To be or not to be a primary hepatic neuroendocrine tumour

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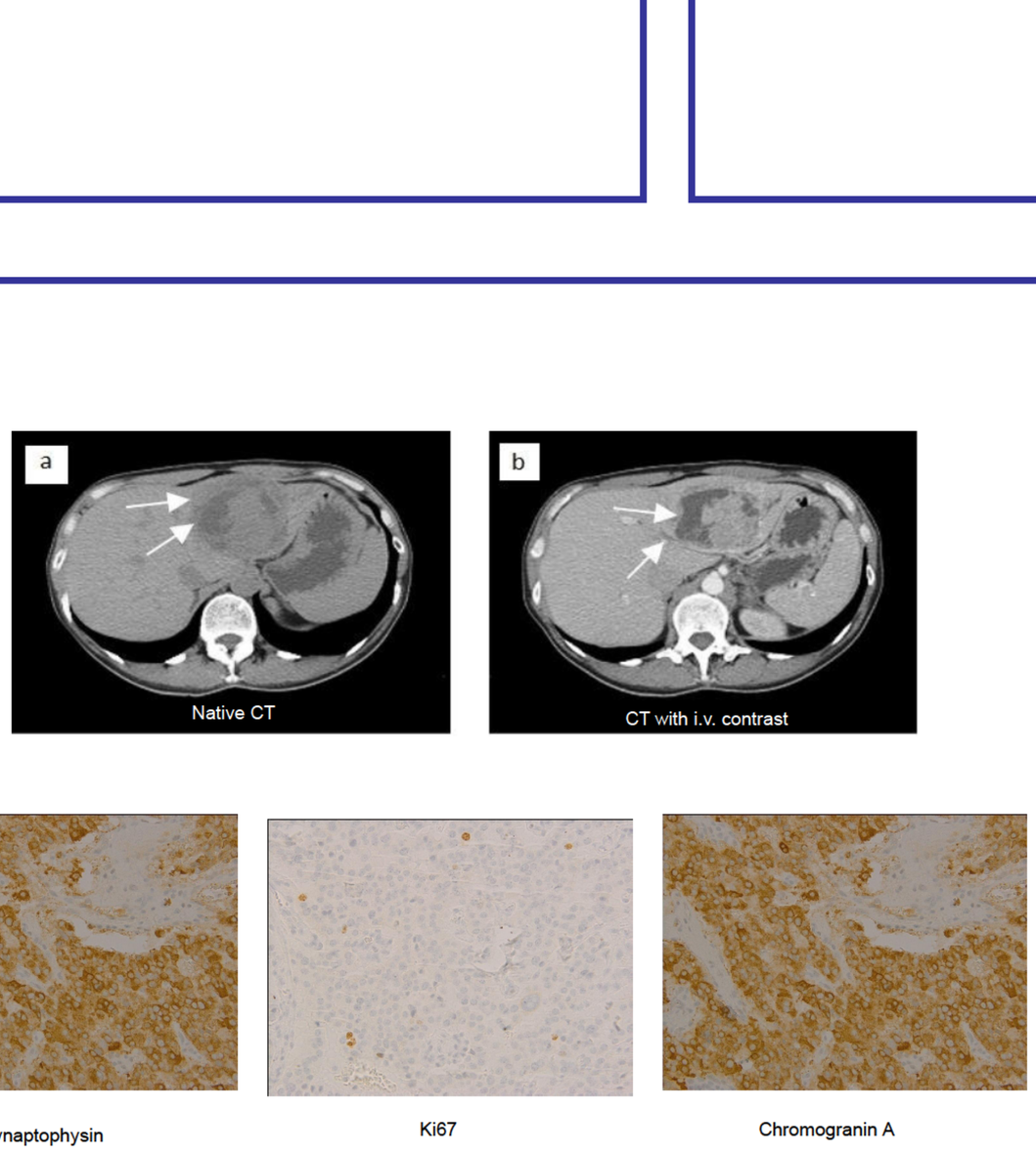
OBJECTIVES

Primary hepatic neuroendocrine tumours (PHNETs) represent approximately 0.3% of all neuroendocrine malignancies. The explanation for this small percentage is due to the fact that the liver is usually the preferential site of metastatis from neuroendocrine tumours (NETs), but not the preferential site of migration of neuroendocrine cells during embryogenesis.
PHNETS are large in size at presentation, indolent in behavior and in most cases they do not associate with carcinoid syndrome. The mean age of occurrence is 49.8 years and women are slightly more affected than men.

METHODS

We present the case of a 45-year-old Caucasian female who was submitted for nausea, vomiting, diarrhea and diffuse abdominal pain. The onset of symptoms occurred 7 days prior to the presentation.

- The patient had no history of liver disease.
- No family history of malignancies.
- Physical examination revealed no abnormalities.



RESULTS

The enhanced computed tomography of the abdomen demonstrated an encapsulated liver mass, measuring 7/8.5/9 cm, that contained multiple daughter cysts separated by a matrix. The initial diagnosis was of an Echinoccocus cyst; left atypical resection was performed.
The histological and immunohistochemical exams revealed a brown-greyish solid cystic tumour outlining a heavily vascularized cell proliferation with trabecular and band disposition, with a homogenous growth pattern that was diffusely positive in the tumour cells for chromogranin A, synaptophysin and neuron specific enolase. **Ki67 index was 2-4%.**
The postoperatory evaluation revealed:

- the specific neuroendocrine markers (serum chromogranin A and 24h urinary 5-hydroxyindolacetic acid) were within normal limits.
- the ^{111}In DTPA-octreotide scan showed no regions that were somatostatin receptor positive and the PET scan with 5HTP demonstrated no tumour mass in the left liver lobe.

CONCLUSIONS

Despite the fact that PHNET is a rare medical entity, we believe that most cases reported in English literature are **overdiagnosed** due to insufficient medical evaluation. It is of the outmost importance to differentiate an ocult primary neuroendocrine tumour from a PHNET, as the two have different therapeutical approaches. Thorough long-term investigations are recommended in all cases of PHNETs.

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