LETTER TO THE EDITOR

Paraganglioma in the renal pelvis

Dear Editor,

Paraganglioma is a rare tumor originating from the primitive neural crest, and can develop at many locations, such as the abdomen, aortic bifurcation, neck, pelvis, and thorax. However, the renal pelvis is an extremely rare location for paragangliomas to occur. Here, we report a case initially suspected to be a renal mass in a 59-year-old patient with paroxysmal hypertension, heart palpitation, and sweating. However, the mass was pathologically confirmed to be a paraganglioma in the renal pelvis. This report describes its rarity as well as its clinical and therapeutic characteristics.

The patient, a 59-year-old Chinese woman, had a 2-month history of paroxysmal hypertension, heart palpitation, and sweating. Her blood pressure measured 137/77 mmHg, and the remaining physical examination results were normal upon admission. Her blood metanephrine and normetanephrine levels were 322 ng/L and 632 ng/L, respectively, and other laboratory test results were normal. The left renal mass, measuring 4 cm in diameter, was detected by a computed tomographic (CT) scan without border differentiation, and enhanced CT results showed images of enhanced kidney only but not the mass. It was difficult, however, to determine the origin of the mass. The patient then underwent a preoperative preparation with α and β blockers for 2 weeks. During the surgery, we determined that both the mass and the left kidney had the same origin; therefore, radical excision of the left kidney and the tumor was performed. Paraganglioma in the renal pelvis was diagnosed pathologically, with Pan Cytokeratin (PCK) being negative, whereas synaptophysin, chromogranin A, and S-100 were positive immunohistochemically (Fig. 1). The patient’s postoperative recovery was without complications. During 3 months of follow-up, her symptoms were controlled and blood catecholamine levels were within normal limits.

Paragangliomas can be classified as either functional or nonfunctional, depending on catecholamine levels. Functional paragangliomas may be detected early if clinical presentations or excess catecholamine is recognized, whereas nonfunctional paragangliomas are difficult to identify preoperatively until they cause symptoms [1]. Preoperative α and β blockers are therefore necessary when paragangliomas are suspected.

The primary symptoms of paragangliomas are headaches, hypertension, palpitation, and sweating. Catecholamine test results are required, whereas CT, magnetic resonance imaging, and ultrasonography can detect tumor locations and metastasis. Iodine-131-meta-iodobenzylguanidine scanning can also detect and treat paragangliomas.

There are a few English language reports on renal hilar paragangliomas. Chandra et al. [2] reported one case of renal hilar paraganglioma complicated with renal artery stenosis. Ahallal et al. [3] reported a 58-year-old woman with renal hilar pheochromocytoma; and Bhandarkar et al. [4] reported two patients with renal hilar pheochromocytomas treated with laparoscopic resection successfully. Melegh et al. [5] reported a case of giant cystic pheochromocytoma in the renal hilum; however, there are few reports on renal pelvic paraganglioma. Radical removal of the kidney or tumor is the optimal surgical procedure to treat paragangliomas in the renal pelvis. The criteria of malignancy cannot be easily distinguished histologically, but a metastasis in follow-up is a sign of malignancy [3].

In conclusion, paraganglioma in the renal pelvis is extremely rare, and mass with hypertension or increased catecholamine levels should lead to suspicion of paraganglioma. We presented this case to demonstrate the
clinical and therapeutic characteristics of this extremely rare tumor.

References


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