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Letter to the Editor

PARAGANGLIOMA OF THE PANCREAS: CASE REPORT AND LITERATURE REVIEW

Paraganglioma de pâncreas: relato de caso e revisão da literatura

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INTRODUCTION

Paraganglioma is a rare neuroendocrine tumor affecting about 1 out of 2.000.000 of the population. It arises from neural crest, including tissues such as the adrenal medulla, carotid and aortic bodies, organs of Zuckerkandl, and other unnamed paragangliomas[6,8].

Only 15 cases, including the present one, have been reported worldwide. The preoperative diagnosis is difficult to obtain, because it is sometimes confounded with other types of tumor. In general, it affects 65-year-old patients. In the contrast-enhanced CT scans, the tumors appear as soft-tissue masses with either homogeneous enhancement or central areas of low attenuation. The presentation is a solid in small tumor or cystic in larger tumors (>6 cm). Most paragangliomas follow a benign clinical course. The treatment of the choice is surgical resection and this case is the first described in Brasil.

CASE REPORT

A 73-year-old woman was admitted to our hospital complaining about right upper abdominal pains, 15 kg weight loss in one year and periodical post-prandial vomiting. She presented an abdominal ultrasonography showing a 2,5 cm mass at the head of the pancreas. The lesion was identified as a 2,5 x 1,7 cm low-intensity mass in T1 and T2 weighted magnetic resonance images. Magnetic resonance imaging with enhanced contrast revealed a homogeneous enhanced image in the arterial phase and venous phase. A contrast-enhanced computed tomography (CT) showed an enhanced lesion of 2,3 x 1,8cm in the head of the pancreas, with well defined limits (Figure 1). Laboratory studies gave normal results except for CA19-9 level, which was in superior limits of normality 35 U/mL (normal range 0-35 U/mL).

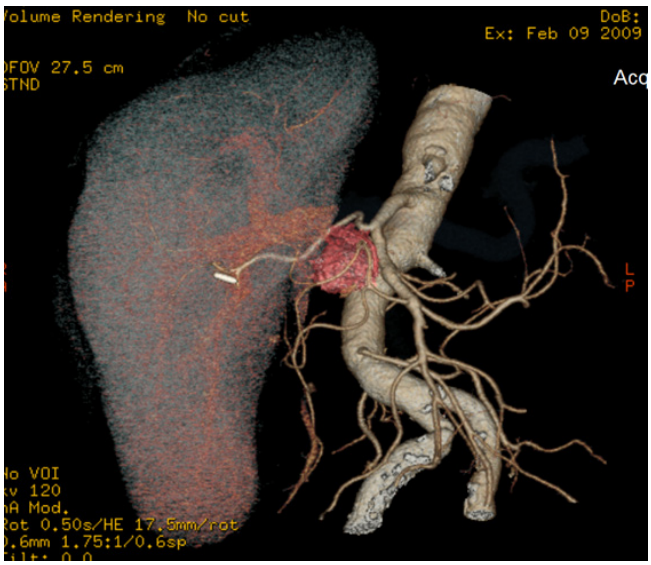

FIGURE 1 – Reconstruction of the lesion showing an arterial vascularization

The patient was submitted to a pancreatoduodenectomy with complete excision of the lesion, (Figure 2) with an uneventful postoperative course, and, in March 2010 one year after her operation, she was feeling well. The histological findings identified the "Zellballen" pattern and the immunohistochemistry revealed positive staining of synaptophysin, chromogranin A and S-100 protein (Figure 3).

