

Diagnosis of a Retroperitoneal Paraganglioma by Endoscopic Ultrasound: A Case Report

M.Kadiri¹, M.Salihoun¹, M.Acharki¹, F. Zoiadia² and N.Kabbaj¹

¹Department of Gastroenterology "EFD-HGE", Hospital Ibn Sina Rabat, Morocco.

²Anatomopathology Department, Ibn Sina University Hospital, Rabat.

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ABSTRACT

Retroperitoneal nonfunctioning paraganglioma are rare tumors. They are defined as an extra-adrenal chromaffin tumors and represent approximately 1/5th of chromaffin tumors. They can develop in contact with the pancreas, whose puncture or surgical resection can lead to serious complications. They are most often asymptomatic, and can reach important dimensions. We report a case of a 49-years-old female patient who was admitted to the hospital for epigastric pain and vomiting. CT-scan showed a posteriorly developing mass. The diagnosis of retropancreatic paraganglioma, was made by EUS-guided fine needle aspiration(FNA), this diagnosis was initially taken for a pancreatic lésion at endoscopy ultrasound(EUS). Complete surgical excision was performed after specific anesthetic preparation and anatomopathological study of the biopsied material. The treatment of paraganglioma is surgical, but management must be multidisciplinary. It is important to search whenever there is a juxtapancreatic tumor, arguments that can guide to the diagnosis of paraganglioma and use reliable means of diagnosis to confirm it.

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Introduction

The paragangliomas, or extra-adrenal pheochromocytomas, are neuroendocrine tumors developed at the expense of the parasympathetic nervous system. They are defined as extra-adrenal chromaffin tumors that comes from the neurovegetative system and represent approximately 1 / 5th of chromaffin tumors.

Retroperitoneal nonfunctioning paraganglioma are rare tumors, and are less frequent than other locations (head, neck). They are often asymptomatic and can reach important dimensions. It is important to make the diagnosis before any invasive action because of the risk of severe hemodynamic or hemorrhagic disorders during surgical resection. We report a case of a retroperitoneal retro-pancreatic paraganglioma in a 49-years-old woman with a literature review.

Case report

A 49-year-old woman with past medical history of arterial hypertension and well-controlled asthma. For 2 months, she presented ongoing epigastric pain, associated with bilious vomiting in a context of apyrexia and general state preservation.

At admission to hospital, the patient was conscious, hemodynamically stable (TA=120/80 mmHg, Hr 74 bpm), with a normal Body Mass Index (23kg/m²). Our patient had not any jaundice or pruritus. The abdominal examination shows the presence of a hard epigastric mass, fixed on the deep plane. The biological assessment revealed an anemia at 7.7g/dl associated with a biological inflammatory syndrome (C-RP :255 mg/l) and an hypoprotidemia. Furthermore, the tumor markers Ca19-9 and CEA were normal.

Radiologically, the abdominal CT scan showed a retroperitoneal mass measuring 48*42*29mm; However, X-ray Imaging did not determine whether this mass was at the expense of the pancreas or not.



Figure 1. radiologic images showing the retroperitoneal mass at CT scan.

The endoscopic ultrasound examination (EUS) revealed a solid heterogeneous hypoechoic tumor in the uncus of the pancreas, measuring 33.7*46.9 mm, well limited, close to the vascular pancreatic axis without invading them. The transgastric endoscopic ultrasound fine needle aspiration (FNA) diagnosed a paraganglioma.

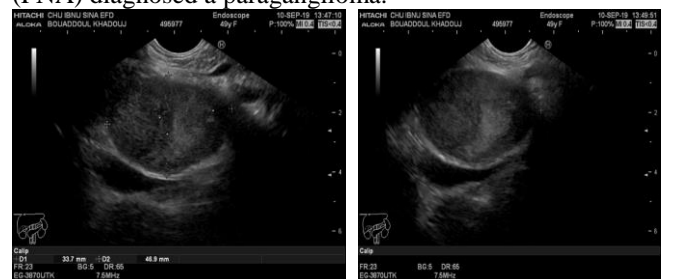


Figure 2. EUS images of a retropancreatic paraganglioma.

Tele:

E-mail address: maryeme.kadiri@gmail.com

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The patient was subsequently operated, and underwent complete resection of the tumor without pancreatic resection. Surgical exploration showed a retropancreatic tumor, presenting a single intimate contact with the pancreas within an area of 1cm.

The histopathological examination of the operative specimen concludes in a tumor measuring 5*2cm with tumor proliferation forming a neuroendocrine architecture, and make the diagnosis of paraganglioma, whose excision was complete.

