Paragangliomas are rare tumors arising from extraadrenal chromaffin cell and accounting for 10~18% of all chromaffin tissue-related tumors and one tenth of pheochromocytomas. Herein we present a case of malignant retroperitoneal paraganglioma with lung metastasis and the computed tomography (CT) findings. Approximately 10% of paragangliomas are discovered on imaging study for evaluation of unrelated symptoms. The combination of radiological findings and clinical characteristics may be helpful to diagnose retroperitoneal tumors, including paragangliomas and other neurogenic tumors and, to determine the local extent of tumor. Preoperative suspicion of paraganglioma mandates biochemical screening and prevention for perioperative hypertensive crisis.

**Key words:** Paraganglioma; Retroperitoneal space, CT; Retroperitoneal space, Neoplasms

**CASE REPORT**

A 79-year-old man was admitted due to right flank soreness and previous history of a right retroperitoneal tumor of unknown nature for 5 years. He also had a history of hypertension under medical control for 15 years, chronic hepatitis C, and peptic ulcer disease with post subtotal gastrectomy status. No headache, cold sweating, paroxysmal palpitation or other family history of malignancy was noted. During the admission, abdominal CT revealed a 10 × 10 × 8 cm heterogeneous mass with strong peripheral enhancement, and central non-enhanced low-density implying cystic change or central necrosis (Fig. 1a, 1b). Encasement of inferior vena cava (IVC), bilateral renal veins and portal vein with splenorenal collateral circulation was also depicted (Fig. 1b, 1c). Abdominal sonography with Doppler study showed one large heterogeneous echoic mass lesion mixed with soft tissue and cystic components. Hypervascularity and displacement of adjacent great vessels was also depicted in the Doppler study (Fig. 2). Under the impression of retroperitoneal tumor of unknown nature, he received exploratory laparotomy, in which only tumor biopsy was done due to severe adhesion to IVC and left renal vein. Pathologic diagnosis was paraganglioma with positive immunohistochemical stain for chromogranin, synaptophysin and S-100. During the hospital course, there was no adverse hemodynamic change after intravenous injection of iodine-containing contrast medium or during operation. Consequently, palliative radiotherapy (4500 cGy) was done. Eight months later, however, routine follow-up abdominal CT showed one 4 × 2cm mass at the lower lobe of right lung (Fig. 1d),
suggesting lung metastasis. According to the clinical behavior, malignant retroperitoneal paraganglioma was diagnosed.

**DISCUSSION**

Tumors arising from chromaffin cells of the sympathetic nervous system are classified into two subsets—pheochromocytomas arising from adrenal medulla and paragangliomas occurring in the extraganglionic sites. The latter can arise from the sympathetic ganglia at the neck, in the mediastinum, along the aorta, in the organ of Zuckerkandl, and in the pelvis or urinary bladder [1, 4]. Most of them occur in the adult with mean age of 47 years and with no sex predilection [2, 3]. When they occur in the child, the possibilities of associated familial syndromes should be considered, including type 2 multiple endocrine neoplasia syndromes, von Hippel-Lindau disease, neurofibromatosis, Carney’s syndrome, Sturge-Weber disease and tuberous sclerosis [1, 4]. In the inherited forms, most of them are bilateral [1-4].

At gross examination, paragangliomas are tan-red, firm, encapsulated masses, adhering adjacent structures and ranging from 1–6 cm in diameter, with malignant tumors tending to be slightly larger [1, 4], just like our case. Central necrosis occasionally occurs in large tumors [1]. Microscopic analysis demonstrates

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**Figure 1.** a. Non-enhanced CT scan at the level of the orifice of superior mesentery artery shows one 10x10x8cm soft tissue mass with central low-attenuation area at right paraaortic retroperitoneum. Large central low-attenuation cystic change implies extensive necrosis of tumor. b. and c. Contrast-enhanced CT scans at the same level of a. and at the level below a. show avid peripheral enhancement of tumor mass with persistent central low-attenuation area. Displacement and suspicious severe adhesion to adjacent great vessels, including abdominal aorta, inferior vena cava and bilateral renal veins are found. Prominent splenorenal shunt is also depicted (arrow in Fig. 1b) d. Eight months later, about 4 months after completion of radiation therapy, contrast-enhanced CT scan at the similar level shows one 4 × 2 cm nodular lesion at lower lobe of right lung, implying a metastatic lesion despite no pathologic diagnosis.
that the neuroendocrine cells arranged in clusters called zellballen and interspersed with fibrovascular stroma, which produces strong contrast enhancement in the CT or MR imaging [1]. Specific antibodies from neuroendocrine markers such as synaptophysin, chromogranin and S-100 protein can be used to confirm the diagnosis [1]. All of them were positive in our case.

The clinical manifestations include paroxysmal or persistent hypertension, throbbing headache, diaphoresis, anxiety, tremor, nausea, vomiting, chest or abdominal pain, visual disturbance and postural hypotension. They are related to excess catecholamine production by the tumors, such as norepinephrine, epinephrine and dopamine [1, 4]. The classic “triad” of headache, palpitation and sweating in a hypertensive patient was found to have a sensitivity of 91% and specificity of 94% [2]. However, about 8% of patients may be completely asymptomatic [2]. Thereafter, certain percentage approximately 10%, of pheochromocytomas and paragangliomas were discovered incidentally at imaging studies or after operation [2, 3, 5]. In the elderly, a special diagnostic challenge exists because of the decreased baroreceptor sensitivity with age, and more concomitant diseases with overlapping symptoms which can confound the diagnosis [1, 2]. The diverse manifestations of the tumors reflect variations in the hormones they release and the patterns of release, and in the individual-to-individual differences in catecholamine sensitivity. The most important thing in the management of pheochromocytomas and paragangliomas is simply to seriously consider them in the differential diagnosis.

