

# Inter Aortic-Caval Paraganglioma Simulating a Pancreatic Mass - A Case Report

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## Abstract

Paraganglioma is a tumor that develops from clusters of neuroendocrine cells located along the vascular and nervous axes called paraganglia. They are present in different locations: the thorax (posterior mediastinum), the abdomen (in the vicinity of the large blood vessels; aorta and vena cava). Some paragangliomas are secreting, most often catecholamines. They are often asymptomatic and can reach significant dimensions. These tumors are benign in more than 80% of cases and localized. Several recent studies have shown that in one third to one half of cases paragangliomas are associated with a hereditary syndrome. We report in this work a particular case of a patient with an inter aorticocaval paraganglioma simulating pancreatic cancer.

**Keywords:** *Paragangliomas; Ganglions; Catecholamine; Pheochromocytoma; Retroperitoneal masse*

## 1. Introduction

Paragangliomas or extra-adrenal pheochromocytoma, are tumors arising from chromaffin tissues. Abdominal localizations are less frequent than head and neck. Retroperitoneal PGLs are more uncommon, causing considerable difficulty in diagnosis and treatment. This case report describes a patient who initially underwent work up for a suspected pancreatic head mass which was discovered to be a paraganglioma [1,2].

## 2. Observation

Mrs. D. M, 40 years old, has been suffering from right hypochondrial pain and vomiting for four years. These pains appeared after copious meals.

The clinical examination found a conscious, afebrile patient. Blood pressure was 130/70 mmHg in standing and lying position with conjunctiva sub icterus and tenderness of the right hypochondrium.

Abdominal ultrasound showed a distended gallbladder with a thin wall and containing micro lithiasis of variable size not exceeding 6.2 mm, moderately dilated main bile duct measuring 12.6 mm and containing two micro lithiasis measuring respectively 6.4 mm and 5.7 mm with a hepatic pedicle solidocystic formation evoking a priori a hilar adenopathy.

A BILI-MRI showed an expansive process of the head of the pancreas suggestive of a mucinous cyst adenoma with probably signs of degeneration measuring 41 cm × 33 cm compressing the vena cava posteriorly with dilatation of the main bile duct and slightly enlarged Wirsung and a micro lithiated gallbladder. The workup showed alkaline phosphatase at 4 times normal, gamma-GT at 5 times normal and conjugated bilirubin at 3 times normal.

The diagnosis of pancreatic head process was retained. The median approach revealed an extra-pancreatic mass between the aortic and the vena cava, compressing the inferior vena cava and pushing the pancreas forward. Intraoperative manipulation caused blood pressure peaks of 22/11 mmHg, and the diagnosis of paraganglioma was suspected, a complete resection of the mass was performed without touching pancreas and duodenum. The anatomopathological study confirmed the diagnosis and concluded that it was a well-differentiated paraganglioma.

### 3. Discussion

Primary retroperitoneal neoplasms are rare and benign [3], Paragangliomas are extra-adrenal pheochromocytoma which originate from fine Chroma cells of the sympathetic system, the latter are located in the retro peritoneum or in the thorax, or from the parasympathetic system (aorta, main vessels) [4].

Paragangliomas metastasize in 20% to 50% of cases (malignant forms) [5], which are higher than that of pheochromocytoma, which are malignant in 10% of cases [6,7].

Functional paragangliomas secrete noradrenaline and Normetanephrine and account for 30-60% of tumors [8]. Clinically, these tumors manifest as paroxysmal episodic hypertension, as well as the typical triad of symptoms associated with pheochromocytoma: palpitations, headaches and profuse sweating, in which case the diagnosis is made by measuring catecholamines [9].

In the case of non-functional paragangliomas, it is most often an isolated pain or a retroperitoneal abdominal mass. The diagnosis of paraganglioma should be made in the presence of any isolated retroperitoneal mass in order to take the necessary precautions to avoid serious and often fatal complications.

A large proportion of these tumors are discovered incidentally in normotensive patients during an imaging workup done for other reasons. The CT scans before and after injection is of major interest. It confirms the extra-adrenal origin when the tumor volume is still moderate and can specify the single or multiple nature of the lesion, the loco regional invasion and confirm malignancy in the case of capsular rupture [10].

