v. 3, n. 13 2023

International Journal of Health Science

ENDOVASCULAR TREATMENT OF RENAL ANGIOMYOLIPOMA RUPTURE: CASE REPORT

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Abstract: Renal angiomyolipoma is a rare type of benign tumor composed of blood vessels, adipocytes and smooth muscle fibers. It is commonly associated with patients with tuberous sclerosis. It presents asymptomatically and is usually diagnosed incidentally. In cases of spontaneous rupture, it may manifest through Wunderlich's syndrome, and may present pararenal retroperitoneal hemorrhage. This article presents the case of an 18-year-old patient who was initially transferred to Hospital Risoleta Tolentino Neves with suspected acute appendicitis, but was diagnosed with probable hemorrhagic angiomyolipoma on the right by computed tomography of the abdomen and pelvis. Embolization of interlobar branches of the upper pole of the right kidney was performed with resolution of the condition, without the need for a surgical approach.

INTRODUCTION

Angiomyolipoma represents a rare tumor that has in its composition three types of tissue in different proportions: fat, blood vessels and smooth muscle[1,2]. It has a great relationship with patients diagnosed with tuberous sclerosis, reaching a prevalence of up to 80% in this group. Its treatment takes into account the size of the tumor, the symptoms and the possibility of complications, with its rupture and evolution to hemorrhagic shock being the most serious complication[2,3].

CASE REPORT

An 18-year-old male patient was transferred to the UPA Risoleta Tolentino Neves Hospital on 12/04/21 with a two-day history of abdominal pain in the flank and right iliac fossa (FID), associated with nausea and vomiting, in addition to two episodes of syncope the day before. A US of the external abdomen was performed on the day of admission, and the report described a non-compressible cecal appendix measuring 5.6 mm, with increased vascularity on Doppler and increased echogenicity of the adjacent fat, suggesting an acute appendicular inflammatory process. On clinical examination, he was pale and dehydrated, tachycardic (HR 115 bpm) and with a slightly distended abdomen, painful on palpation in FID, positive Blumberg.

This is a patient with tuberous sclerosis diagnosed at the age of 1 year and 8 months, with no other comorbidities. Due to this prior context, an urgent computed tomography scan of the abdomen and pelvis was performed, showing a voluminous pararenal retroperitoneal hematoma on the right, in addition to a heterogeneous round mass measuring 9.0 x 6.5 cm, with soft tissue and fat density, strong enhancement and blush due to arterial extravasation of contrast on the lateral aspect and upper pole of the right kidney, without signs of acute appendicitis.

A diagnostic hypothesis of spontaneous rupture of renal angiomyolipoma was suggested and an evaluation by the vascular surgery team was requested in order to attempt embolization of the lesion. The patient was referred to the operating room on 12/04/21 and underwent embolization of interlobar branches of the upper pole of the right kidney with two 4 mm Nester coils, obtaining immediate control of contrast extravasation in fluoroscopy. Tomography performed the day after the procedure showed low contrast in the upper and middle third of the kidney, with no signs of active bleeding.

After two days in the ICU and two days in the ward, maintaining hemodynamic stability and clinical improvement with no signs of recurrence, the patient was discharged from hospital for outpatient follow-up with the service's urology team.

DISCUSSION

Renal angiomyolipoma is a benign tumor strongly associated with patients with tuberous sclerosis, a clinical entity that has mental retardation, sebaceous adenoma and the presence of renal angiomyolipoma in its spectrum of manifestations. These patients commonly present the neoplasm in a multifocal, bilateral form, and with larger lesions[1,2,4].

Tumors are usually asymptomatic, and the diagnosis is made incidentally. However, as in the patient in question, up to 10% of cases may manifest with retroperitoneal hematoma and hypovolemic shock. This presentation is called Wunderlich syndrome, an emergency condition characterized by hemorrhage in the perinephric space, which manifests through Lenk's triad: acute pain in the affected flank, palpable mass in the flank and hypovolemic shock[1].

The diagnosis can be established by imaging methods such as abdominal ultrasonography, computed tomography or magnetic resonance imaging. The follow-up of patients remains a controversial subject, but there are recommendations established in the literature. Asymptomatic patients with masses smaller than 4 cm must be monitored with ultrasonography or computed tomography every 6 months[5].

Among the criteria for intervention we have a diameter greater than 4 cm, intratumoral aneurysms greater than 4-5 mm, occurrence of pain or active hemorrhage, multiple or bilateral angiomyolipomas, unilateral angiomyolipoma in a single kidney, and patients with complex tuberous sclerosis[6]. In these cases, arterial embolization has been used as an alternative to surgical treatment[7].

CONCLUSION

It is concluded, therefore, that angiomyolipoma is a differential diagnosis of cases of renal masses, and that it may pose a risk to the lives of patients in cases of retroperitoneal hemorrhage, often requiring urgent treatment. Arteriography with embolization represents an effective treatment for this condition, and is also a less invasive modality than operative treatment.

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