For patients with symptoms related to excess catecholamine production, it is important to confirm or to exclude the presence of hyperactive catecholamine-secreting tumors by measuring catecholamines and their related metabolites in the plasma and urine. The available tests include plasma catecholamines, 24-hour urine fractioned metanephrines, 24-hour urine catecholamines, serum chromogranin-A, and plasma free metanephrine [1-4]. Among these tests, plasma free metanephrine is considered the most sensitive [2]. However, no single analysis can achieve 100% accuracy because pheochromocytomas are heterogeneous group of hormone-secreting tumors [2]. Care should be taken when interpreting the results of these tests because several medications and foodstuffs may produce false-positive results, including the beta-blockers which may be prescribed for patients with presumptive diagnosis of essential hypertension [1, 2, 4]. On the other hand, high urinary metabolites of catecholamines with near normal plasma catecholamines may be present in the patients with large cystic tumors, which mainly secret metabolized catecholamines. In the equivocal cases, pharmacological tests such as stimulation or depression tests may be considered even they are now seldom used [2, 4].

Biochemical confirmation of the diagnosis should be followed up by radiological evaluation to locate the tumor. Pheochromocytomas are referred as “10% tumors”—10% are bilateral, 10% are extraadrenal, i.e. paragangliomas, 10% are in the children and 10% are malignant [1]. Moreover, approximately 10% of paragangliomas are discovered at imaging examination for evaluation of unrelated symptoms [2, 3, 5]. Being
familiar with the radiological findings and clinical characteristics are helpful in the differential diagnosis of retroperitoneal tumors, including paragangliomas and other neurogenic tumors, and in the determination of the local extent of tumor for appropriate treatment planning. Preoperative suspicion of paraganglioma mandates biochemical screening and prevention for perioperative hypertensive crisis. On CT images, there are no specific imaging finding for paragangliomas [1, 4, 6]. They are usually large tumors with areas of hemorrhage or necrosis and avid enhancement after contrast medium injection [1, 4, 6]. Sometimes fluid-fluid level is found [4]. Because of the risk to incite a hypertensive crisis after intravenous injection of contrast medium, it is a common policy at many institutions that only non-enhanced CT with peroral contrast medium is done for request to “rule out” pheochromocytomas or paragangliomas [1, 4, 6, 7]. On the other hand, enhanced CT scan is helpful in detection of small tumors and detail evaluation of local invasion. If enhanced CT is crucial, preventive medication with alpha- and beta-blocker is recommended for the reason of safety [1, 4]. Mukherjee et al [7] reported no statistically significant increase in catecholamine level after intravenous injection of non-ionic contrast medium in 10 patients with pheochromocytomas as compared to the control subjects and they suggested that non-ionic iodinated contrast medium may be used safely without premedication in patients with suspicious pheochromocytomas or paragangliomas. However, in our case, there was no adverse reaction from ionic contrast medium enhanced CT scan.

MR imaging has been considered as a sensitive and safe imaging modality for patients with pheochromocytomas or paragangliomas in tumor localization especially when the tumors are functional [1, 4]. Multiplanar imaging also offers a more comprehensive demonstration of the relationship between the tumors and adjacent structures such as kidneys, inferior vena cava and aorta [4]. In most cases, they are hypointense or isointense on T1-weighted images (T1WIs), marked hyperintense on T2-weighted images (T2WIs) and strongly enhanced after gadolinium injection [1, 4-6]. Marked hyperintensity on T2WIs may be essential for differential diagnosis of adrenal tumors because most of other adrenal tumors tend to be isointense [1]. However, it should be kept in mind that significant minority up to 30% of these tumors do not have such typical appearance [1, 4]. Tumors with hemorrhage show the typical MRI features of hemorrhage of hyperintensity on T1WIs at acute or subacute stages [4]. The imaging features are not reliable to distin-

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惡性副神經節瘤：病例報告

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副神經節瘤為罕見的原發於腎上腺外嗜鉻細胞的腫瘤，約佔所有原發於嗜鉻細胞腫瘤的10～18%；發生率也只有腎上腺嗜鉻細胞瘤的十分之一。本文藉由腹部電腦斷層的發現，我們提出一位罕見的惡性副神經節瘤合併有肺部轉移之病例報告。由於副神經節瘤的臨床表現非常多樣化且缺乏專一性，因此約有十分之一的副神經節瘤是在影像學檢查中意外發現的。因此熟悉副神經節瘤的影像學表現及其可能的臨床特徵，將有利於後腹腔腫瘤的鑑別診斷及腫瘤局部侵犯的評估。若術前有考慮到副神經節瘤的可能性，則應該針對其代謝產物做詳細的生化學檢驗及針對術中可能發生的髙血壓危象做預防性的藥物投與。

關鍵詞：副神經節瘤；後腹腔，電腦斷層；後腹腔，腫瘤