Primary hepatic neuroendocrine tumor: A case report

Vélez Pinto Juan Felipe*; Domínguez Prieto Víctor; Meliga Cecilia; Escanciano Manuel; Jiménez Fuertes Montiel; Guadalajara Labajo Héctor

Department of general and Digestive Surgery, Hospital Fundación Jimenez Diaz, Madrid, Spain.

Abstract

Primary neuroendocrine tumors (NET) of the liver are extremely rare, with approximately 150 published cases [2]. For the diagnosis is necessary to exclude its origin and presence in other locations and histological confirmation. When resection is feasible surgery is usually indicated. we present the case of a 57 year old patient with a primary NET on the liver.

Case presentation

A 57-year-old patient presented with dyspepsia. The abdominal ultrasound revealed a hyperechogenic lesion of 2cm on segment VIII of the liver. Liver MRI (Figure 1) showed a 22mm lesion of well-defined margins, hyperintense on T1 and T2, with significant restriction; in the dynamic study, the lesion had peripheral enhancement without central uptake on all phases.

Thoracoabdominpelvic CT scan was performed, which revealed no new lesions. Biopsy of the liver mass revealed a neuroendocrine tumor with positivity for Cytokeratin AE1/AE3, CD56, and chromogranin, and it was negative for CK7 and CK20, with Ki67 1%. Octreoscan (Figure 3) was performed, which only revealed uptake on the hepatic lesion. Gastrosopy, colonoscopy, and MRI enterography were performed, all of them without findings.

Figure 1: Liver MRI, arrow pointing the lesion with peripheral enhancement without central contrast.

Surgical resection of the lesion was performed; there were no other findings on the abdominal cavity. Thus, an appendectomy was performed also during the procedure. Histological examination of the resected lesion confirmed the diagnosis of neuroendocrine neoplasia, formed by midsize cells with eosinophilic cytoplasm, central nucleus, moderate pleomorphism, 20 mitoses per filed (x40). The tumor cells were arranged in solid nodules and occasionally rose like structures. On the immunohistochemistry the tumor was positive for CD56 Chromogranin, synaptophysin, Ki67 <1%, and low histological grade (G1). The patient is still on follow-up 22 months after surgery, but he remains free of disease.

Figure 2: Octreotide scan, shows uptake on a single hepatic mass, no other lesions are seen.
Discussion

Neuroendocrine tumors usually arise in the gastrointestinal tract (50%) or bronchopulmonary 30% [1]. The liver is a usual site for neuroendocrine metastasis. Upon diagnosis, it is important to differentiate between a primary and metastatic tumor. Primary neuroendocrine tumors of the liver are usually an incidental finding, without the characteristic carcinoid syndrome. Patient may present unspecific symptoms such as abdominal pain or discomfort, abdominal distention, or less frequently jaundice [1, 2]. Although there is no specific classification for primary NET of the liver, to determine tumor aggressiveness, WHO classifies NET as G1 G2, or G3, depending on the proliferation index or Ki67. Recurrence and survival at 5 years are 18% and 78% respectively. Given that most hepatic neuroendocrine carcinomas usually are metastasis from other organs, the diagnosis of a primary neuroendocrine tumor is based on the exclusion of the disease in every other location. Radiological findings are similar in primary and metastasic NET, bronchopulmonary, and gastrointestinal tract should be explored to exclude extrahepatic disease [1].

For resectable tumors, surgery is the treatment of choice, either wedge resection or partial hepatectomy, with a 5-year survival rate of approximately 70%. In non-resectable tumors, therapy can vary from arterial embolization, somatostatin analog therapy, systemic 5fluoracyl, and liver transplantation. Tumors of higher histological grade, Ki 67 and poorly differentiated have a worse prognosis [1, 3].

Conclusion

Primary neuroendocrine tumors of the liver are rare, the diagnosis is made usually as an incidental finding, patient may present unspecific symptoms such as abdominal discomfort, pain, mass effect, or jaundice. Diagnosis is based on the exclusion of other NET locations. Surgical excision is the treatment of choice when possible.

References

