EXTENSIVE SPONTANEOUS PERIRENAL HEMATOMA SECONDARY TO RUPTURED ANGIOMYOLIPOMA: CASE REPORT

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SUMMARY – A rare case of extensive spontaneous perirenal hematoma from a ruptured angiomyolipoma is described. A 55-year-old woman was admitted to our department with a complaint of acute left flank pain. The images and clinical data of the case are presented. The imaging diagnostic techniques employed were abdominal ultrasonography, computed tomography scan and magnetic resonance imaging, which indicated the diagnosis of a large left perirenal hematoma and showed its size and extension. No tumor was suspected preoperatively. The operation was completed with nephrectomy and definite diagnosis was made on the basis of pathology findings, which revealed a relatively small angiomyolipoma on the upper pole of the left kidney wherefrom the hemorrhage had occurred. Therapeutic approach, i.e. nephrectomy or conservative therapy, remains controversial, however, concerning the prevalence of renal neoplastic lesions, most recent reports recommend nephrectomy in case of even slight doubt of renal neoplasm. The clinical, diagnostic and therapeutic aspects of this rare disease are discussed.

Key words: *Hematoma, diagnosis; Hematoma, surgery; Kidney neoplasms, complications; Angiomyolipoma, complications; Case report*

Introduction

Spontaneous retroperitoneal hemorrhage is an uncommon entity and even rarer when the underlying cause is from ruptured angiomyolipoma. Renal tumors account for the majority of atraumatic kidney rupture. Renal cell carcinoma and angiomyolipoma are the most common diseases in this group[1]. Oral anticoagulant therapy and hemodialysis could be responsible for a few cases. With the help of modern facilities, diagnosis can be made preoperatively and conservative surgery is indicated in these patients. However, nephrectomy is the treatment of choice in patients presenting with hemorrhagic shock as the initial symptom or with solid renal mass with perirenal hematoma. Preoperative tissue specific diagnosis of renal angiomyolipomas is now frequently made by computed tomography (CT)[2]. Although hemorrhage of these hypervascular tumors is a common cause for presentation, small tumor is rarely recognized preoperatively, as in this case[2].

Case Report

A 55-year-old woman was admitted to our department with a complaint of acute left flank pain. Prior to admission, physical examination showed a palpable, painful mass in the left upper quadrant of the abdomen and in the left flank. The patient denied recent trauma or use of anticoagulant therapy. There was no hematuria. Ultrasonography was remarkable showing extensive inhomogeneous mass

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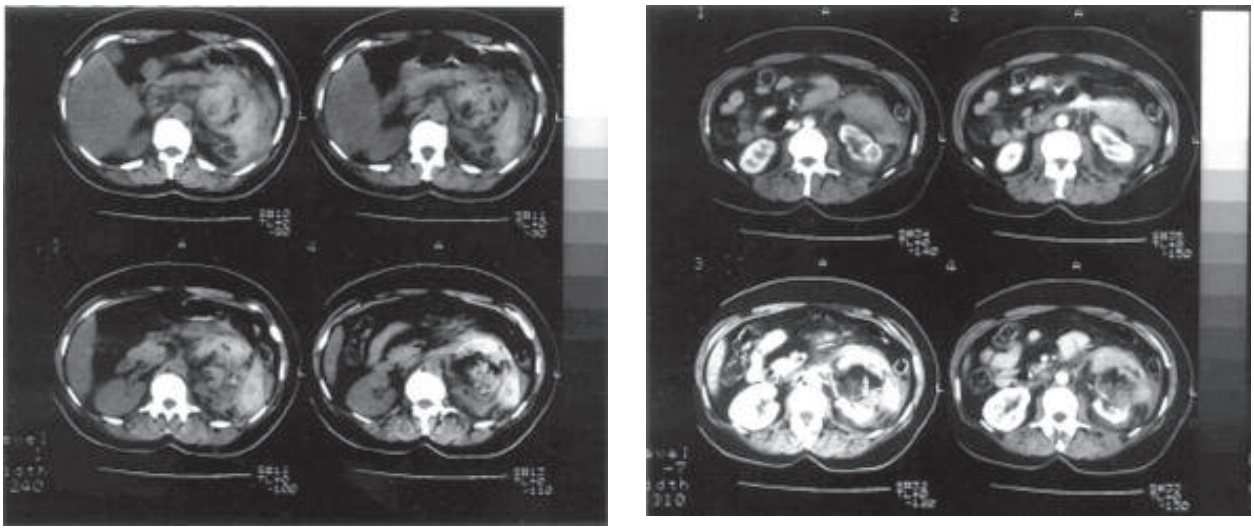