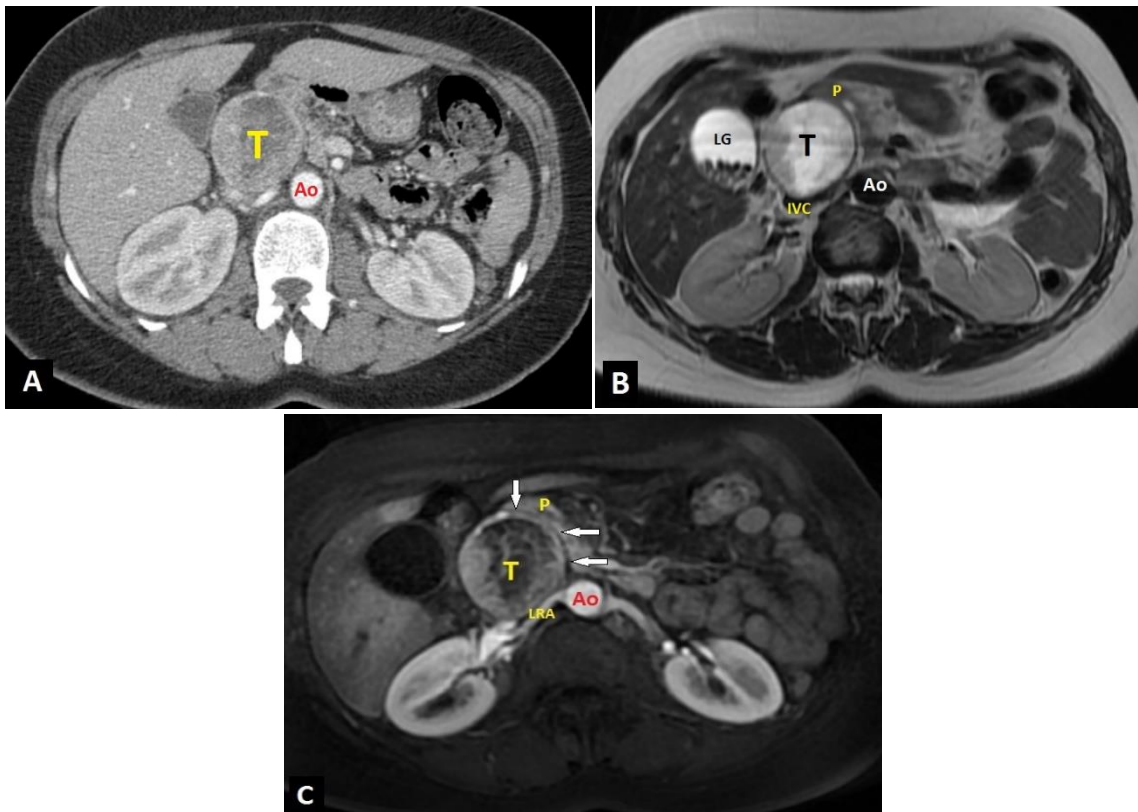
As for magnetic resonance imaging (MRI), it detects an intermediate weak signal in T1 spin echo and a marked hyper signal in T2 spin echo, which is reinforced in the second echo, which would be very suggestive [11].

Surgery is the mainstay of treatment, and total excision is necessary to cure the disease, although it may also be necessary to extend the disease to adjacent organs. The possibility of radical surgery is estimated at 75% of cases [12].

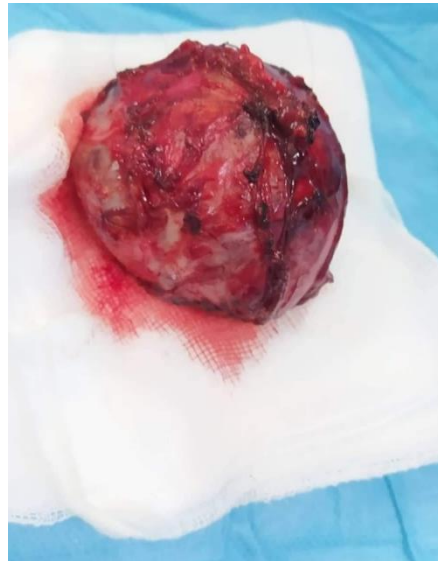
The role of MIBG scintigraphy in malignant pheochromocytoma and paragangliomas is crucial. Indeed, by establishing complete lesion topography, it guides the therapeutic course of action [13].

All in all, retroperitoneal paragangliomas are rare tumors, most often benign and with A good prognosis, but which may present a loco regional invasion with a metastatic potential. Clinically these tumors are very atypical especially when it comes to non-functional masses.

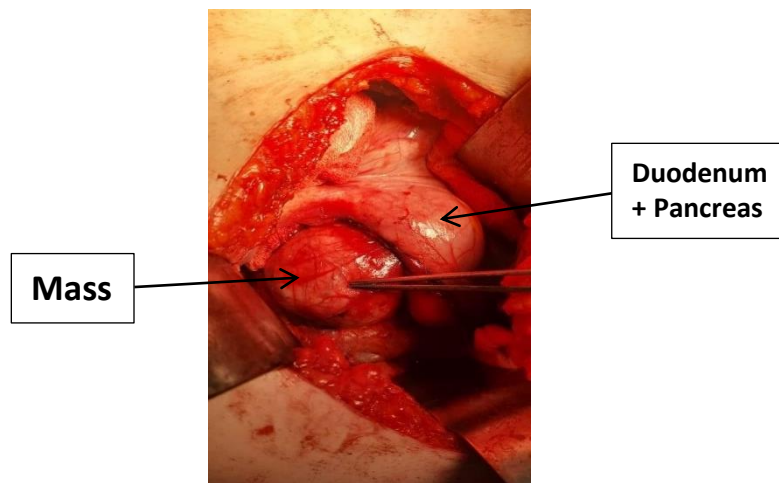
The diagnosis of paragangliomas is based on the dosage of catecholamines, followed by diagnostic imaging (a CT or even an MRI) in order to locate the primary lesion. Surgery remains the best weapon and is the mainstay of treatment as long as the mass is resectable (FIG. 1-3).



**FIG. 1. CT scan (A) and MRI (B,C) showing a paraganglioma which was surrounded by aorta, inferior vena cava, and left renal artery. A cleavage plane is noted between the pancreas and the mass (arrows). T: tumor, Ao: aorta, IVC: inferior vena cava, LRA: left renal artery, P: pancreas, LG: lithiasis gallbladder.**



**FIG. 2. Inter aortico-caval paraganglioma.**



**FIG. 3. Intraoperative inter aortico cava paraganglioma.**

#### **4. Conclusion**

The clinical presentation of paragangliomas can be atypical and misleading, it is necessary to know how to evoke the diagnosis of paragangliomas in front of any isolated retroperitoneal mass in order to undertake the necessary precautions to avoid serious complications often mortal. Surgical removal is the treatment of choice, with additional therapies being used mainly for symptomatic purposes. Their benign or malignant nature can only be confirmed by long-term follow-up, based on CT scan, MRI and MIBG scintigraphy, which can detect recurrence or metastases.

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