaptophysin and chromogranin A which confirms the diagnosis of primary neuroendocrine tumour of liver.

CONCLUSION: PHNEC should be considered as a possible differential diagnosis in management of hepatic tumours. Surgical resection must be considered for curative treatment.

KEYWORDS

INTRODUCTION

Neuroendocrine tumours have extremely been rare in liver with only about 150 cases [1] having been reported, it represents about 0.3% [2] of all neuroendocrine tumours.

NET originates from dispersed neuroendocrine cells distributed throughout the body.

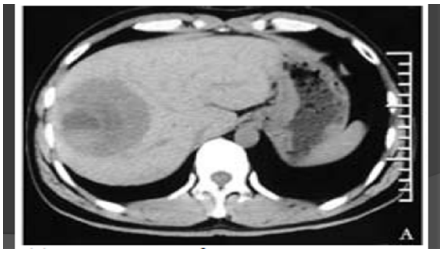
Neuroendocrine tumours primarily arise in the bronchopulmonary or gastrointestinal tracts (stomach, ileum, duodenum, jejunum, pancreas, appendix, meckel diverticulum, rectum, colon) and account for 70% of all NETs found in the body [3].

CASE REPORT

A 55 year old male complaining of weight loss, intermittent fever and associated with anorexia for last 2 months.

Routine haematological investigations and thyroid scan were within normal limits. No abnormality detected in gastrointestinal endoscopy and chest X-ray. On whole body CT scan a hypo dense, round, solitary space occupying lesion in liver is noted of size 3.4 X 3.5 cm.

Fig: 1 CT scan showing hypodense solitary liver mass



Ultrasound guided FNAC was done. MGG stained smear shows highly cellular material with discohesive, small and round cells with scanty cytoplasm, high nucleo-cytoplasmic ratio, round nuclei, regular nuclear outline, stippled chromatin and small indistinct nucleoli with clear background.

Fig 2: MGG stained smear at low power

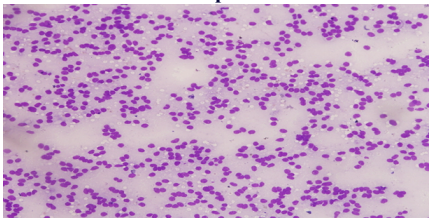


Fig 3: MGG stained smear at high power

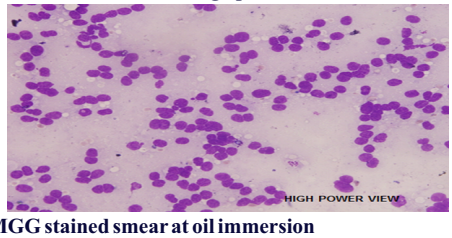


Fig 4: MGG stained smear at oil immersion

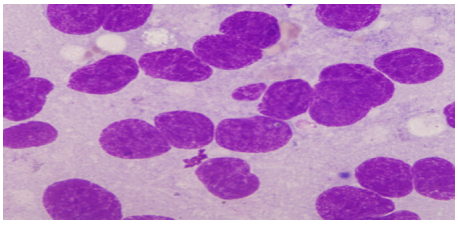
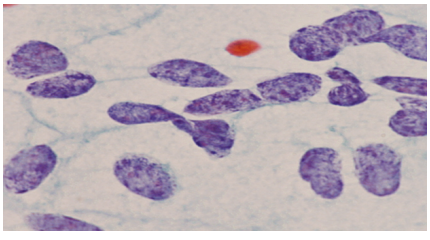


Fig 5: PAP stained smear



Tumor markers AFP, CEA and CA19-9 and liver function test were within normal limits.

Further Immunocytochemistry was advised which showed positivity with synaptophysin and chromogranin A, which rules out HCC and confirms the diagnosis of neuroendocrine liver tumor. Surgical resection of the tumor is done. There is no recurrence till now.

Fig 6: Showing Chromogranin A positive

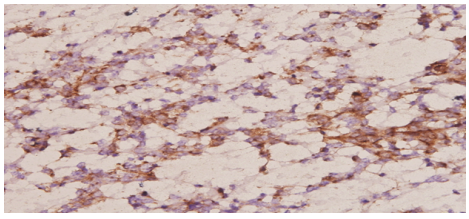
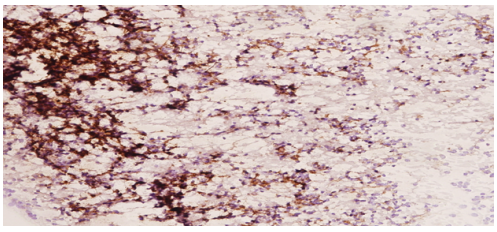


Fig 7: Showing Synaptophysin positive



DISCUSSION

PHNEC are uncommon and usually presents as solitary large hepatic mass associated with vague symptoms or abdominal pain. Its considered slow growing and mildly aggressive. 5% of NET presents with carcinoid syndrome. Rarely metastasis is to bone, lymph node and lung may occur.

When a NET is found in the liver, it must be first treated to exclude metastasis from extrahepatic primary site, because liver is the most common site of NET metastasis. Primary can be ruled out by performing gastrointestinal endoscopy, CT of chest and abdomen, whole body PET-CT and octreotide scanning. Regular screening at interval is required to rule out any recurrence.

Diagnosis can be confirmed by testing immunoreactivity for chromogranin A (89.1%), neuron specific enolase (74.1%) and synaptophysin(48.9%) [1]. In our case showed positivity for chromogranin A and synaptophysin.

Aggressive treatment is not required. When lymph node and distant metastasis are absent, liver resection is adequate for treatment. If metastasis is present, adjuvant chemotherapy may be needed. Potential chemotherapy includes 5-Flurouracil, adriamycin, mitomycin, etoposide and cisplatin. Transarterial chemoembolization, liver transplant or radiotherapy treatments also have been reported as effective treatment.

Prognosis is favourable. Five year survival and recurrence rate after surgical resection have been reported as 74% and 18% respectively [4].

Differential diagnosis are metastatic hepatic neuroendocrine tumor, hepatocellular carcinoma small cell type, metastatic adenocarcinoma, small cell lymphocytic lymphoma, small cell undifferentiated carcinoma, tuberculosis, hydatid cyst, amoebic abscess etc.

CONCLUSION

Primary hepatic neuroendocrine carcinoma should be considered as a possible differential diagnosis in management of hepatic tumors. And if the tumor are diagnosed as PHNEC, surgical resection must be considered curative.

Long term follow up and further investigations are needed for better understanding the outcomes and nature of this rare tumor.