Fig. 1. Precontrast (left panel) and postcontrast (right panel) section of abdominal CT showing extensive retroperitoneal hematoma originating from the upper pole of the left kidney.

in the left upper retroperitoneal region, overlying the left kidney that could not be identified clearly. A large retroperitoenal hematoma was suspected and abdominal CT scan (Fig. 1) and magnetic resonance imaging (MRI) were done immediately upon admission to establish the extent and origin of hematoma. The imaging diagnostic techniques did not raise any suspicion of tumor but revealed a large retroperitoneal hematoma originating from the upper pole of the left kidney, extending throughout the left retroperitoneum and into the left pelvis major. Urgent surgery ended with nephrectomy due to the extent of renal damage, and definite diagnosis was made on the basis of the pathology findings that revealed a relatively small (measuring about 2 cm) angiomyolipoma on the upper pole of the left kidney wherefrom the hemorrhage had started (Fig. 2). The diagnosis was confirmed immunohistochemically with HMB-45 staining, which is the best marker for the diagnosis of angiomyolipoma (Fig. 3).

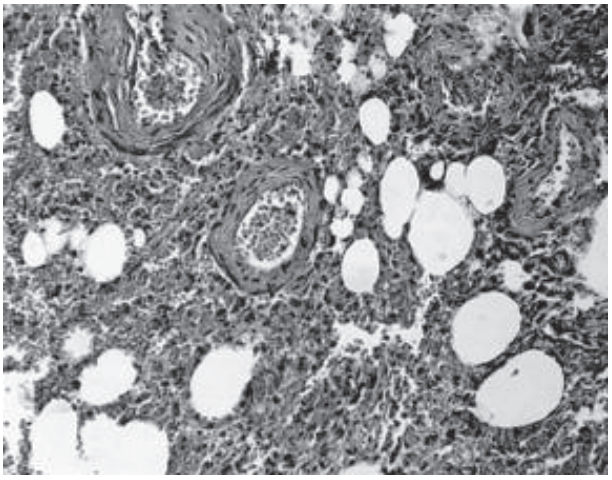

Fig. 2. Typical histologic pattern of angiomyolipoma with prominent vessels intermixed with smooth muscle cells and adipocytes. (HE, X40)

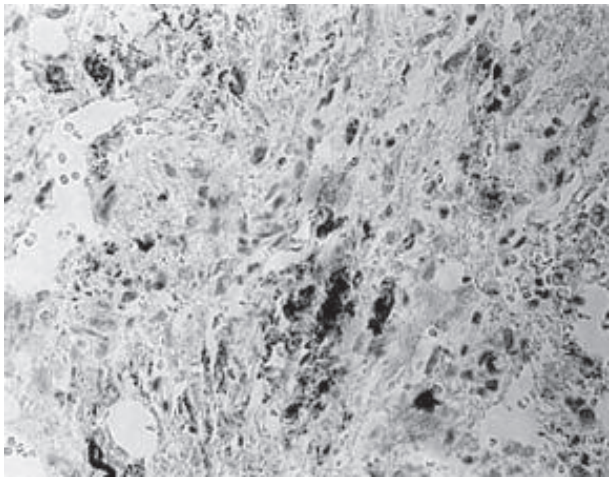

Fig. 3. Histologic specimen with immunohistochemical analysis showing the lesions to be positive for HMB-45, which is the best marker to confirm the diagnosis of angiomyolipoma.

