Cavernous hemangioma of the adrenal gland mimicking a hepatic hemangioma

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Adrenal cavernous hemangiomas are a rare, benign, and non-functional tumor. We report a case of a 62-year-old male who presented with right upper quadrant and flank pain. Physical examination revealed a fullness of the right upper quadrant. Both computed tomography and magnetic resonance imaging suggested a hemangioma originating from the liver. During angiography with the intent of embolization, it was discovered that the vascular supply was consistent with an adrenal mass rather than a hepatic origin. The patient was referred to Urology and underwent curative right open adrenalectomy and nephrectomy. Histopathology confirmed the diagnosis of an adrenal cavernous hemangioma.

Key Words: benign adrenal mass, adrenal cavernous hemangioma

Introduction

Cavernous hemangiomas of the adrenal gland are a rare, benign, and functionally inactive tumor. Since the first case was described in 1955, approximately 70 cases have been reported in the literature.[1] The rare nature of this disease along with the non-specific clinical findings and investigations make the preoperative diagnosis exceptionally difficult for clinicians.

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Case presentation

A previously healthy 62-year-old male presented with right upper quadrant (RUQ) and right flank discomfort. On physical exam, the abdomen was soft and non-tender with subtle fullness in the RUQ. The patient appeared well, was not jaundiced, and showed no signs of liver disease. An ultrasound revealed a large lesion in the right flank area measuring approximately 9.5 cm. The origin was unclear and further imaging was arranged. Following both a CT, Figure 1, and MRI, it was concluded that this patient had a hemangioma originating from the liver. Arrangements were made for the patient to return for embolization of the presumed hepatic hemangioma a few months later.

At the time of angiography, it was discovered that the celiac trunk and superior mesenteric artery were

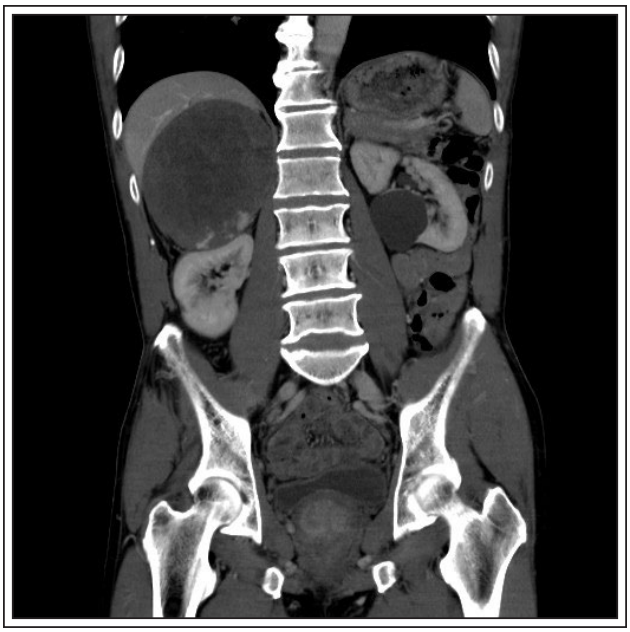

Figure 1. Coronal CT scan showing a large heterogenous RUQ mass ablating the liver and renal upper pole.

non-contributory to the tumor. In fact, the vascular supply of the tumor originated exclusively from a branch off the right renal artery, suggesting the mass was adrenal in origin, Figure 2. The planned embolization was not performed, and the patient was consulted to Urology. Subsequently, an adrenal functional work up and chest x-ray were ordered, both of which were negative. Given the size of the tumor and possibility of malignancy, the patient was scheduled for an open adrenalectomy. Prior to the operation, a CT was repeated, which again showed a 9.5 cm enhancing mass in the RUQ.

At the time of the operation, the patient underwent an open right adrenalectomy and nephrectomy via a subcostal incision. We could not identify a clear plane between the adrenal mass and upper pole of the kidney, so the adrenal gland and kidney were resected en-bloc to maintain an oncologic margin in case this was an adrenocortical carcinoma. Pathologic examination revealed a well circumscribed, encapsulated 9.5 cm mass arising from the adrenal gland, with extensive hemorrhage and necrosis, Figure 3. Microscopically, the mass was comprised of blood-filled thin-walled, dilated, and interconnecting vascular channels, consistent with cavernous hemangioma.

The patient had an unremarkable postoperative stay in hospital and was discharged home 5 days later. He was seen in follow up 6 weeks postoperatively. His recovery was unremarkable, he maintained normal renal function, and had returned to his baseline activities.

