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Retroperitoneal paraganglioma—Is pre operative embolization useful?

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ABSTRACT

INTRODUCTION: Paragangliomas (PG) are rare tumors derived from chromaffin cells that are located outside the adrenal gland and are capable of producing catecholamines. The treatment is based on a surgical resection, and there is controversy regarding the usefulness of previously carrying out an embolization and what is the most adequate surgical approach.
CLINICAL CASE: We will present a 17-year-old woman with a retroperitoneal tumour in contact with the aorta and the inferior vena cava, treated with embolization prior to the surgical resection via laparotomy.
DISCUSSION: The PG tumors are very infrequent and originate in the extra-adrenal chromaffin cells that exist in the vicinity of the components of the autonomic nervous system. Most of them (86%) produce catecholamines, are unique, sporadic, benign and more frequent in middle-aged women. Since they are radioresistant tumors, the only possibility for a cure is by a complete surgical excision. The preoperative embolization has been described mainly as the treatment of cervical PG, although its use in abdominal PG is more controversial and is not done in a systematic manner.
CONCLUSION: We can conclude that the embolization of abdominal PG is not free of risks and that it has not been demonstrated that it significantly reduces the peri-operative bleeding or the surgical time. Probably, the embolization should be reserved for intensively hypervascularized and larger PGs.

1. Introduction

Paragangliomas (PG) are rare tumors derived from chromaffin cells that are located outside the adrenal gland and are capable of producing catecholamines. The treatment is based on a surgical resection, and there is controversy regarding the usefulness of previously carrying out an embolization and what is the most adequate surgical approach. We will present a case treated with embolization before the surgical resection via laparotomy and a review of the existing literature. The present work is reported in line with the SCARE criteria [1].

2. Clinical case

A 17-year-old woman, without any interesting notes, that was assessed due to brief palpitations, hydrorrhea and cephalalgia, without suffering from high blood pressure, which has evolved over one year. The physical exam, the electrocardiogram monitoring, the blood pressure and the treadmill test were normal. The laboratory exams show an increase of the plasma normetanephrine (566 pg/ml), urinary noradrenaline (592mcg/24 h) and urinary dopamine (632 mcg/24 h). The rest of the test were normal, including the oncogene markers and the chromogranin-A. In a computerised tomographic scan (CT) there can be seen a 3 × 5 cm retroperitoneal tumour in contact with the aorta and the inferior vena cava, with a homogenous uptake of the contrast medium (Fig. 1). The magnetic resonance imaging (MRI) of the abdomen shows a 49 × 30 × 25 mm right retroperitoneal nodular lesion at the level of the inferior vena cava, which it compresses, with an increase in signal in the potentiated sequences in T2 and restriction to the diffusion and enhancement with contrast medium (Fig. 2). The study was completed with a scintigraphy with somatostatin receptors (OctreoScan) SPECT CT in which the retroperitoneal mass shows a pathological uptake.

Based on the suspicion raised from the retroperitoneal paraganglioma, the decision to operate on the patient was made, after a two-week of alpha-adrenergic blockade, and beginning the beta-adrenergic blockade 48 h before the operation; while the patient remained hospitalized to carry out a safe embolization of the lesion.

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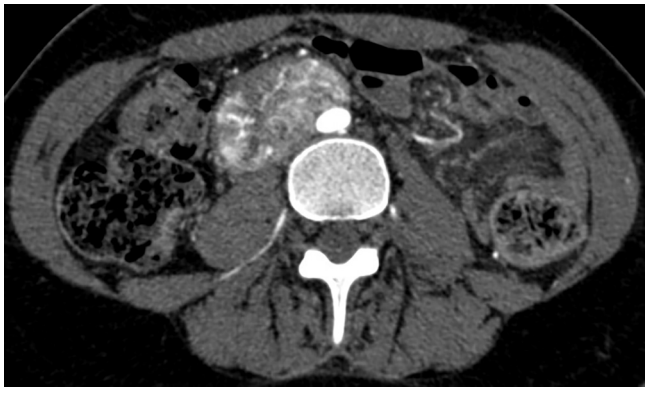

Fig. 1. CT scan showing a 3 × 5 cm retroperitoneal tumour in contact with the aorta and the inferior vena cava, with a homogenous uptake of contrast.

Accessing it through the right femoral artery, a selective angiography of the right lumbar arteries was carried out, being able to complete the vascularization of the tumour by direct branches from the distal third of the aorta (Fig. 3). The arteriography from the upper mesenteric artery shows small arterioles that rise from a branch of the ileocolic artery and irrigate the tumour (Fig. 4). Embolization of the lumbar branches is carried out without subsequent highlighting of the tumour. They did not embolize the arterioles from the ileocolic to prevent a possible ischaemia of the bowel loops. The patient was stable during the procedure and the following hours.

