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Case Report
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Adrenal Cavernous Hemangioma: An Uncommon Cause of Adrenal Gland Enlargement

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Abstract

Adrenal cavernous hemangioma is a benign, usually non-functioning tumor. It is often completely asymptomatic and incidentally discovered on radiologic imaging. CT-scan and MRI are useful tools for the preoperative study of the tumor, but they usually do not let to a clear preoperative diagnosis because of the lack of pathognomonic signs and because adrenal cavernous hemangiomas are rarely encountered lesions [1]. The most valuable tool remains histopathological examination after surgical resection. We herein report the case of a 74-year-old man referred to our institution for a large right adrenal mass. The biological workup was normal, and the imaging studies did not permit to completely characterize the lesion. A surgical resection was performed and histopathological examination diagnosed a cavernous hemangioma.

Introduction

Among incidental adrenal masses, cavernous hemangiomas are benign, usually non-functioning lesions, arising from vascular endothelial cells. Since the first surgical reported case in 1955, to our knowledge there are 67 unilateral and 1 bilateral case described in literature [1,2]. They are often incidentally discovered in asymptomatic patients, and pre-operative diagnosis is challenging because of the lack of specific findings on imaging studies. We herein report an additional case with the biological and radiological workup, surgical management and histopathological examination.

Case Presentation

A 74-year-old man was referred to our institution for evaluation of a right adrenal mass. In his past medical history we could notice recent surgery for aortic valve replacement, achalasia treated by pneumatic endoscopic dilation, and recent cigarette smoking cessation. He was under treatment by beta-blockers, acetylsalicylic acid and alprazolam. The mass was incidentally discovered on a thoraco-abdominal CT-scan during the pre-operative workup of an aortic valve stenosis. On interrogatory, the patient did not mention any mechanical or functional symptom. On clinical examination there were no signs or symptoms of a functioning adrenal tumor, and the lesion was not palpable. An exhaustive hormonal biological workup was undertaken: all the laboratory tests were in normal ranges (Table 1). On the CT-scan previously performed, a well-defined heterogeneous right adrenal lesion of 98 mm large, with attenuation values varying from 5 to 52 Hounsfield Units (HU) on non-contrast series (Figure 1), with one small calcification. The left adrenal gland was normal. A MRI was subsequently performed: the 9.5 cm × 8.5 cm × 9 cm lesions showed a heterogeneous high intensity signal on T2-weighted sequences with peripheral enhancement after contrast administration and a large central necrotic area (Figure 2); heterogeneity was less important in T1-weighted sequences with spontaneous peripheral high signal areas corresponding to hemorrhagic zones (Figure 2). A Positron Emission Tomography (PET) -CT was then performed and showed no hypermetabolic area in the right adrenal mass with few peripheral calcifications. On revision of all past imaging studies of the patient, the mass was retrospectively already present on a chest CT-scan realized in 2014 after endoscopic pneumatic dilatation for achalasia, measuring 32 mm × 25 mm at that time, but unfortunately no follow up was scheduled after that. Due to the heterogeneous aspect of this right adrenal non-functioning lesion, the presence of necrosis, and the size, a surgical resection was scheduled. The patient underwent a right adrenalectomy through a right open subcostal approach. We did not experience any significant blood loss. The post-operative course was uneventful and the patient was discharged home on the fourth post-operative day. Macroscopic pathological examination showed a specimen of 10 cm × 6 cm × 8.5 cm weighing

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Table 1: Laboratory results.

	Results	Normal ranges
Plasma		
Morning cortisol	17.1	3.7-19.4 µg/dl
ACTH	10.7	5.0-49.0 pg/ml
Aldosterone	16.6	<39 ng/dl
Renin	15.8	4.0-50.0 µU/ml
Progesterone	0.3	0.13-0.97 µg/L
DHEA	97	10-285 µg/dl
Testosterone	15.37	9.7-38.14 nmol/L
Urine		
Norepinephrine	32	10-100 µg/day
Epinephrine	3.2	2-20 µg/day
Dopamine	221	60-400 µg/day

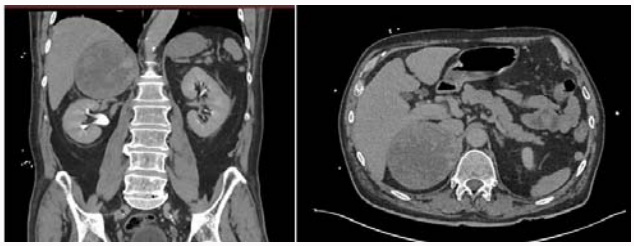

Figure 1: Coronal and transverse sections of the CT-scan, showing the well-encapsulated right adrenal lesion. On the right, image from the non-contrast series.

