Renal hilar paraganglioma: A case report

Zeinab Ali Abou Yehia, Rashid Khalid Sayyid, Ali Ahmad Haydar

Zeinab Ali Abou Yehia, Rashid Khalid Sayyid, Ali Ahmad Haydar, Department of Radiology, AUBMC, Riad El Solh, Beirut 1107 2020, Lebanon

Abstract
Paragangliomas are extra-adrenal pheochromocytomas that derive from chromaffin cells and arise along the sympathetic paraganglia in the body. In the majority of cases, they are secretory tumors and most commonly present with palpitations. Plasma metanephrines are the standard screening tests for making the diagnosis which is confirmed by pathology. Imaging plays a very important role in establishing the diagnosis. However, there is no specific feature on imaging for paragangliomas; the vascularity of the tumor should show as hyper-enhancing lesions but this is not always the case. The diagnostic value of PET is yet a matter of debate. We present a very rare case of a paraganglioma arising at the renal hilum, splaying the renal artery and vein and causing vascular compromise to the left kidney. The patient presented with an atypical presentation of unrelenting fever that was followed by acute colicky pain. Based on imaging and blood metanephrine levels, the diagnosis of paraganglioma was made. Resection of the tumor was achieved and the patient is now asymptomatic.

INTRODUCTION
Paragangliomas comprise a rare category of chromaffin cell tumors. They are extra-adrenal pheochromocytomas usually arising along the sympathetic paraganglia in the body. The rarity of these tumors can easily mislead the pre-operative diagnosis, leading to a drastic outcome especially when it arises in atypical locations. There is no specific well described feature on imaging for paraganglioma; the vascularity of the tumor should reveal a hyper-enhancing mass but this is not always the case. Despite premedication, a significant percentage of patients experience considerable intraoperative hemodynamic complications; it is therefore essential to have a high index of suspicion in making the diagnosis of a paraganglioma in the light of atypical tumor location and unspecific symptoms.

CASE REPORT
The patient is a previously healthy 32-year-old woman. She presented with a 10 d history of headache, nausea, vomiting and generalized weakness, followed by acute onset colicky renal pain and low grade fever. Physical
examination was non-remarkable except for tachycardia (HR: 130-140).

Patient had leukocytosis (WBC: 17000), elevated inflammatory markers (CRP: 11.485 mg/dL, normal: 0-1 mg/dL) and the other tests were unremarkable. CT abdomen revealed a 3.3 cm × 3.3 cm × 2.5 cm mass with hypodense center and ring enhancement at the renal hilum splaying the renal artery and the renal vein causing vascular compromise to the left kidney with secondary focal lower pole infarction (Figure 1).

Blood metanephrine level was then obtained and found to be elevated (0.88; normal: less than 0.37). PET-CT revealed radiotracer uptake at the periphery of the mass (Figure 2). CT guided FNA was done and showed atypical epithelioid malignant cells.

The patient was pre-medicated with an alpha blocker and underwent surgery four days later for a suspected extra-adrenal pheochromocytoma. Total left nephrectomy and excision of the mass was done. Pathology revealed a paraganglioma (Figure 3).

Her post-operative course was uneventful. Her headache disappeared completely and her pulse returned to normal, despite discontinuation of the beta blocker.

**DISCUSSION**

Paragangliomas usually present along the sympathetic chain in the abdomen, particularly in the vicinity of the aorta and less commonly in the thorax or head and neck. They can be functional-secreting catecholamines or nonfunctional. Paragangliomas are secretory in more than 50% of cases, the majority of which present with palpitations.

Other typical symptoms include episodic hypertension, headache and diaphoresis. Paroxysmal hypertension is seen only in 50% of these patients. Unspecific presenting symptoms such as fever and lumbar pain have been reported with cases of retroperitoneal paragangliomas. The laboratory workup of patients with pheochromocytomas and extra-adrenal paragangliomas (PPGLs) has conventionally relied on biochemical measurements of tumor secretory products or their metabolites. Histopathology and immunohistochemistry remain the gold standard for making the definitive diagnosis.

Approximately 10% of pheochromocytomas and paragangliomas are malignant; nevertheless, this often cannot be determined on a biochemical or histological basis. Malignancy in these tumors is defined by the presence of local invasion on gross or microscopic examination at the time of resection, or much more commonly by the presence of metastases.

Our patient presented with a 10 d history of headache and fever, followed by acute onset colicky renal pain. She was tachycardic but her blood pressure was normal on all measurements taken. The acute pain, in association with fever, headache, nausea and vomiting, together with the hypodense mass in the kidney, led us to consider an infectious ongoing process.

Plasma free metanephrine and normetanephrine or urine metanephrine are the first screening tests to make the diagnosis of a paraganglioma. They are functional-secreting catecholamines or nonfunctional. Paragangliomas are secretory in more than 50% of cases, the majority of which present with palpitations.

Plasma free metanephrine and normetanephrine or urine metanephrine are the first screening tests to make the diagnosis of a paraganglioma. Our patient had elevated plasma metanephrine level with no elevation of normetanephrine. Imaging studies such as CT or magnetic resonance imaging are essential before surgery for confirmation and for further assessment of the location, size and possible metastasis. PET-CT provides precise anatomic and metabolic information and recently has become increasingly relied upon. In the initial evaluation of pheochromocytomas, it was validated that PET-CT complements other modalities. However, the value of

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**Figure 1** Axial computed tomography scan of the abdomen in the porto-venous phase at the level of the kidney. Black double-arrow head points to hypodense tumor with ring enhancement at the renal hilum. White arrow shows the displaced renal vein; black arrow shows the compressed renal artery. White double-arrow head shows anterior renal infarct.

**Figure 2** Positron emission tomography computed tomography coronal view. Arrow head pointing to tumor showing peripheral radio-tracer uptake.

**Figure 3** Pathology slide, arrow pointing to synaptophysin positive cells.
Treatment
The patient was pre-medicated with an alpha blocker and then 4 d later under-
grew left nephrectomy and excision of the mass was performed.

Related reports
The Joynt et al article “Paragangliomas, etiology, presentation and management” published in 2009 provides a brief but cumulative overview on the case topic.

Term explanation
A paraganglioma is a rare tumor that originates from chromaffin cells of sympathetic chain; it can be found anywhere along the sympathetic chains in the body.

Experiences and lessons
One lesson that the authors learned from this case is to consider paraganglioma in the differential when a patient presents with vague symptoms and an abdominal mass.

Peer review
The manuscript presents a rare case of a renal hilar paraganglioma demonstrating the steps to making the diagnosis and it provides some literature review regarding the management and follow up of such a rare tumor. The case may not add much to the existing literature but it certainly highlights an original substance to learn from.

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