Bilateral Cavernous Hemangiomas Causing Liver and Inferior Vena Cava Compression: A Case Report

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Introduction

This case highlights extensive progression of a relatively rare disease. In our case summary we highlight the progression of a bilateral adrenal hemangioma progressing to a size significant enough to cause severe pain and partially compressing the IVC. Adrenal hemangiomas of this significance are rare, and mass effect to this degree is also an underreported phenomenon. To our knowledge this is the first case of a symptomatic bilateral adrenal hemangioma causing mass effect.

Case Report

A 68 year old male with a history of a large renal cyst underwent evaluation for abdominal fullness and persistent dry cough. As part of his workup he received a CT scan of the chest, abdomen and pelvis (Figure 1) which revealed bilateral large irregular and heterogeneous enhancing mass involving the adrenal glands which was new compared to the imaging performed years prior. On the right, the largest lesion measured 14.8 cm. The mass on the right demonstrated several areas of punctate calcifications and had irregular areas of peripheral enhancement with central necrosis. The mass displaced the inferior vena cava anteriorly and compressed/displaced the liver significantly. The left adrenal gland measured approximately 9 cm in diameter and was also a heterogeneously enhancing non-calcified mass. There was evidence of interlesional bleeding and peripheral nodular enhancement bilaterally on MRI (Figure 2). Core biopsy of the lesion revealed features of a possible vascular lesion or possibly organized hematoma; malignancy could not be excluded due to the nonspecific histological and radiographic features. Upper and lower endoscopy was performed prior to surgery as well as blood and urine studies to rule out functional indications and methods for their removal.

During laparotomy, there was no evidence of a primary malignancy within the abdomen or pelvis, however a large mass was encountered involving the right adrenal gland that measured approximately 15 cm in diameter and significantly displaced the retro hepatic vena cava and the liver along with the bile duct consistent with imaging. The left adrenal gland revealed a well-encapsulated mass that was approximately 10 cm in diameter also consistent with imaging. A frozen section of the right adrenal tumor revealed no evidence of malignancy but extensive necrosis limited the sensitivity of this intraoperative evaluation; a bilateral adrenalectomy was performed.

Discussion

Cavernous hemangiomas are lesions most often found in the liver and skin. Hemangiomas of the adrenal gland are rare. Although adrenal cavernous hemangiomas are benign in nature, their diagnosis is an important consideration in adrenal masses. Like most other masses, histological classification is the gold standard for diagnosis [1]. A fairly accurate differential can be made, however, by considering common trends in previously diagnosed masses of the adrenal gland by using modern imaging studies. Here, we discuss the most commonly found features of cavernous hemangiomas of the adrenal gland, as well as indications and methods for their removal.

Abdominal CT (Figure 1) and ultrasound of adrenal cavernous hemangiomas reveal a heterogeneous mass enclosed in a thick capsule; there may be diffuse areas of calcification and cystic spaces [2,3]. The peripheral areas of the mass may show patchy enhancement with enhanced CT where residual areas of the compressed gland persist. Findings on dynamic CT may show pooling of contrast material at

Figure 1: CT of abdomen and pelvis demonstrating heterogeneously enhancing mass with central necrosis.

Figure 2: MRI of Adrenal Mass demonstrating extensive hepatic compression: (a) axial (b) and coronal sections.

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the periphery of the lesion, which is consistent with the large venous sinuses seen on histological examination [2,3]. MRI is the preferred diagnostic imaging modality (Figure 2), with the most common finding of peripheral enhancement of the tumor [4]. Characteristic findings typically show multiple areas of hemorrhage and necrosis, as well as calcified areas [5]. Angiography may reveal ring-like vascular channels that retain contrast material in delayed films, which is characteristic of the neovascularity of angiomas. In general, although the incidence of adrenal mass “incidentalomas” have increased with the increased utilization of cross-sectional imaging (i.e., CT and MRI), the dedicated scans that are being performed today rarely require percutaneous biopsy. Unfortunately, a negative core needle biopsy does not exclude a malignancy. It is also important to differentiate incidentalomas compared to adrenal lesions identified during workup for staging cancer patients or symptomatic patients as the treatment can be significantly different.

Adrenal cavernous hemangiomas are typically asymptomatic until they reach sizes greater than 10 centimeters in diameter [3]. Indications for the removal of adrenal tumors are size, mass-effect, and complications such as hemorrhage, necrosis, or thrombosis. The treatment for cavernous hemangiomas is conservative with periodic follow up for smaller and asymptomatic lesions. Surgical resection is required, however, to exclude malignant disease, relieve pressure-related symptoms, and prevent hemorrhage [4]. Adrenalectomy can be performed laparoscopically if the lesion is less than 6 cm [6]. Most recently with robotic, image-guided surgery, partial adrenalectomies can be performed but in limited settings. A biopsy also can be useful, in clarifying an uncertain clinical picture and for confirming the presence of suspected metastatic disease [7]. Larger tumors of uncertain etiology should be removed through an open technique. Consequences of bilateral adrenalectomy include the need for supplementation of glucocorticoids, mineralo corticoids, and possibly adrenal androgens.

Conclusions

This case demonstrates several crucial concepts. First the differential diagnosis of adrenal tumors is vast, and the workup does require a systematic approach. Specific to this case however, benign lesions can have very significant consequences and even drastic clinical outcomes. This case highlights the need to consider this rare etiology among the diagnostic possibilities in symptomatic adrenal masses.

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References