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Nonfunctional paraganglioma of the head of the pancreas: A rare case report

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ABSTRACT

BACKGROUND: Paragangliomas are rare neoplasms that originate from the neural crest. They are malignant in approximately 10% of cases, with a 50% survival rate at 5 years from diagnosis. In most cases, manifestations of malignancy (such as metastasis) are lacking, and paragangliomas are considered benign lesions. Pancreatic paragangliomas are extremely rare, with only 31 cases described in the scientific literature to date.

CASE SUMMARY: Here we describe a case of a 55-year-old Caucasian male patient referred to our institution in September 2013 for lumbar pain lasting five months. The ultrasound and the CT scan revealed a 2.5 cm solid nodule located in the uncinate process of the pancreas. On the basis of this evidence, the preoperative diagnosis was a pancreatic neuroendocrine tumor (NET), which was further confirmed by a subsequent In-Pentetreotide Scan examination. A pylorus-preserving duodenocephalopancreasectomy was performed. Pancreatic paraganglioma was the final pathological diagnosis. Rare localizations of paraganglioma are often discovered casually, during imaging examinations for other clinical reasons, as happened in the case of our patient. It appears evident that the preoperative diagnosis of pancreatic paragangliomas is extremely challenging. Surgery represents the cornerstone of the clinical management of these neoplasms, primarily for the need of a definitive diagnosis, which is difficult to assess preoperatively in most cases.

CONCLUSIONS: Our strategy is the same as that adopted for the management of pancreatic NETs; the dimensional limit for a conservative resection is 2 cm, while major resections (Whipple's approach or distal pancreatectomy) should be employed in larger tumors, which are generally associated with a worse prognosis.

1. Introduction

Paragangliomas are rare neoplasms that originate from the neural crest and involve both the sympathetic and parasympathetic ganglia [1]. They are malignant in approximately 10% of cases, with a 50% survival rate at 5 years from diagnosis. In most cases, manifestations of malignancy (such as metastasis) are lacking, and paragangliomas are considered benign lesions. Pancreatic paragangliomas are extremely rare, with only 31 cases described in the scientific literature to date.

2. Case report

A 55-year-old Caucasian male patient was referred to our institution in September 2013 for lumbar pain lasting five months. Inflammatory, degenerative and rheumatic alterations were excluded by clinical examination, while the clinical examination of the abdomen did not evidence any manifestation of disease. The subsequent ultrasonographic (US) examination revealed a 2.5 cm solid nodule, located in the uncinate process of the pancreas. The lesion was further studied by contrast enhancement ultrasound (CEUS) and computed tomography (CT) scan (Fig. 1A and B). Arterial phase hypervascularity and slow wash-out were observed (Fig. 1C and D), leading to the hypothesis of a neuroendocrine pancreatic tumor (NET), which was further confirmed by a subsequent In-Pentetreotide Scan examination. The patient had no personal or family history of neoplastic diseases. All the biochemical tests returned results within normal ranges, including