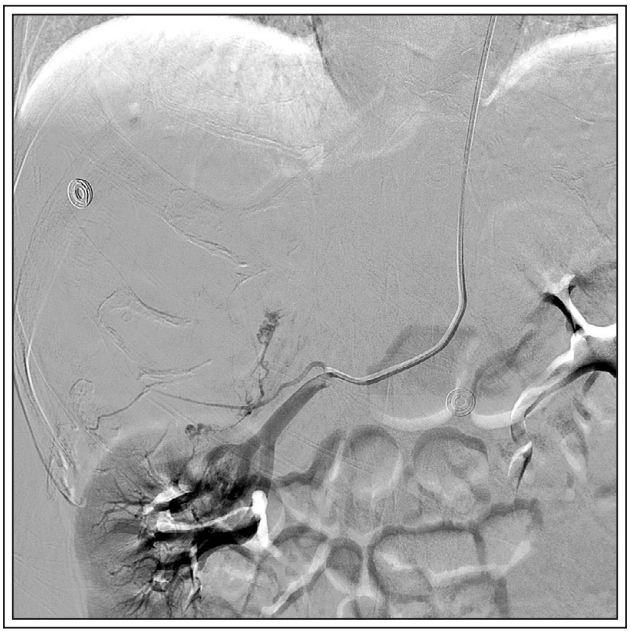

Figure 2. Right renal angiogram.

Discussion

Preoperative diagnosis of adrenal cavernous hemangiomas (ACH) is difficult for many reasons. The exceedingly rare nature of the disease, a lack of

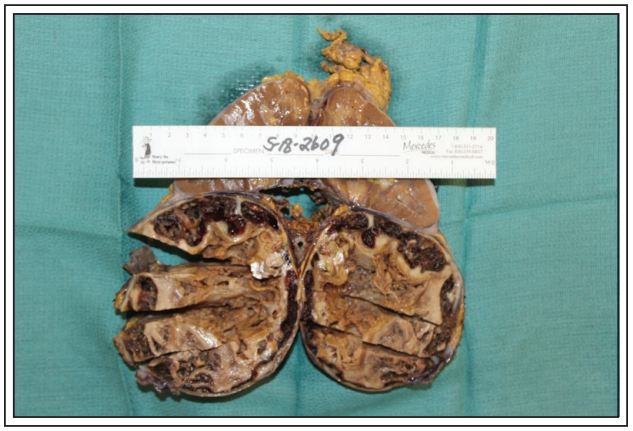

Figure 3. Gross photograph showing a large (9.5 cm) encapsulated mass with extensive hemorrhage and necrosis. Attenuated residual adrenal gland is seen at the periphery of the mass.

distinguishing features on history and physical exam, and non-specific investigation findings all contribute to this challenging diagnosis.[1-6] In this case, the diagnosis was made on histopathology following surgical resection.

The widespread use of imaging has led to the incidental detection of an increasing number of ACH.[2-9] Despite this, it is still estimated that these tumours represent only 0.01% of all adrenal tumors.[1] The fact that this disease is exceptionally rare makes it unlikely that it would be included in the differential diagnosis for most clinicians.

With ACH, patients tend to be asymptomatic until the tumor grows to a size that causes mass effect and compression on adjacent structures.[3,4,9] Prior cases have reported tumor sizes ranging from 2 to 25 cm, with the majority measuring greater than 10 cm.[1,4,9] When symptoms do develop, flank pain, early satiety, and a palpable mass are some of the features that have been described.[5] These history and physical findings are seen in countless abdominal diseases therefore limiting their utility when attempting to make the correct diagnosis preoperatively.

RUQ masses often require further investigation, and in the case of an adrenal mass, this includes imaging and a functional work up. On an enhanced CT, ACHs have been described as having a heterogenous internal structure with peripherally patchy enhancement in over 50% of cases.[3,5] Additionally, speckled calcifications that are secondary to phleboliths have been observed on both CT and radiographs in up to two-thirds of cases.[1] On MRI, dynamic studies with peripheral spotty and centripetal enhancement are characteristic. Marked hyperintensity on T2-weighted images combined with focal hyperintensity on T1-weighted images corresponds to areas of calcification and hemorrhage that are seen in ACHs.[9] Although helpful, these imaging characteristics are not pathognomonic and can also be seen with other adrenal tumors. With regards to the functional work up, the vast majority of ACHs are inactive.[3,4,6,8] These results allow certain diagnoses to be excluded (i.e pheochromocytoma), however, the differential diagnosis remains broad and includes worrisome diagnoses such as adrenocortical carcinomas.

With the lack of specific findings on history, physical and subsequent investigations, it is difficult to propose strategies for clinicians that would allow of a preoperative diagnosis. Current guidelines for adrenal incidentalomas recommend surgical resection for masses > 6 cm, and considering results from initial and follow up evaluations for masses between 4 and 6 cm.[10] Given that the majority of ACHs previously reported have measured greater than 10 cm, these masses would be subject to surgical resection based on their size alone due to the high risk of adrenal malignancy (35%-98%). For smaller masses, clinicians should expand their differential diagnosis for adrenal masses to include all possibilities, including rare entities such as ACHs.

Conclusion

In summary, we present a case of an ACH that was initially thought to be a hepatic hemangioma. Only once the vascular supply was identified on angiography was the diagnosis of an adrenal mass established. The preoperative diagnosis of ACHs remains extremely challenging for clinicians for several reasons. However, as the number of adrenal incidentalomas continues to rise, it is important for all diagnoses to be considered. □

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