Intravascular Papillary Endothelial Hyperplasia: Regarding a Case

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Abstract

We report a case of a patient who presented with right renal colic pain. Complete workup for colic pain was performed and a heterogeneous lesion was found that seemed to originate from the right adrenal gland. Tumor in the right adrenal gland was suspected and the patient underwent radical nephrectomy. Pathological diagnosis of intravascular papillary reports endothelial hyperplasia, one rare benign lesion. The patient is currently asymptomatic and without findings in follow-up visits.

Keywords: Intravascular papillary endothelial hyperplasia; Renal tumor; Nephrectomy

Introduction

Intravascular papillary endothelial hyperplasia (IPEH) was first described as a malignant lesion by Pierre Masson in 1923 [1], who termed it vegetant intravascular hemangioendothelioma and observed its histological likeness to angiosarcoma. Later, Henschen [2] described a similar intravascular endothelial proliferation, interpreted as a reactive process secondary to vascular inflammation and stasis. We present a case of a patient diagnosed with intravascular papillary endothelial hyperplasia of renal vein.

Case Presentation

A 61-year old male patient was referred to our department with symptoms compatible with right renal colic. The patient had history of hypertension, obstructive sleep apnea syndrome and benign prostatic hyperplasia treated with alpha-blocker therapy. The only symptom reported by the patient was intense pain in the right flank, sometimes radiating to the ipsilateral inguinal region. Physical examination revealed right flank pain with percussion. Results of blood and urine analysis, abdominal X-ray and ultrasound study were normal. The persistence of symptoms led to the performance of a CT scan with contrast that revealed a lobulated solid mass sized 3.5 cm × 3 cm × 4 cm, with heterogeneous necrotic core, located at the right renal hilum, and infiltrating the renal vein (Figure 1). This mass was adjacent to the lower portion of the adrenal gland, to the kidney, and the posterior portion of the duodenum, and its origin could not be clearly identified. MRI was requested in order to identify the origin, and the mass was described as a tumor both T1-hypointense and T2-hyperintense (Figure 2), of probable adrenal origin.

Suspecting adrenal injury, hormonal and metabolic workup was carried out with blood and 24-hour urine samples, and the results were: serum cortisol 12.3 μg/dl; 24-h urine cortisol 102.2 μg/dl. The patient was treated with glucocorticoids and mineralocorticoids and did not present with hypertension.

Figure 1: CT: heterogeneous mass with necrotic core located in the right renal hilum.
ng/dl; DHEA-S: 1.2 μg/ml; 24-h urine dopamine 362 μg/24-h; serum aldosterone 30 pg/ml; vanillylmandelic acid in urine 5.1; ACTH 11 pg/ml. MIBG-scintigraphy was also requested but no enhanced lesions suggestive of pheochromocytoma were found. Surgery was decided given the findings described and the likelihood of a nonfunctional adrenal malignancy. Subcostal laparotomy was performed, and a tumor about 4 cm in diameter was found firmly adhered to the renal hilum. Simple tumor excision was impossible, so right radical nephrectomy was carried out. No incidences were reported during the postoperative follow-up, and the patient was discharged on the fourth day. During microscopy examination of the surgical specimen, the kidney and the adrenal gland were unaltered. The mass was close to the renal hilum and partially contained within a dilated vein, with thrombosis and endothelial reactive area proliferation (capillary and papillary), which was compatible with intravascular papillary endothelial hyperplasia (Figure 3). Visits are scheduled every six months, and analytical tests and CT images are normal at present.

Discussion

Several morphological features were suggested by Enzinger and Clearkin [3] that serve in the differential diagnosis of IPEH and angiosarcoma. These include intraluminal location of the lesion, the absence of necrotic tissue and the presence of thrombotic material. These authors also proposed the term intravascular papillary endothelial hyperplasia. Etiology of IPEH is still unknown, although trauma has been proposed as the main etiological factor. However, traumatic history is exceptional. Several authors agree with Clearkin and Salyer, who believe the cause to be an alteration in the thrombosis process, consisting of an unusual and peculiar way of thrombus organization [4,5].

IPEH can occur at any age, and is more frequent in female patients. The lesions are most often localized in skin vessels, head and neck, where they appear as small hard masses in the skin of bluish red color. However, less frequent locations such as jejunum, central nervous system, liver, and lungs have been reported [5,6].

Three different types of IPEH have been described: Primary IPEH affecting dilated vascular lakes; secondary or mixed IPEH, after a preexisting vascular lesion such as hemangioma, arteriovenous malformations or pyogenic granuloma; and extravascular IPEH, less frequent and presenting as the result of a hematoma [7]. IPEH in the renal vein is rare, and very few cases have been reported in the literature. Its symptoms are variable, ranging from the incidental finding in an otherwise asymptomatic patient, to colic pain and hematuria. When imaging techniques are used, CT scan with contrast reveals a solid lesion with heterogeneous contrast uptake, and a lesion hypointense on T1 and hyper intense on T2. These observations do not exclude malignancy [8,9].

Kidney preservation was possible in one of the cases reported in the literature [10,11] after preoperative diagnosis and lesion excision. However, this was not possible in most cases, as malignancy could not be ruled out, so radical surgery is usual: proximity to the renal vessels poses technical impossibility, and the suspicion of malignancy recommends it. Cases with metastases or malignant degeneration have not been reported.

Preoperative diagnosis of IPEH is challenging because there are no characteristic symptoms and no imaging technique allows for adequate differential diagnosis [12]. The latter is very limiting, since several neoplastic and non-neoplastic lesions can be found in the renal hilum, including renal carcinoma, angiomylolipoma, schwannoma, myelolipoma, hemangiopericytoma, lymphoma, cysts, Castleman disease or lipomas. Therefore, this condition, although rare, must be considered among the diagnostic possibilities, especially if the origin is elucidated or proximity to vessels is detected.

References

