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, 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.
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 extravasal 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 hyperintense 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, angiomylipoma, schwannoma, myelolipoma, hemangioendothelioma, 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**