DISCUSSION

Although paraganglioma can occur anywhere in the para-aortic region, the tumours are frequently found in the infrarenal area near the origin of the inferior mesenteric artery, where the organs of Zuckerkandl are

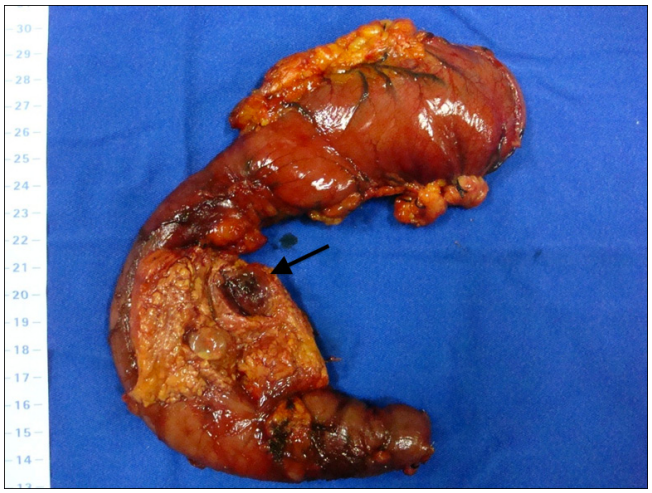

FIGURE 2 – Surgical specimen showing a well delimited lesion with free surgical limits in the pancreas head

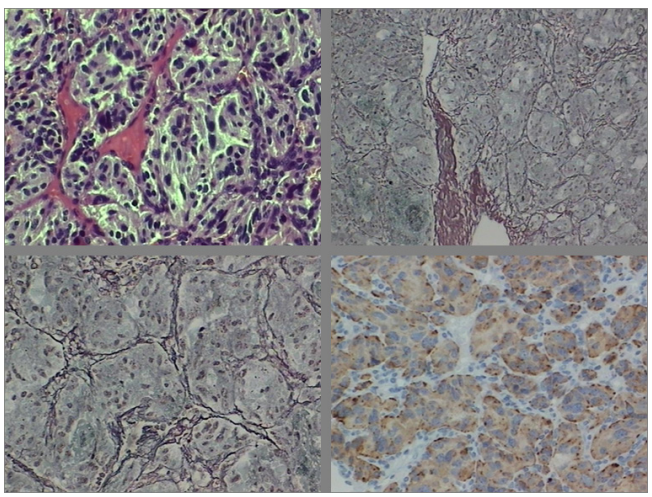

FIGURE 3 – Histopathologic with immunohistochemistry identifying the "Zellballen" pattern

located[6,7,8]. Other less common locations for abdominal paragangliomas include gallblader, urinary bladder, prostate, spermatic cord, uterus, and duodenum. Paraganglioma of the pancreas is extremely rare, and only 14 cases, including the present one, have been reported worldwide[2,3,4,56,7,8]. When the diagnosis of paraganglioma is made, it's necessary screening other tumours, because until now this type of neoplasm has been considered part of multiple endocrine neoplasia syndrome, secundary of genetic alteration predisposing presence of multiple tumors[1].

The mean age of the 15 patients was 63 years, ranging from 41 to 85 years. Five patients were men, and 10, women. In 10 out of the 14 cases, the tumor was located in the head of the pancreas, and 5 were located in the body or tail.

Because paraganglioma of the pancreas is sometimes confounded with pseudocyst or endocrine tumor, it is difficult to obtain a preoperative diagnosis, especially in nonfunctional cases[2,8].

The diagnosis was established by imaging exams revealing a well-defined mass with frequent areas of hypoechogenicity in ultrasonography, a well-marginated, hypervascular tumor with cystic areas of low-attenuation on contrast-enhanced CT[3,8]. Smaller tumors are more likely to show homogeneous attenuation and they can be sharply marginated, as compared to larger ones, demonstrating a rupture of vascular trauma due to the neoplasia and tumoral necrosis[4,6,8].

The majority paragangliomas follow a benign clinical course. However, long-term follow-up studies suggest that the incidence of malignancy may be higher than suspected, especially in certain locations. The treatment of choice for paraganglioma is surgical resection[8]. In this series, the pancreatoduodenectomy occurred in 4 out of the 14 cases, because the lesion proximity to major vessels or the technical impossibility of surgical enucleation[6]. The other patients showed an equally good outcome after simple tumor enucleation[8]. In general, the resection of paragangliomas of the pancreas is technically difficult due to the possibility of a sudden catecholamine release during the operation. Furthermore, this lesion seems to grow slowly and long-term survival can be expected, even when metastasis is present[2].

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