Case Report

Abdominal Ultrasound in the Detection of an Incidental Paraganglioma

CME Credits

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Abstract

Paraganglioma is a tumor that originates from neuroendocrine cells of the sympathetic or parasympathetic systems. Patients may suffer from headaches, palpitations, diaphoresis, and hypertension due to catecholamine excess or symptoms from the mass effect of the tumor. In the absence of typical symptoms of catecholamine excess, the diagnosis of a nonfunctional paraganglioma is often delayed. Herein, we report a case of a 63-year-old female patient with a nonfunctional paraganglioma which is an accidental finding during investigation of a fever. Abdominal ultrasonography incidentally detected this lesion as a complex, solid, cystic mass in the left suprarenal retroperitoneum.

Keywords: Paraganglioma, retroperitoneum, ultrasound

Introduction

Incidentaloma is a mass lesion that is discovered incidentally during a radiologic examination. Because of the widespread use of abdominal ultrasonography (US), computed tomography (CT), and magnetic resonance imaging (MRI), such imaging findings have been increasingly frequent.[1-3] "Incidentaloma" is used to describe a wide and heterogeneous spectrum of pathologies, including benign or malignant tumors, hormonally active or inactive lesions, metastases, infections, granulomas, infiltrations, cysts and pseudocysts, and hemorrhages.[1] We present the case of nonfunctional retroperitoneal paraganglioma discovered during investigation of a fever.

Case Report

A 63-year-old female with a history of hypertension under medication control for 3 years went to the emergency department of our hospital because of fever and chills for 2 days. She went to the emergency department of our hospital because of fever and chills for two days. Physical examination disclosed a soft abdomen without flank knocking pain. Blood examination showed elevated C-reactive protein (14.8 mg/dL; normal < 0.7 mg/dL). Urinalysis was positive for leukocyte esterase and bacteria. The remaining systemic inquiry was unremarkable. There was no leukocytosis at presentation.

Abdominal US was performed to evaluate the urinary system and showed a mass lesion with cystic contents in the left upper retroperitoneum behind the pancreas and just lateral to the abdominal aorta [Figure 1]. Bilateral kidneys showed no hydronephrosis. The patient underwent abdominal CT that confirmed a mass lesion in the left retroperitoneal location that was very close to the left adrenal gland [Figure 2]. An incidentaloma was considered.

Antibiotic treatment was prescribed for the urinary tract infection, fever, and chills subsided. One month later, the patient underwent laparoscopic surgery, and the retroperitoneal incidentaloma about 6.4 cm × 5.5 cm was excised totally. Neither ascites nor retroperitoneal lymph node was noted. The left adrenal gland was free of the tumor. The pathology showed a paraganglioma [Figure 3]. The postoperative course

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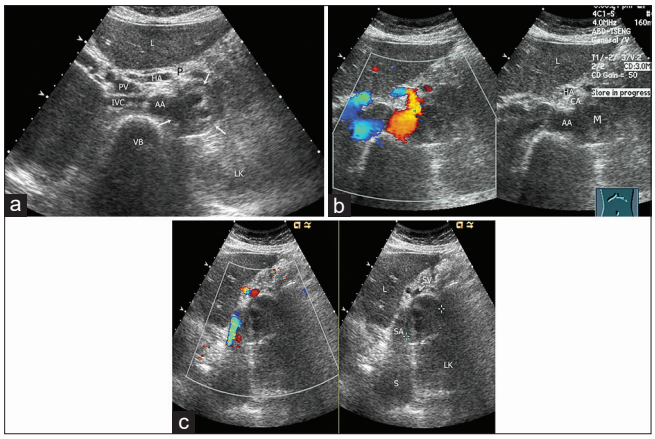

