A 55-year-old woman had a right renal tumor incidentally diagnosed by ultrasonography. CT revealed a perirenal low density mass 3 cm in diameter. Dynamic CT showed peripheral enhancement of the tumor in early phase and internal homogeneous enhancement in delayed phase. Since hemangioma was considered most likely, we performed tumor resection and spared the right kidney. The tumor was supplied by the superior ureteral artery from the right main renal artery which was considered to be derived from the renal sinus. The tumor was diagnosed as intravascular papillary endothelial hyperplasia (Masson’s tumor). This is the first report of intravascular papillary endothelial hyperplasia existing in the perirenal space. Although preoperative diagnosis of intravascular papillary endothelial hyperplasia is difficult, intra-operative pathology and kidney-sparing treatment should be considered in such a case.

Key words: kidney – intravascular papillary endothelial hyperplasia – Masson’s tumor – perirenal – renal sinus

INTRODUCTION

In 1923, Masson (1) described a bizarre endothelial proliferation including anastomosing vascular channels with endothelium-lined tufts and papilla-like formations extending into the lumen of a blood vessel and termed it ‘vesiant intravascular hemangioendothelioma’. Henschen interpreted it as a reactive process rather than a neoplasm (2). The term intravascular papillary endothelial hyperplasia (IPEH) was first used by Clearkin and Enzinger (3). IPEH was classified into two subtypes; a mixed form, papillary endothelial hyperplasia changing focally within hemangioma, and a pure form without a vascular lesion.

In general, the lesions are most commonly located within veins on the head, neck, fingers and trunk, where they appear as small, firm, superficial masses imparting a red to blue discoloration to the overlying skin. There are about 200 well documented cases of IPEH; however, only two cases involving the urinary tract have been reported. We report a case of IPEH considered to be derived from the renal sinus.

CASE REPORT

A 55-year-old woman had a right renal tumor incidentally found by ultrasonography. The patient was asymptomatic and both physical examination and routine evaluation were unremarkable.

Ultrasoundography showed an isoechoic, partially hypoechoic, mass of the right kidney (Fig. 1). CT revealed a mass measuring 3 cm in diameter with homogeneous low density structure. Dynamic CT showed peripheral enhancement of the tumor in early phase and internal homogeneous enhancement in delayed phase (Fig. 2). T1 weighted magnetic resonance imaging (MRI) displayed a hypointense tumor mass compared to the renal parenchyma and T2 weighted MRI displayed a mass of great hyperintensity. Coronal MRI view showed an indistinct upper margin against the renal parenchyma (Fig. 3). Selective angiography showed that the tumor was supplied by a supra-ureteral artery and showed marginal fragmented stains which spread gradually to the entire tumor.

From these findings, renal hemangioma was considered; however, we could not exclude the possibility of renal cell carcinoma. Therefore, operative removal of the tumor and rapid histological tissue examination was planned. The tumor was connected to the renal sinus and supplied through the right superior ureteral artery from the right renal artery, and a vein from the tumor went into the renal sinus. Histological tissue diagnosis, using frozen material, revealed hemangioma and the right kidney was spared.
Macroscopically, the resected tumor was well-demarcated and was $2.7 \times 2.0 \times 1.6$ cm. It was partly encapsulated by fibrous connective tissue which was thought to be a vessel wall. The cut surface of the lesion was protruding, soft and colored dark red (Fig. 4). Microscopically, the tumor was composed of proliferating endothelium without significant pleomorphism or mitotic figures (Fig. 5). In addition, pericytes could be identified on the antiluminal aspects of the endothelial cells. The central area was relatively acellular and composed of degenerated and edematous amorphous tissue. The lesion was partly surrounded by a fibrous pseudocapsule containing residual smooth muscle and elastic tissue of the preexisting vessel wall, but in some parts of the boundary the proliferating endothelium was directly attached to surrounding connective tissue. The tumor was diagnosed as IPEH.

DISCUSSION

IPEH may occur at any age and has a slight female preponderance. Most pure forms have been found in the dermis or subcutis of the fingers, head and neck area, extremities or trunk (3,4), though visceral locations are extremely rare. Only two cases of IPEH relating to kidney have been reported. Gerber et al. reported the first IPEH case in the kidney in 1990 (5) and Steffe et al. reported it in a thrombosed renal allograft vein (6).

As visceral IPEH is extremely rare, there is no consensus about the radiological findings of IPEH. In the present case, radiological examinations suggested hemangioma. Differential diagnosis of IPEH from hemangioma is difficult to make radiologically. Generally, hemangiomas may mimic abscess, hematoma or even adrenal mass in ultrasonography. Both CT and MRI findings are nonspecific when differentiating hemangiomas from other benign or malignant tumors (5). In hemangiomas MRI usually displays as hypointense on T1 weighted images and hyperintense on T2 weighted images and there is a marked signal increase after gadopentetate dimeglumine application (7,8). Most hemangiomas display a characteristic enhanced pattern; they show peripheral enhancement of the tumor in the early phase and internal homogeneous enhancement in the late phase (9).

IPEH is believed to be a primary endothelial proliferation occurring in response to inflammation and stasis in a vascular bed. Most of the lesions are thought to be unusual forms of
Figure 3. Coronal view of early phase dynamic MRI (T1-weighted) shows peripheral enhancement. The margin is unclear to the right kidney at the upper pole of the tumor.

Figure 4. The tumorous lesion is 2.7 × 2.0 × 1.6 cm, had dark red compact cellular areas and myxomatous areas on its cut plane. It was partly encapsulated by fibrous connective tissue considered to be vessel wall.

Figure 5. The tumorous lesion grew from the venous endothelium without significant pleomorphism or mitotic figures. (a), Elastica stain; (b), HE.

organizing thrombus. The excised tissue has usually been described as a gray-tan or purplish red multicystic mass that contained hemorrhagic or clotted material surrounded by more or less distinctive fibrosis. It is an exuberant intravascular endothelial proliferation that in many respects mimics an angiosarcoma. The most distinctive aspect was the presence of endothelial-lined papillary structures within a vascular space. Thrombus and papillary structures were usually found in the center of a well-defined area of endothelial cells. Mixed forms can be seen as a focal change within another vascular lesion such as a pyogenic granuloma, cavernous or capillary hemangioma (10), venous lake (11), arteriovenous malformation and blue rubber bleb nevus (12).

In the present case, we could not find characteristic papillary growth of the endothelial cells and associated organizing
thrombus material but we suspected that the lesions were in the late stage and the characteristic papillae might be fused and clumped (13). Although the possibility of true capillary hemangioma still remained, in view of the benign endothelial cell proliferation connected to the inner side of the vascular wall, and no tendency to extend beyond the vessel wall, we diagnosed the lesion as late stage IPEH.

Preoperative diagnosis of IPEH of the renal sinus is difficult because there is no particular symptom and no radiological consensus. However, no case of metastasis or malignant degeneration has been reported with IPEH. When operative treatment is chosen, intraoperative tissue diagnosis and kidney-sparing treatment are recommended.

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References