Hemorrhage; Unusual Presentation of Adrenal Hemangioma

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Abstract  Cavernous hemangioma is a vascular tumor that are usually incidentally discovered during routine radiologic evaluation. Adrenal cavernous hemangioma is extremely rare benign tumor of the adrenal gland. We report a case of a 75 year old female presented to the emergency department with a bleeding adrenal cavernous hemangioma. A review of the literature, multimodality imaging features, and differential diagnosis of adrenal hemangioma is discussed.

Keywords: atypical hemangioma, adrenal mass, CT


1. Case Report

75 year-old female presented to the emergency with a three week history of right upper quadrant pain radiating to her back associated with nausea and constipation. She underwent CT of the abdomen that revealed a large right upper quadrant mass, questionably arising from either the liver or right adrenal gland. The mass demonstrated peripheral nodular discontinuous enhancement with some centripetal filling on delayed images, however, there was a large amount of central necrosis and small amount of central calcification. Imaging findings were suggestive of a hemangioma (Figure 1). The mass showed extrinsic mass effect on the adjacent liver parenchyma and right kidney.

Figure 1. 75 years old female with periodic right upper quadrant pain radiating to her back

Technique: Multiphasic CT scan of the liver after injection of 90 ml Omnipaque 350 IV contrast obtained in porto-venous phase axial images (after 60 sec) (A) and delayed phase axial images (after 3 mins) (B). Coronal reconstruction of the venous phase (C).

Findings: A large 15.8 x 15.1 cm mass within the expected region of the right adrenal gland in the upper right quadrant demonstrate peripheral nodular enhancement on venous phase (A, C) and centripetal filling on delayed phase (B) Note the presence of high dense calcification within the mass (Arrow head). The mass exhibits extrinsic mass effect on the upper pole of the right kidney and inferior border of the liver.
On clinical presentation, the patient was hemodynamically stable. She was evaluated by general surgery and referred for follow-up as an outpatient for possible resection. Outpatient endocrinology work-up demonstrated a non-functional adrenal tumor. Complete surgical resection was performed and pathology revealed a large necrotic benign neoplasm arising from the right adrenal gland with internal bleeding (Figure 2), consistent with complicated cavernous hemangioma.

Figure 2. 75 years old female with periodic right upper quadrant pain radiating to her back

Histopathology Findings: (A: 10x objective magnification [o.m], hematoxylin and eosin) and (B: 20x.o.m., hematoxylin and eosin): Few areas retain endothelial cell lining. There is abundant stromal hemorrhage with fibrin. These areas have a pseudopapillary appearance, not typical of the dilated thin-walled vessels seen in cavernous hemangiomas. There are eosinophilic bodies suggestive of ghost cells. Overall, much of the neoplastic stroma has been obliterated by coagulative necrosis with concomitant hemorrhage and fibrin deposition. Sparse collections of intravascular erythrocytes and fibrin indicate the presence of intravascular blood and thrombosis that was likely washed out during tissue processing.

(C: 20x.o.m., hematoxylin and eosin) and (D: 10x.o.m., hematoxylin and eosin): This chronic organization can be seen throughout 90% of the neoplasm. Fibroblastic hyperplasia can be appreciated, and there is haphazard deposition of collagen indicative of organization (granular tissue). Hemosiderin-laden histiocytes can be seen interspersed throughout the organizing granulation tissue. Notably, there is a focus of hemorrhage within the lower-right corner that is not lined by endothelial cells. This pattern was prominent in the neoplasm, and indicates leakage of blood from the intravascular compartment into the surrounding interstitium.

2. Discussion

Adrenal hemangiomas are rare benign tumors with less than 100 cases documented in literature. The first case was described in 1955. They are usually incidentally identified, found in up to 10% of patients undergoing abdominal imaging. There is increasing frequency of detection with the widespread use of CT, MRI, and ultrasonography [1].

The vast majority of adrenal incidentalomas are benign adenomas (80%). The remaining 20% is made up of both malignant and benign lesions. Malignant lesions include adrenal cortical carcinoma, metastatic disease, and pheochromocytoma. Benign entities include cysts, myelolipoma, hematoma, ganglioneuroma, and hemangioma [4]. Neuroblastoma is also a consideration in the pediatric population.

Adrenal hemangiomas are well-encapsulated and found in the adrenal cortex. Histologically, they are composed of dilated vascular spaces lined by endothelium, surrounded by a collagenous wall. Similar to hemangiomas elsewhere in the body, they contain areas of hemorrhage, necrosis, degeneration, and calcification [5]. Peripherally located vascular cavities are also a feature, which correlates to the characteristic early peripheral enhancement seen on CT and MRI. They are most commonly seen in the 5th to 7th decade, with females outnumbering males by a factor of 2.
Incidentally discovered hemangiomas are usually asymptomatic and non-functioning tumors. Occasionally, they may bleed and become symptomatic. Symptoms typically include abdominal or flank pain. Rarely they may show endocrinology symptoms related to hypercortisolism or hyperaldosteronism. It has been suggested that the hemangiomas do not directly cause endocrinopathies, but rather arteriovenous malformations within the lesions lead to enhanced entry of active endocrine metabolites into the body [1,2,3,7].

Because the majority of adrenal hemangiomas are asymptomatic, imaging plays an important role in narrowing the diagnosis. Generally, hemangiomas tend to be unilateral. Ultrasound findings are nonspecific, often demonstrating a mass of variable size and echotexture. Unenhanced CT will typically show a soft tissue attenuating mass, varying in size. Larger masses frequently will have calcifications which represent either phleboliths or dystrophic calcification from prior hemorrhage. On Contrast enhanced CT, adrenal hemangiomas will enhance similar to hemangiomas elsewhere, with heterogeneous peripheral enhancement early that gradually fills in over time. MRI usually demonstrate high T2 signal intense, heterogeneous low T1 signal intense lesion, post-contrast appearance is similar to CT, with peripheral nodular enhancement that gradual fills on delayed images [2,3,9,14].

While certain imaging features aid in narrowing the diagnosis, the differential diagnosis can be challenging [13]. Lesions less than 3.5 cm with typical imaging characteristics can be monitored with follow-up imaging. For larger or atypical lesions, the treatment of choice is surgical excision due to difficulty in excluding malignancy [3,11]. Resection is also advocated for all functioning adrenal lesions, and to preclude future apparent hormonal hypersecretion. Endocrine Practice. 2008; 14(1):104-108.

References


3. Conclusion

Although rare, adrenal hemangioma should be included in the differential diagnosis of enhancing adrenal masses especially with atypical imaging findings.