Figure 1: A retroperitoneal incidentaloma in a 63-year-old female patient with a urinary tract infection. (a) Transverse sonogram of the upper abdomen shows a mass lesion (arrows) with central cystic change in the left upper retroperitoneum, behind the pancreas (p). (b) Color Doppler (left panel, velocity setting of 53 cm) and corresponding gray-scale (right panel) transverse sonograms at the level of the celiac artery (CA) show the mass (m) in the left lateral to the abdominal aorta (AA). The mass lesion is thought to be in the left adrenal fossa. No detectable Doppler flow signal within the lesion. (c) Color Doppler (left panel) and corresponding gray-scale (right panel) longitudinal sonograms reveal that the mass (marked by + cursors) is posterior to the splenic vein (SV) and caudal (distal) to the splenic artery (SA) and between the liver (l), spleen (s), and left kidney (LK). HA, hepatic artery; IVC, inferior vena cava; PV, portal vein; VB, vertebral body

was uneventful. There was no tumor recurrence at the 2-year follow-up on CT.

Discussion

Paraganglioma is a tumor that originates from the neuroendocrine cells of the sympathetic or parasympathetic systems.[4-6] These tumors can occur at the skull base, neck, chest, and abdomen.[7] When the lesion occurs in the abdomen, the most common site is the adrenal gland, and it is then specifically called a "pheochromocytoma." The term, "paraganglioma," refers to a tumor that is outside the adrenal gland.[8]

The classical presentation of a paraganglioma is excess catecholamine production.[6,9,10] Paragangliomas produce, store, synthesize, and metabolize catecholamines.[11] A value three times the upper range of normal is a positive result.[6] However, because of fluctuating release of catecholamines, a false negative may occur in the presence of low catecholamine level.[11] Urine and plasma metanephrines, which are the metabolites of catecholamines, last more consistent in the body, so measurement of these metabolites in plasma is now considered the more accurate test.[12,13] There is another biochemical marker called chromogranin A (CgA) that is used in patients with paraganglioma. CgA is a polypeptide commonly secreted by chromaffin cells, typically with catecholamines.[14] In the absence of typical symptoms of catecholamine excess, diagnosis of a nonfunctional paraganglioma is often delayed.[15,16]

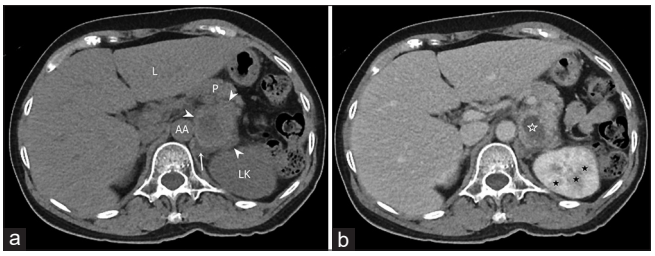

Figure 2: The transverse views of nonenhanced (a) and contrast-enhanced (b) computed tomography scan confirm the location of the mass (arrowheads) behind the pancreas (p) and left lateral to the abdominal aorta (AA). The mass is composed of an enhancing solid part peripherally and a nonenhancing cystic part centrally (open star). The mass is very close to the left adrenal gland (arrow) and the fat plane between them is not clearly visible. Note that some areas with poor contrast enhancement in the left renal parenchyma (stars) are consistent with acute pyelonephritis, which is not obvious on the sonogram. L, liver; LK, left kidney

Diagnostic imaging plays a critical role in paragangliomas [Table 1]. On US imaging, paragangliomas in the neck usually appear as hypoechoic, well-defined, inhomogeneous, hypervascular masses in the area of the carotid bifurcation, which separates the internal and external carotid arteries.[17,18] However, there are few reports of US imaging of abdominal paragangliomas. Hashimoto *et al*. revealed eight abdominal paragangliomas (size range: 2.3–11.9 cm; median: 6.5 cm) and showed that six of eight were in contact with the inferior vena cava or pancreas.[19] They found that all eight lesions had distinct boundaries with near-spherical (six of eight), polygonal (one of eight), or irregular (one of eight) shapes. Half of the lesions (four of eight) had predominantly cystic components, and seven of eight showed blood flow signals. In our case, the tumor was close to the pancreas and exhibited a well-defined margin and cystic change. We did not detect Doppler flow signals within the lesion, which may have been because of the high-velocity scan setting (53 cm) for which a high-flow vessel, such as the abdominal aorta [Figure 1b], is not contaminated with aliasing artifacts, but the sensitivity to low blood flow within a tumor declines.

