Renal angiomyolipoma rupture: Case report

Rotura de angiomiolipoma: Relato de caso

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ABSTRACT

Generally benign, renal angiomyolipomas are neoplasms composed of mature adipose tissue, smooth muscle, and thick-walled blood vessels. Although asymptomatic, massive retroperitoneal hemorrhage from angiomyolipomas accompanied by hypovolemic shock has been found in up to 10% of patients, usually those with large tumors. We report the case of a 33-year-old woman with spontaneous rupture of an angiomyolipoma, who was initially treated with embolization of the lesion in order to stabilize her clinical condition, and later submitted to a partial nephrectomy.

Keywords: Angiomyolipoma/surgery; Rupture; Hemorrhage; Kidney, neoplasms; Case reports [Publication type]

INTRODUCTION

Angiomyolipomas (AML) are generally benign lesions (1-3), although the epithelioid angiomyolipoma, a subtype that occurs in about 3% of cases, can have an aggressive behavior. This tumor is characterized by mature adipose tissue, smooth muscle, and thick-walled blood vessels (3-4). The true nature of these lesions is unclear, but they are usually classified as hamartomas. Thirty-three percent of patients with AML have tuberous sclerosis (TS), a dominant autosomal disease characterized by mental retardation, epilepsy, and sebaceous adenomas; more than 80% of patients with tuberous sclerosis have AML (1).

CASE REPORT

We report a case of a 33-year-old woman at our institution who presented with a history of intense abdominal pain in the left flank, associated with nausea.

Initially interpreted as renal colic, an ultrasound of the urinary tract identified a cystic lesion in the upper pole of the left kidney, leading to a hypothesis of a renal abscess. The patient’s hemoglobin was 11.0 g/dL. A computed tomography of the abdomen revealed a solid 80.1 mm diameter solid nodule, compatible with AML of the upper pole of the left kidney, with signs of recent bleeding (figure 1). The patient was hospitalized for observation and had a reduction in her hemoglobin to 8.0 g/dL within 4 hours. She was submitted to renal arteriography that showed contrast leakage in the upper pole. Embolization of the upper pole of the left kidney was performed, and her clinical picture stabilized (figure 2) and progressed with no complications. On day 25 post-embolization, the patient underwent an upper left polar nephrectomy, and the pathological report confirmed AML with a

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Study carried out at Hospital São Camilo – Pompéia, São Paulo (SP), Brazil.

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The predominance of the lipomatous component (figure 3). She was discharged on the fourth post-operative day, and her clinical progress was uneventful.

**DISCUSSION**

Angiomyolipoma [AML] was originally described by Fischer in 1911, and was named by Morgan in 1951. Cases of AML are found in 0.3% of all autopsies and in 0.13% of the population when evaluated by ultrasonography².

In the group of patients that present AML associated with TS, the mean age of onset is 30 years, and the female gender predominates at the ratio of 2:1. Female predominance is even more accentuated in the group with no TS association, occurring at an older age (fifth and sixth decades of life). AML associated with TS is more commonly bilateral and multicentric, with a tendency for accelerated growth². Typically, lesions are asymptomatic, but there may be flank pain, a palpable mass, hematuria, or a combination of these signs and symptoms, especially in patients with TS. In 10% of patients, generally those with large tumors, there may be intense bleeding accompanied by hypovolemic shock¹, typifying the Wunderlich syndrome. Pregnancy seems to increase the risks of hemorrhaging.

The presence of fat inside a renal nodule, although not pathognomonic, virtually leads to the diagnosis of AML. Therefore, the ideal radiological method for distinguishing AML is computed tomography (density less than 10 UH), that presents a 95% accuracy rate. The presence of microaneurisms on the arteriography is the safest prognostic factor for determining the possibility of AML rupture⁴.

Embolization should be the first treatment measure implemented in patients with an AML hemorrhage, however, in large tumors there may be no tumor mass reduction after this procedure even though new bleeding may be prevented¹.

Prophylactic surgery is justified in cases of large tumors, women at childbearing age, or patients whose follow-up may be difficult or who have little access to emergency medical care. Several reports in literature suggest that surgical treatment should be used in tumors larger than 4.0 cm². Recent advances include a better genetic understanding of the TS-AML complex, and the identification of possible molecular markers that facilitate the histopathological diagnosis and refinement of embolization techniques¹.

**REFERENCES**