NEUROENDOCRINE TUMOUR OF LIVER: A CASE REPORT

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ABSTRACT
Neuroendocrine tumours of the liver are rare. This is the case report of a woman, 28 years of age, who presented with right upper abdominal pain, fever and abdominal mass for six weeks duration. Ultrasonography of the abdomen revealed hepatomegaly with two cystic mass lesions diagnosed as hydatid cysts, though her Echinococcus antibody titre was normal. A laparotomy was carried out which showed enlarged liver with a knobby mass, partially cystic and partially solid, arising from the inferior surface of the right lobe of the liver. Partial right hepatectomy was performed. The histopathology report revealed it to be a neuroendocrine tumour of the liver.

KEYWORDS: Primary Hepatic Tumours, Neuroendocrine Tumours, Immunohistochemistry

INTRODUCTION
Most of the neuroendocrine tumours of the liver arise from the gastrointestinal tract, especially from the appendix, ileum and pancreas. Primary hepatic neuroendocrine tumour is a rare clinical entity, requiring strict exclusion of possible extra-hepatic primary sites for its diagnosis.

Histopathologically the tumours are well-differentiated (classified as carcinoids) to poorly differentiated anaplastic type (classified as neuroendocrine carcinoma). However, microscopy alone does not necessarily predict their biological behaviour. Hence they are characterized by their contents of intracytoplasmic secretory granules shown by the electron microscopy and by chromogranin immunocytochemistry.

CASE REPORT
A female aged 28 years, resident of Karachi presented with history of pain in the right upper abdomen for six weeks. The pain was dull, continuous in nature, mild to moderate in severity, aggravated by movements and cough, and decreases on taking analgesics. The pain was associated with nausea, retching and fever, which was low grade and intermittent without any rigors and chills. There was no history of weight loss or altered bowel habits, but she did complained of decreased appetite and insomnia.

She had a Caesarean section about eight years ago, but no history of jaundice or any other problem in the past. She had a history of taking analgesics (NSAIDS) and antacids (H₂ blockers).

On examination she was a young lady of average height and built, lying uncomfortably on bed, with normal vital signs. Chest examination revealed harsh vesicular breathing with slight decreased entry in the right lower lung.

The abdomen showed a bulge in the right upper abdomen, striae gravidarum and a lower midline (Caesarean) scar on inspection. There was tenderness in the epigastric and right hypochondriac region. The liver was enlarged four fingers below the costal margin with a vague knobly lump at the right lower end.

 Investigations including blood complete picture, LFTs, urea and creatinine, blood sugar, hepatitis viral markers, and X-ray chest and abdomen were normal. An ultrasound of the abdomen showed hepatomegaly with two well defined cystic mass lesions (9.2x6.9 cms and 5.1x5 cms) having a thick capsule around them and daughter cysts within, in the right lobe of the liver. The conclusion was that these were hydatid cysts, though the Echinococcus antibody titre turned out to be within normal limits.

After necessary preparations the patient underwent a laparotomy. At operation the liver was found enlarged

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with a large (8x6cms) bossellated mass, partially cystic and partially solid, arising from the inferior surface of the right lobe of the liver. The mass was excised with a wide margin of healthy tissue around it, a partial right hepatectomy being performed (Figs. 1 & 2). The abdomen was closed with drainage, after securing haemostasis. There was no significant post-operative complication and the patient was discharged home in a satisfactory condition.

The histopathology report of the excised mass revealed a neoplastic lesion, composed of cells arranged in a trabecular architecture with predominately round nuclei showing mild pleomorphism, salt and pepper chromatin, conspicuous nucleoli and occasional mitotic activity (1 to 2 mitosis/10HPF). No definitive foci of necrosis were identified, but multiple small foci of similar lesions were seen scattered in the surrounding liver parenchyma. The sections were stained with a panel of following monoclonal antibodies using envision system and the tumour cells were found to be:

- Cytoplasmic glycogen (PAS±Diastase) Negative
- Cytokeratin AE1/AE3 Positive
- Cytokeratin 7 Positive
- Cytokeratin 20 Negative
- Chromogranin Positive
- Synaptophysin Positive
- TTF-1 Negative
- CEA Negative

The conclusion of the report was - Liver parenchyma showing involvement by neuroendocrine carcinoma. She was referred to an oncologist for further opinion, but she never returned back.

DISCUSSION

The incidence of primary neuroendocrine tumours of the liver has been under-reported because of the assumption that it represents a metastatic lesion. Only 55 cases have been reported in the literature so far. The analysis of these published cases revealed that it occurs more frequently in females and in the middle age. Our case was also that of a female, but she was young (age 28 years).

The patients of these cases usually present with right upper abdominal pain, weight loss, progressive enlargement of the liver, as well as a palpable mass. Moreover, biochemical effects of the hormones produced by the tumour may be evident. In our case, the patient presented with right upper abdominal pain, nausea, retching, fever and hepatomegaly; but there were no hormonal manifestations.

Pre-operative diagnosis of primary neuroendocrine tumours of the liver is difficult, as radiological appearances on ultrasound, CT scan and MRI can mimic other pathologies. Hence primary carcinoid tumours of the liver have been described as hepatocellular carcinoma and cholangiocarcinoma on imaging, as reported by Fenwick et al in their series of eight cases. It can also simulate cystic lesions like the hydatid cyst as was seen in our patient, and previously mentioned by Schlauri and associates. The diagnosis of these cases is based on histopathology and immunohistochemistry for chromogranin and synaptophysin; both found positive in our patient.

The mainstay of curative treatment of primary hepatic neuroendocrine tumours is liver resection. This was observed by Donadon et al and Ruckert et al in their series. In our case also we performed a partial right hepatectomy. The data on the role of palliative chemotherapy in inoperable cases is insufficient. Andreola et al reported down-staging of the tumour by intensive systemic chemotherapy and subsequent resection in one
out of three patients.

CONCLUSION

Recognition of primary neuroendocrine tumours continue to be a challenge. An enhanced awareness about the rare sites of these tumours will result in an optimal management. Moreover, non-parasitic cystic lesions in liver should have a firm diagnosis by invasive means before definitive surgical intervention.

REFERENCES