On contrast-enhanced CT, these tumors appear as paraaortic soft-tissue masses with either homogeneous contrast enhancement or central areas of low attenuation, as in our case.[20] Compared with larger tumors, smaller ones are more likely to be homogeneous in attenuation and sharply marginated. Punctate calcification or focal areas of high attenuation caused by acute hemorrhage are seen in some tumors.[20] On MRI, these tumors are usually hypointense or isointense compared with the liver parenchyma on T1-weighted images and are markedly hyperintense on T2-weighted images.[21-23] Either CT or MRI images may exhibit similar appearances between functioning and nonfunctioning paragangliomas.[21]

The diagnosis of malignant paraganglioma has stringent criteria according to the World Health Organization (WHO)

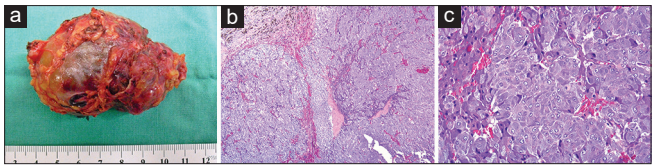

Figure 3: Histopathology of the paraganglioma. (a) Gross view of the specimen shows a yellowish to brownish elastic and encapsulated mass. (b and c) Histopathological views show the paraganglioma composed of sheet, zellballen growth of epithelioid, polygonal, tumor cells with amphophilic, finely granular, cytoplasm, and centralized nuclei with conspicuous nucleoli(b: [H and E, ×40] and c: [H and E, ×100]). The immunostains (images not shown) show synaptophysin (+), chromogranin (+), and Melan A (−) in tumor cells. The S100 highlights sustentacular cells. The PHH3 shows a mitotic rate <1 mitosis/10 HPF in tumor cells

Table 1: Imaging characteristics of paragangliomas

Image modality	Finding
US	Distinct boundaries, various shapes (near-spherical in the majority) , presence of blood flow signals, with or without cystic change
CT	Sharply marginated, punctate calcification, with or without hemorrhage, homogeneous contrast enhancement in small tumors; central necrosis in large tumors
MRI	Hypointense or isointense to liver parenchyma T1-weighted images; markedly hyperintense on T2-weighted images

classification in 2004 and requires evidence of metastases at nonchromaffin sites distant from the primary tumor, such as lymph nodes, bones, liver, and lung.[24] However, the WHO classification changed in 2017. Currently, the term metastatic paraganglioma has replaced malignant paraganglioma.[25,26] Previous reports have shown that the malignant rate of paragangliomas range from 7% to 50%[27-29] and tumor malignancy can be diagnosis at the first presentation or during follow-up. In addition, Hamidi *et al*. reported that approximately 21% of malignant paragangliomas are nonfunctional tumors.[30] The clinical, biochemical, and radiological features alone are inadequate to predict malignancy.[10] Some biochemical data are thought to be a factor to predict malignancy, but they remain controversial.[31-33]

Concerning treatments for paraganglioma, patients with biochemically active paraganglioma should immediately be placed on antihypertensive medications to control symptoms and reduce the risk of hypertensive crises. After antihypertensive medications, treatment options are surgical resection, radiofrequency ablation, radiotherapy, chemotherapy, and molecular-targeted therapies; however, surgical resection remains the only curative treatment for patients with paraganglioma.[11,29,34] A previous report described apparently benign disease that returned with metastases ≤15 years after resection,[27] so life-long follow-up of patients with resected paragangliomas is imperative.[29]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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