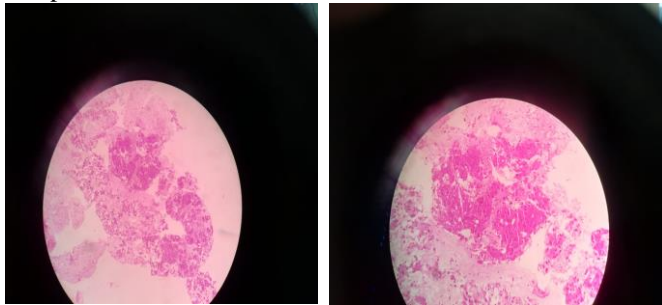


Fig 3. The anatomopathological analysis of material obtained by FNA biopsy showing the paraganglioma.

Discussion

Paragangliomas are rare tumors that develop from chromaffin cells and secrete catecholamines. These tumors develop most often in the adrenal medulla and then take the name of pheochromocytoma. Chromaffin cells are also found in sympathetic ganglia. These paraganglions are distributed symmetrically from the base of the skull to the bladder in close association with the autonomic nervous system. Other preferred sites for paragangliomas are retroperitoneum, carotid glomus, jugular foramen, middle ear, aortopulmonary region, and bladder wall (1, 2). Paragangliomas can occur at any age with a predominance in the fourth and fifth decades of life as the case of our patient. They are rarely multiple or bilateral (10%), rarely malignant (10%), this malignant potential seems more important for retroperitoneal localizations (20 to 42%).

→Clinical data:

Classical clinical manifestations include arterial hypertension, which may be episodic or refractory, affecting approximately 85% of subjects. The hypersecretion of catecholamines is responsible for headaches, palpitations and cardiovascular manifestations (myocardial infarction, cerebral haemorrhage or malignant hypertension) that may be life-threatening. About 10% of paragangliomas are clinically silent and incidentally detected by imaging performed for another pathology (2). Clinically silent tumors, tend to be larger in size.

→Biological data:

The diagnosis of paraganglioma is strongly suspected of an elevation of urinary methoxamines: metanephrines and normetanephrines. Their threshold values range from 1000 to 2880 nmol/24 h for metanephrine and from 3000 to 6550 nmol/24 h for normetanephrine, which makes it possible to define the areas of unlikely diagnosis below these thresholds, possible diagnosis between these thresholds, and very likely above. The sensitivity of this assay is 96.9% (3), the test being negative for the rare non-secreting paragangliomas. Metaiodobenzylguanidine scintigraphy (MIBG-I131) may show hyperfixation even in the absence of elevation of

urinary metoxyamines. It has a sensitivity of 90%. It can locate the tumor and detect another location. Finally, it can be used to define the neoplastic nature of the tumor by detecting metastases defined by the involvement of a site where there is normally no residual embryonic enterochromaffin tissue. The sensitivity of urinary methoxamine dosing combined with MIBG scintigraphy would be 100%.

→Morphological data:

The abdominal CT scan shows the characteristics of the tumor including the retroperitoneal seat, the size, whether it's a single or multiple tumor, locoregional and remote invasion, the most common aspect is a solid round or oval mass, homogeneous, but can be cystic or necrotic at its center, or calcified (4,5,6). However, there is no specific tomodensitometric criterion of paraganglioma which makes it possible to differentiate it from other retroperitoneal (neurofibroma, sarcoma) and also pancreatic (adenocarcinoma, neuroendocrine tumor) tumors. In fact, the location of the tumor in our patient misled the initial diagnosis on imaging.

The MRI shows a hypo-intense signal in T1, strongly reinforcing on the second spin echo sequence in T2. MRI, however, does not provide a benefit compared to CT in the characterization of paraganglioma (7). Imaging is especially interested to clarify the relationship of the tumor with the adjacent organs to guide the surgical procedure.

Endoscopic ultrasonography makes it difficult to determine whether the tumor is intrapancreatic or extrapancreatic (8), which is the case in our case report, but has made it possible to make the diagnosis of certainty by needle aspiration; Histology confirms the diagnosis of paraganglioma, but does not define whether the tumor is benign or malignant. Indeed, only the appearance of a distant new localization will retrospectively make the diagnosis of malignancy.

→Therapy data :

Surgery is the basis of treatment with radical resection in 75% of cases. The choice of the surgical procedure between the conventional and laparoscopic pathway, remains very controversial considering the undesirable side effects of laparoscopy. Complementary therapies such as chemotherapy and external radiotherapy, could find their place in metastatic forms with a positive response in about 50% of patients, however, without significantly influencing the prognosis. Only surgical excision allows a significant improvement, with a 75% recurrence-free survival rate at 5 years and 45% at 10 years (9). the average survival is 3 years in metastatic forms and 4 years in case of incomplete resection (9).

Conclusion

The paragangliomas, are a rare neuroendocrine tumors developed at the expense of the parasympathetic nervous system. They are often asymptomatic and can reach important dimensions. Endoscopic ultrasound-guided fine needle aspiration (EUS-FNA) has emerged as an effective tool in the diagnosis of these lesions. The assessment of anatomically adjacent structures by EUS may provide useful information before surgical resection (10). In the present case, the main differential diagnosis was a pancreatic lesion. Nonetheless, EUS-FNA was performed uneventfully and this most unusual diagnosis was possible.

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