Forty-eight hours later the patient is operated on through a median infra-umbilical laparotomy. There are no peritoneal implants, regional pathologic adenopathies nor ectopic locations of chromaffin tissue. After mobilising the right colon medially a 6 cm retroperitoneal tumour is identified. It had a nodular appearance and a brownish colour and was closely adhered to the distal

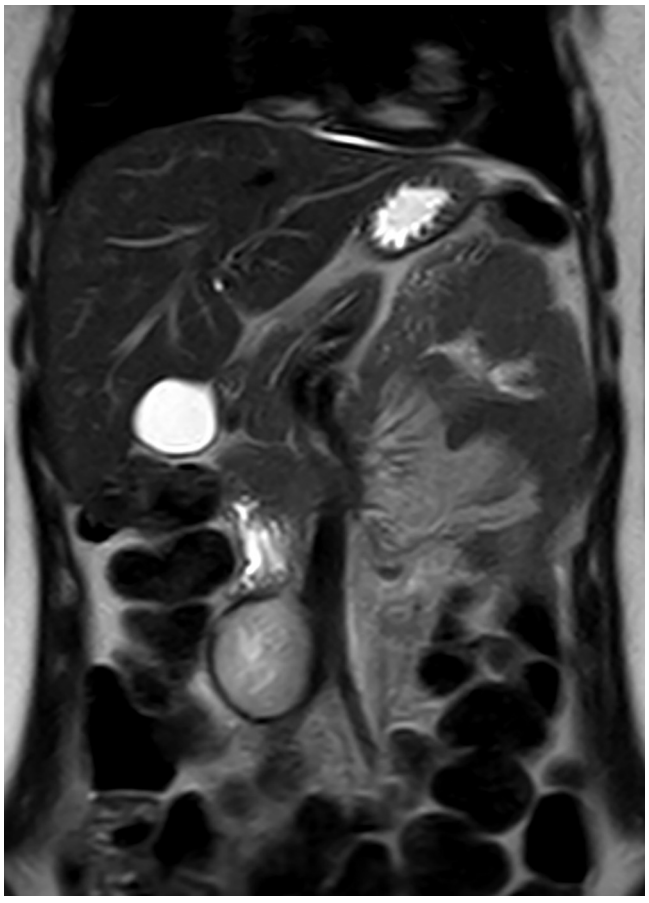

Fig. 2. MRI showing a retroperitoneal node, with an increase in signal in the potentiated sequences in T2 and restriction to the diffusion and enhancement with contrast.

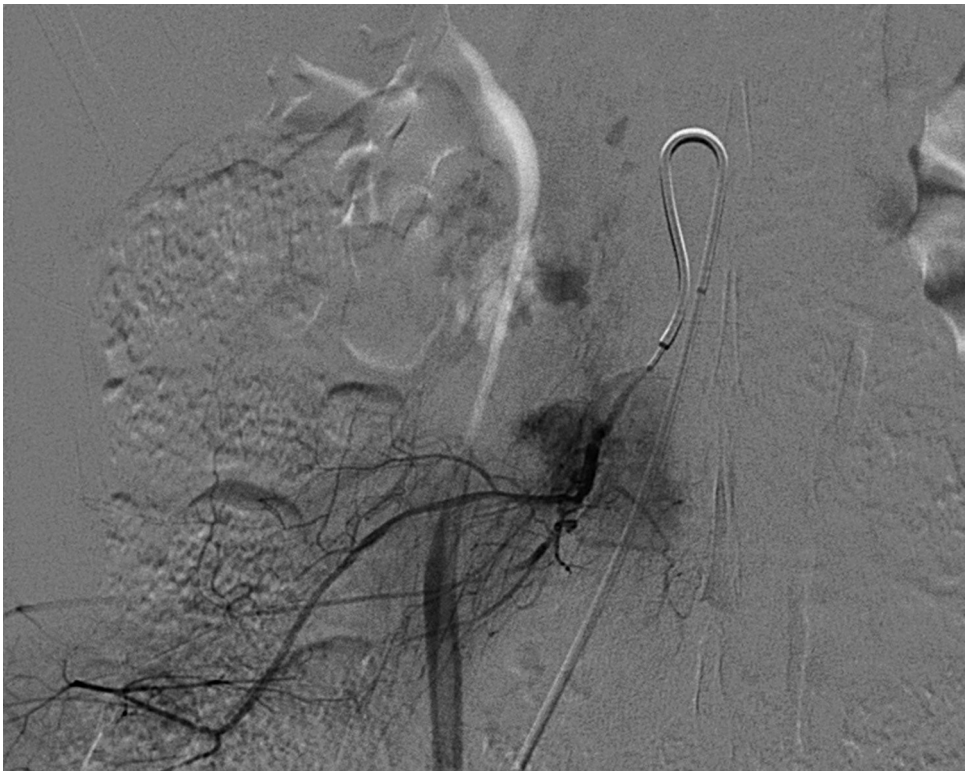

Fig. 3. angiography of the right lumbar arteries.