Discussion

To our knowledge, less than 50 cases of spontaneous retroperitoneal/perirenal hematoma caused by ruptured angiomyolipoma have been reported in the literature so far, and this report is the first such case reported in a Croatian patient. Angiomyolipomas are usually found in women aged 50-70 years, and are considered benign mesenchymal tumors. Occasionally, angiomyolipomas may be characterized by cellular pleomorphism and mitoses, and even regional lymph node involvement[3,4]. Complications from angiomyolipomas are rare but often severe depending on the size and content of the lesion. Complications of angiomyolipomas reported in the literature to date included compression of the pyelocaliceal system, intratumoral bleeding, rupture with subcapsular, perirenal or pararenal hematoma, and extensive perirenal/retroperitoneal hematoma, like in this case[5]. Generally, it has been accepted that patients with isolated angiomyolipoma smaller than 4 cm should be followed up, whereas those with lesions greater than 4 cm and moderate or severe symptoms should undergo surgery. The possibility of follow-up every 6 months prevents the development of complications from the tumor[6]. Whenever there is collection detected by ultrasound in various anatomic renal or perirenal spaces in a patient with acute flank pain and low hemoglobin or hematuria, it is advisable to perform abdominal CT or MRI and search for hematoma. Small amounts of fat, detected by ultrasound, CT and MRI, may lead to the diagnosis of underlying angiomyolipoma that can spontaneously rupture, or in case of minor forces applied to the kidney. In cases where surgery is necessary, a kidney sparing technique should be performed whenever possible, such as enucleation of the lesion or partial nephrectomy[6]. Unfortunately, the most rational treatment in case of isolated, operatively detected angiomyolipoma still consists of nephrectomy because of the possible diagnostic mistakes and concomitance of malignant neoplastic diseases[6]. When imaging procedures fail to reveal the cause of spontaneous perirenal/retroperitoneal hemorrhage, exploration and biopsy are mandatory to exclude renal cell carcinoma[7]. Spontaneous perirenal hematomas essentially raise the problem of etiologic diagnosis which may occasionally prove incorrect. Therapeutic approach, nephrectomy or conservative treatment, remains controversial, however, in the light of the prevalence of renal neoplastic lesions, many authors recommend nephrectomy in case of even slight doubt about renal integrity[8]. When no cause can be found, the assessment can be completed postoperatively with long-term, close surveillance because of the risk of an undiagnosed neoplastic lesion[7,8]. Our case clearly shows that a relatively small angiomyolipoma can rupture spontaneously. Also, there is a possibility that a primary tumor was much bigger prior to hemorrhage, indicating that the tumor is just a residuum following intratumoral bleeding. Despite the conservative approach in the management of angiomyolipomas smaller than 4 cm, it still remains controversial whether to do follow up or to perform a surgery.

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Sažetak

VELIKI SPONTANI PERIRENALNI HEMATOM UZROKOVAN RUPTUROM ANGIOMIOLIPOMA: PRIKAZ SLUČAJA

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Prikazan je slučaj velikog perirenalnog hematoma uzrokovanog rupturom angiomiolipoma u 55-godišnje bolesnice primljene na kliniku zbog akutne boli lijeve lumbalne regije. Uporabljene dijagnostičke tehnike bile su ultrazvuk, kompjutorizirana tomografija i magnetna rezonanca, a prikazale su veliki perirenalni retroperitonealni hematom, te njegov opseg i odnos prema ostalim strukturama. Prijeoperacijski se nije posumnjalo na tumor lijevog bubrega. Hitna operacija završila je nefrektomijom, a definitivna je dijagnoza dobivena patohistološkom analizom koja je otkrila relativno malen angiomiolipom na gornjem polu lijevog bubrega iz kojega je krvarenje započelo. Cilj je prikaza pokazati kako još uvijek postoje proturječja o terapiji angiomiolipoma u odnosu na njegovu veličinu te moguće komplikacije. Raspravlja se o kliničkim, dijagnostičkim i terapijskim vidovima ove rijetke bolesti.

Ključne riječi: *Hematom, dijagnostika; Hematom, kirurgija; Neoplazme bubrega, komplikacije; Angiomiolipom, komplikacije; Prikaz slučaja*