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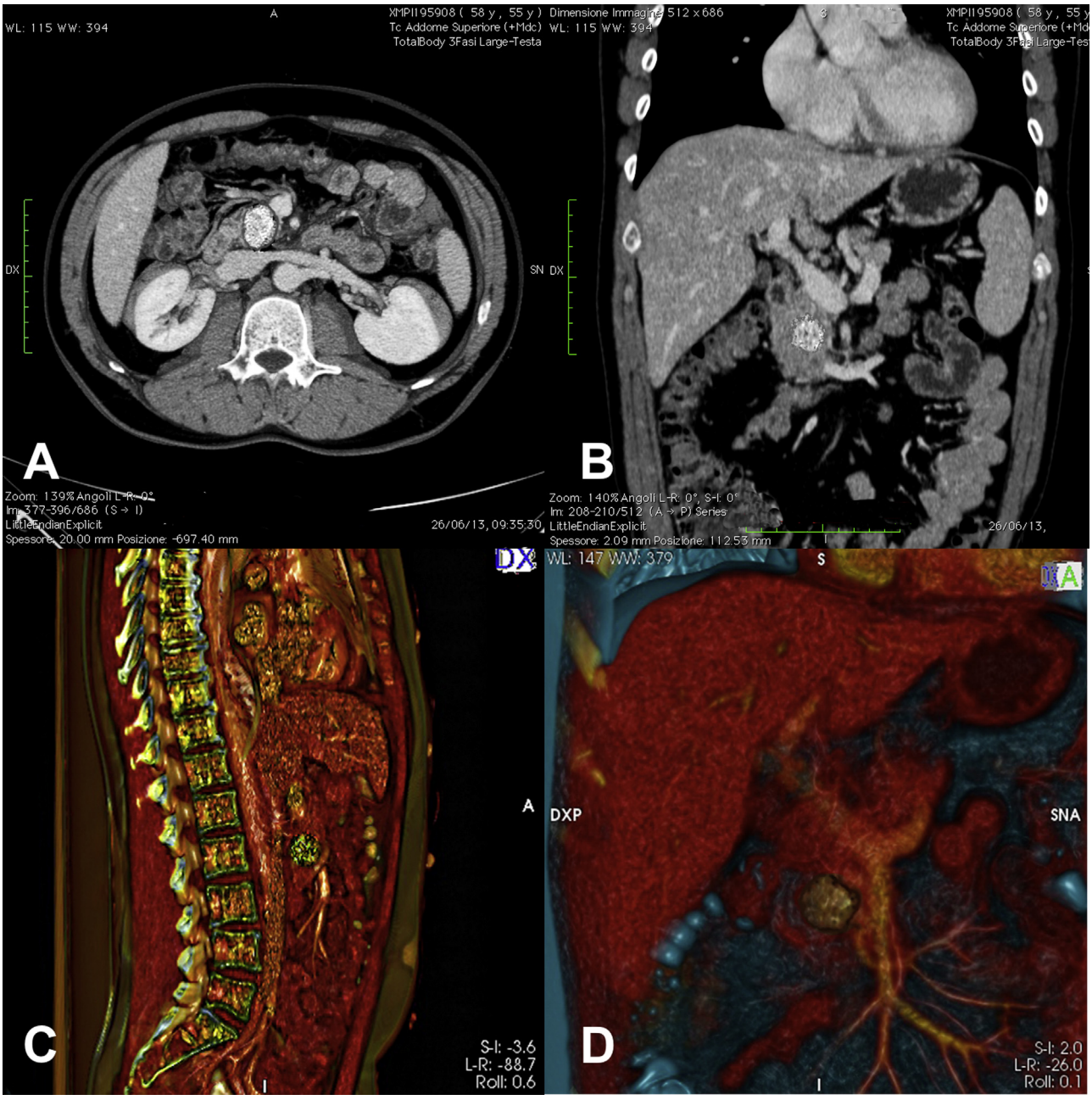

Fig. 1. A–B Computed tomography (CT) scan "arterial phase" showed a hypervascularity and slow wash-out. B–C Computed tomography (CT) scan showed a 2.5 cm solid nodule located in the uncinate process of the pancreas.

chromogranin, blood catecholamines, and urinary metanephrines, as well as serum tumor markers (Ca19-9, CEA, Ca15-3). On the basis of this evidence, the preoperative diagnosis was a pancreatic NET. A pylorus-preserving duodenocephalopancreasectomy was performed in September 2013.

On gross pathological examination the pancreatic lesion measured 1.3 cm, was brownish in color, solid consistency, and well-circumscribed. Microscopically, the lesion was organized in nests of variable dimensions, with numerous blood vessels and hemorrhagic areas (Fig. 2a). The neoplastic cells showed high variability in size and shape. The nuclei were pleomorphic, sometimes very large, and the cytoplasms abundant, finely granular and eosinophilic (Fig. 2b). Strong immunohistochemical positivity for chromogranin A and synaptophysin was observed (Fig. 2c), as well as focal positivity for S100 and somatostatin, while cytokeratins (CAM 5.2) (Fig. 2d) and AE1/AE3 were negative. The final pathological diagnosis was pancreatic paraganglioma.

The patient was discharged on the 15th postoperative day. He underwent a CT scan of the lungs at periods of six months, one year and two years postoperatively, with no evidence of progression of the lung nodule discovered. He is currently alive and disease free. Informed consent for the publication of the present report was obtained; the CARE guidelines were employed for reporting [2].

3. Discussion

Paragangliomas are neoplasms originating from the ganglia of the autonomous nervous system, which usually involve both the