A Rare Case among Hypertensive Patients: How Do We Handle Reninomas?

Hiroshi Okuda, Michiaki Abe and Hideyasu Kiyomoto

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Juxtaglomerular cell tumors (JCT), also known as reninomas, were first reported over 50 years ago (1). These extremely rare tumors can cause severe hypertension via their hyper-secretion of active renin (2). The afflicted patients, who are commonly adolescents or young adults, usually show malignant hypertension with hypokalemia. A pre-operative diagnosis of JCT is difficult, although a high plasma renin activity (PRA) without any renal arterial stenosis or occlusions can be indicative of the disease. Imaging studies (enhanced CT/MRI) and selective venous catheterization are usually performed to define the function of the tumor. However, selective renal vein sampling is not always successful in elucidating the root causes of increases in PRA, and may be affected by the anatomical variations of the renal veins. In a review of 47 published case reports, 85 renal vein catheterizations were preformed, but a pre-operative determination of the presence of lateralization could not be made in 26 of these cases (1). The lateralization of the tumor can be confirmed if the lateralization ratio (LR) is greater than 1.5, because the sensitivity and specificity were 56 and 94%, respectively, for an LR of 1.5 based on 50 previously reported renal vein catheterizations (2, 3). In order to support an accurate diagnosis by renal vein sampling, an oral administration of an angiotensin-converting enzyme inhibitor or a diuretic is necessary to improve the sensitivity of the catheterization procedure. These agents enhance renin secretion and increase LRs via the negative feedback of the renin-angiotensin system.

In this issue of Internal Medicine, Osawa et al. report a rare case of JCT with secondary hypertension, where the cause of the hypertension had been unknown for 17 years (4). This case report is an excellent example of the successful use of selective renal venous sampling in proving the existence of the small tumor. In addition, they also confirmed the characteristics of tumor as a functional JCT based on the combination of a saline infusion test, a furosemide-upright test and a captopril challenge test.

Reninoma is a rare disease, but it should be considered as a possible diagnosis when physicians observe malignant hypertension in young adults or adolescents. In order to diagnose this condition accurately, dynamic enhanced CT and selective renal venous sampling should be performed prior to any surgical intervention.

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References