Rare Case of Reninoma with Double Inferior Vena Cava

Yan Jiang,1 Lian Duan,1 Lin Lu,1 Wei-Gang Zhao,1 Zheng-Pei Zeng,1 Han-Zhong Li,2 Xiao-Bo Zhang3

1 Department of Endocrinology, Key Laboratory of Chinese Health Ministry, Peking Union Medical College Hospital, Peking Union Medical College, Chinese Academy of Medical Science, Beijing, China
2 Department of Urology, Peking Union Medical College Hospital, Peking Union Medical College, Chinese Academy of Medical Science, Beijing, China
3 Department of Radiology, Peking Union Medical College Hospital, Peking Union Medical College, Chinese Academy of Medical Science, Beijing, China

Abstract
A 16-year-old boy suffered from headaches and dizziness for 2 years. He was found to have remarkably elevated blood pressure (BP) of 180/110 mmHg. Laboratory findings showed a low level of serum potassium and markedly increased plasma renin activity. A solid mass at the periphery of the right kidney and double inferior vena cava (IVC) were detected by abdominal computer tomography (CT). Right partial nephrectomy via laparoscopy was performed on the patient. The histologic and electron microscopic findings confirmed a diagnosis of juxtaglomerular cell tumor. The patient had no headache or dizziness with normal BP after surgery.

Keywords: reninoma, double inferior vena cava

INTRODUCTION
Reninoma is a renal juxtaglomerular cell tumor which is a rare cause of hypertension. There were only about 89 cases of reninoma that have been reported over the past 40 years (1). Vascular anomalies of vena cava are uncommon with the prevalence rate of 0.5 percent to 3 percent (2, 3). To our knowledge, the co-existence of reninoma and double inferior vena cava (IVC) have not been reported up to now. We presented the first case of reninoma with double IVC.

CASE REPORT
A 16-year-old boy suffered from headaches and dizziness for 2 years. He was found to have remarkably elevated blood pressure (BP) of 180/110 mmHg. Although he was administered therapy of felodipine and metoprolol, his BP was 150/90 mmHg. His biochemical evaluations revealed that serum potassium was 3.4∼3.7 mmol/L, 24-h urinary catecholamines were within the normal range. Stenosis of renal artery was ruled out by a renal arterial Doppler study. A solid mass at the periphery of the right kidney and double IVC (Figure 1) were showed in abdominal computer tomography (CT). Vasography of IVC also demonstrated double IVC (Figure 2). The peripheral plasma renin activity (PRA) was markedly elevated for more than 12 ng/ml h (normal range 0.05∼0.79 ng/ml.h) and plasma aldosterone concentration (PAC) was normal (11.86 ng/dl, normal range 5.9∼17.4 ng/dl). Selective renal vein sampling was performed to define the function of this lesion, which showed symmetric renal vein PRA (34.5 ng/ml.h in the left renal vein, 28.5 ng/ml.h in the right vein, and 22.8 ng/ml.h in the periphery vein).

Right partial nephrectomy via laparoscopy was performed on this patient. The histologic examination demonstrated a well-encapsulated, grey-pink solitary lesion measuring 3 cm without hemorrhage or cystic degeneration. Hematoxylin eosin (HE) staining showed uniformly round cells with indistinct cell borders and a moderate amount of eosinophilic cytoplasm (Figure 3). Immunostaining for smooth muscle actin (SMA) was positive in these cells. Electron microscopic studies revealed characteristic rhomboid-shaped renin protogranules and irregular rounded renin-containing granules in the cytoplasm of tumor cells (Figure 4). The histologic and electron microscopic findings supported the diagnosis of juxtaglomerular cell tumor.
Figure 1. Computed tomography scan showing a solid, well-circumscribed mass lesion in the right kidney (white arrow) and two rounded structures of almost equal size, on each side of aorta. This is double IVC (black arrows).

Figure 2. Vasography of IVC showing double IVC.

The operation was successful without any damage to renal function. One week following surgery, his BP was approximately 125/80 mmHg without any antihypertensive agents. After 2 weeks, his serum potassium was 4.6 mmol/L, peripheral PRA returned to a normal range of 0.4 ng/ml.h, and PAC was 8.9 ng/dl. At his 4-month follow-up, the patient’s BP and serum potassium remained in normal range. The patient no longer had headache or dizziness.

Figure 3. HE staining: uniformly round cells with indistinct cell borders and moderate amount of eosinophilic cytoplasm (color figure available online).

Figure 4. Electron microscopic photograph showed characteristic cytoplasmic rhomboid-shaped rennin protogranules and irregular rounded rennin-containing granules ($\times 15000$).

COMMENTS

Reninoma is a renal juxtaglomerular cell tumor that produces excessive amounts of renin, resulting in secondary hyperaldosteronism. It is a rare form of secondary hypertension. It occurs primarily in children and young adults with a mean age of 27 years. The most common presenting symptom is headache (1). Hypokalemia is not a necessary finding for diagnosis. However, all patients had elevated plasma renin activity. Most of the patients had elevated PAC levels, but in a few cases, PAC levels were reported as normal. This case showed normal PAC level.

The criteria for defining PRA lateralization during renal vein sampling are unclear. Previous studies cited that a renin lateralizing ratio of 1.5 was useful for predicting tumor location. In this case, segmental sampling of PRA did not demonstrate the lateralization.
Since performing renal vein sampling in a double IVC patient is very difficult, it may affect taking blood samples from each renal vein. Chuang et al. (4) also showed blood flow carried by the left vena cava diluted the left renal vein sample. Therefore, double IVC may affect the results of samplings. Another possible reason for the symmetric renal vein PRA from previous studies is that the tumor is usually small and located on the surface of the kidney and most of the tumor blood is collected in the peripheral veins and not in the renal vein. Therefore, renal venous sampling for renin assay is not mandatory due to high false negative results (5).

Anomalies of inferior vena cava are uncommon. Duplication of the IVC occurs as the result of persistence of either the right or left supracardinal veins or the subcardinal veins. There were a few reports of unilateral aplasia of the kidney associated with IVC abnormalities. It has been postulated that the developing kidneys profoundly affected the formation of the IVC (6, 7). Some case reports also revealed that renal cell carcinoma combined with double IVC (8, 9). However, the association of double IVC with reninoma has not been reported. Lack of preoperative recognition of retroperitoneal venous anomalies can have potentially disastrous consequences. Intraoperative trauma may cause a life-threatening hemorrhage. Thrombus may extend into the anomalous vena cava. Extension of renal cell carcinoma into duplications of the IVC has also been reported (8–10). Computed tomography is the most reliable method for detecting tumor localization and venous anomalies in retroperitoneum. Because reninoma in this patient was a benign lesion, and at the periphery of the right kidney, the anomalous vascular structure did not present any difficulty at surgery.

In conclusion, we report a case of reninoma with double IVC. Partial nephrectomy via laparoscopy led to a cure of reninoma. Recognition of venous anomalies is important in the evaluation and surgical treatment.

Declaration of interest: The authors report no conflict of interest. The authors alone are responsible for the content and writing of the paper.

REFERENCES

学霸图书馆

www.xuebalib.com

本文献由“学霸图书馆-文献云下载”收集自网络，仅供学习交流使用。

学霸图书馆（www.xuebalib.com）是一个“整合众多图书馆数据库资源，提供一站式文献检索和下载服务”的24小时在线不限IP图书馆。

图书馆致力于便利、促进学习与科研，提供最强文献下载服务。

图书馆导航：
图书馆首页 文献云下载 图书馆入口 外文数据库大全 疑难文献辅助工具