INTRODUCTION

The neuroendocrine tumor, also known as carcinoid tumor, is a neoplasm of the diffuse neuroendocrine system. The occurrence of this type of tumor in the small intestine is rare and has a genetic influence in its etiology. It has been estimated that the deletion of the tumor suppressor gene PLCβ3 causes the uncontrolled growth of the neuroendocrine cells. The incidence ranges from 1 to 2 per 100,000 and affects men and women equally. Most of these tumors are well-differentiated and have an indolent course. Consequently, the onset of symptoms is late and, in most cases, the diagnosis is made at advanced stages of the disease. The chosen therapeutics is tumor resection. For the initial stages, the method aims at healing; while in advanced phases, the cytoreductive operation, associated to the multidisciplinary treatment, provides an increase of the survival time.

The objective of this study was to present a case of a metastatic neuroendocrine tumor of small intestine with a characteristic course of carcinoid syndrome.

CASE REPORT

A 57-year-old woman, referred to Hospital Santa Casa in Curitiba, PR, Brazil due to the presence of hepatic nodules that suggested of hemangiomas on abdominal ultrasonography (Figure 1) and diarrhea episodes with sporadic facial flushes.

During ambulatory investigation, the presence of 15 mm solid liver nodules in segments II and VIII was confirmed by computed tomography of the abdomen, with evidence of contrast washout in the late arterial phase. Magnetic resonance imaging (Figure 2) showed, in addition to nodules in segments II and VIII, other 5 mm nodules in segments V and VI. Due to the suspected malignancy of the hepatic tumor, the patient underwent biopsy of the liver, omentum and peritoneum.

The omentum and peritoneum presented no neoplasia. In hepatic tissue, the nature of the lesion was confirmed by immunohistochemistry, that showed positivity for synaptophysin (clone SY38), chromogranin A (clone DAK-A3), CDX-2, AE1/AE3 diffusely and Ki-67 positive on 2% of cells. The immunohistochemical panel was accordant with an infiltrative and well differentiated neuroendocrine tumor in the liver, with its origin in the digestive tract. After the biopsy results, was requested a 24 h urine 5-hydroxyindolacetic acid test.

Through computed tomography enterography and OctreoScan (Figure 3) was, respectively, confirmed the presence of polypoid lesions in the ileal region and mesenteric lymph node metastasis. The patient underwent an intraoperative enteroscopy and a tumor resection in the small intestine, with the removal of 80 cm of ileum and the realization of an enteroenteroanastomosis. Furthermore, were removed nine lymphnodes. She has evolved well in the postoperative period and was discharged with a prescription of Sandostatin LAR (Octreotide 20 mg at 4-week intervals).
A carcinoid syndrome, as it is known the clinical syndrome, is composed by a series of symptoms such as diarrhea, facial flushes, bronchospasm, cyanosis and inconstancy of the blood pressure as a result of the serotonin production. The syndrome affects around 5-7% of patients. In this case report, the patient represents the minority group that is affected by the clinical syndrome.

The chronic course is fairly typical of these tumors. Some patients might present less specific symptoms, like abdominal pain, small enterorrhages and intestinal obstruction. When such symptoms and the carcinoid syndrome are present, 12% of the individuals already show distant metastasis, mainly in the liver.

Regarding the hepatic metastasis, there is no specific protocol for therapeutics. Some studies argue that, in case of unresectable metastasis, the use of somatostatin analogs along with cytoreduction surgery promotes an increase in patient survival and an improvement in quality of life. The use of somatostatin analogues as a method of prevention of the carcinoid crises remains very controversial. The study made by Guo et al argue that these medications are inefficient. However, Gregersen et al concluded that, in addition to an improvement of the diarrheal episodes, the somatostatin analogs also reduce the serotonin, chromogranin A and urinary 5-hydroxindoleacetic acid levels. For the correction of the primary site, radical resection of the neuroendocrine tumor is the therapy of choice. In the present case, the approach chosen was the radical resection and the use of Sandostatin Lar, with a significant improvement in the clinical status and reduction of the urinary levels of 5-hydroxindoleacetic acid.

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