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Inter Aortic-Caval Paraganglioma Simulating a Pancreatic Mass

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1. Summary

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) [1]. 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 [2]. We report in this work a particular case of a patient with an inter aorticocaval paraganglioma simulating pancreatic cancer.

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, apyretic patient. Blood pressure was 130/70 mmHg in standing and lying position with conjunctival subicterus 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 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 cystadenoma with probably signs of degeneration measuring 41x33 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 and confirmed by anatomopathological study, which concluded that it was a well-differentiated paragangliom.

3. Discussion

Primary retroperitoneal neoplasms are rare and benign [3], Paragangliomas are extra-adrenal pheochromocytomas which originate from fine chroma cells of the sympathetic system, the latter are located in the retroperitoneum 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 is higher than that of pheochromocytomas, 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 pheochromocytomas: 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 tumours are discovered incidentally in normotensive patients during an imaging workup done for other reasons.

The CT scan before and after injection is of major interest. It confirms the extra-adrenal origin when the tumour volume is still moderate and can specify the single or multiple nature of the lesion, the locoregional 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 pheochromocytomas and paragangliomas is crucial. Indeed, by establishing a complete lesion topography, it guides the therapeutic course of action [13].

All in all, retroperitoneal paragangliomas are rare tumours, most often benign and with a good prognosis, but which may present a locoregional 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 (Figure 1 and 2).

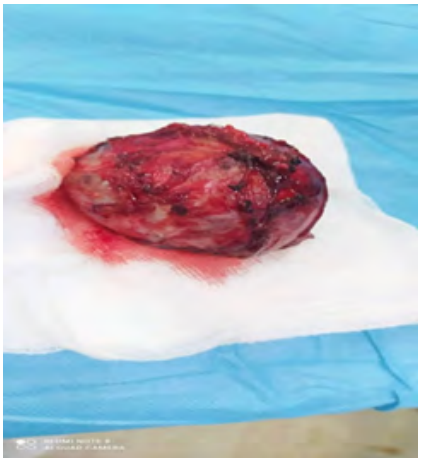

Figure 1: Inter aortico-caval paraganglioma

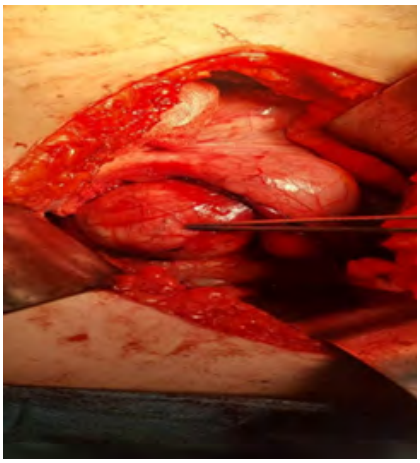

Figure 2: Intraoperative inter aortico cava paraganglioma

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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