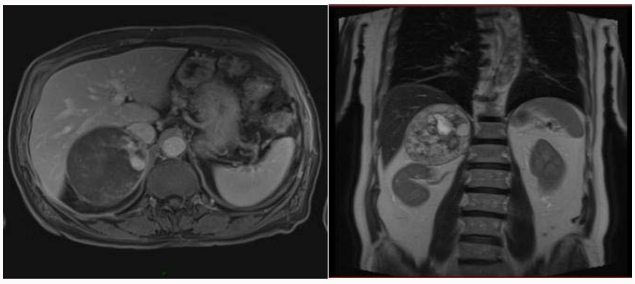

Figure 2: On the left, T1-weighted MRI showing a quiet homogeneous adrenal mass with spontaneous peripheral high signal intensity. A thin rim surrounds the lesion. On the right, T2-weighted MRI showing heterogeneous signal with highly vascularized nodular areas.

377 g. The lesion was well-encapsulated without any invasion of the peri-adrenal fat tissue. It looked widely cystic and hemorrhagic, with a large central necrotic area (Figure 3). Microscopic pathological examination showed a partially necrotic vascular lesion with a wide vascular cavernous pattern; the endothelium showed positive staining for ETS-related gene ERG, a highly specific endothelial marker (Figure 4), weak positivity was noted for Ki67. A small amount of normal adrenal cortex was still present (Figure 3 and 5). Microscopy confirmed no invasion of the peri-adrenal fat tissue. The final report of histopathological examination was a widely necrotic cavernous hemangioma of the right adrenal gland, which underwent complete resection with no argument for malignancy.

Discussion

Adrenal cavernous hemangiomas are benign tumors, arising from the endothelial linings in the adrenal cortex. A recent review of the

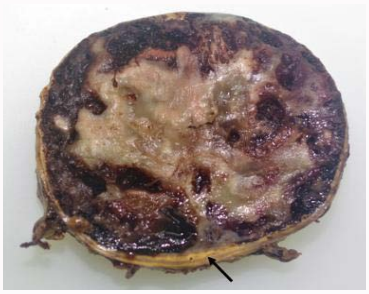

Figure 3: Macroscopic section of the lesion: we can see the capsule, some residual normal cortical parenchyma (yellow tissue, black arrow), and the vascular lesion with large hemorrhagic areas surrounding a central yellow zone (necrosis).

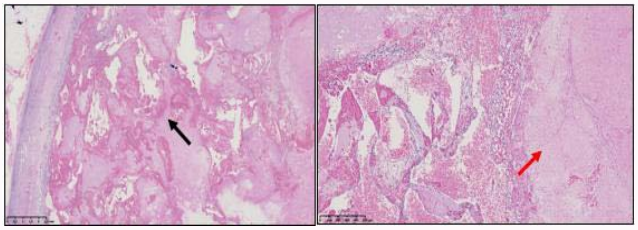

Figure 4: Left: Hematoxylin-eosin staining (0.88x). Starting from the left: The peri-adrenal fat tissue, the adrenal capsule, residual normal adrenal parenchyma and the vascular lesion (black arrow). The vascular structures follow a cavernous pattern. Right: Hematoxylin-eosin staining at higher magnification (5x). Large cystically dilated and interconnected vessels lined by endothelial cells. On the right, large necrotic areas (red arrow).

Figure 5: Immunohistochemistry 5x. Nuclear positive staining for ETS-Related Gene (ERG) on the left, a highly specific endothelial marker. On the right, a large necrotic area.

literature showed that they are most frequent in women, with a ratio of 3 to 2, and the median age is around 60-year-old [1]. There is no preferred side. Only one bilateral case has been reported in literature [2]. These tumors are completely asymptomatic in approximatively 60% of cases, but patients can also present with vague abdominal symptoms or discomfort. A rare presentation is spontaneous rupture with severe retroperitoneal hemorrhage [3].

While usually non-functioning tumors, six secreting cases of cavernous hemangiomas are described in the literature, three cases presenting signs of primary hyperaldosteronism and three cases presenting with subclinical Cushing syndrome [1]. Reactive hyperplasia of the normal cortex is sometimes associated to the presence of cavernous hemangiomas and highlighted during histological examination [2,4]. It could be responsible of the metabolic secretion described in these patients. Another explanation suggested by other authors is that arteriovenous malformations within the hemangiomas may release endocrinologically-active metabolites into

the bloodstream [5].

In our patient, biological workup excluded any adrenal secretion, consistently with the majority of the reported cases.

Pre-operative diagnosis is challenging because of the lack of specific findings on imaging studies. Speckled calcifications are common findings in adrenal hemangiomas, but they are lacking in almost half of the patients [1]. Some authors affirm that they could be phleboliths in dilated vascular spaces but their presence seems not to be pathognomonic for adrenal gland hemangiomas [6]. In our case, only few peripheral calcifications were present. On CT imaging adrenal cavernous hemangiomas commonly appear as heterogeneous complex masses, associating hemorrhage and necrosis, and high attenuation values on non-contrast CT with minimal contrast washout in contrast series [4]. All these features make adrenal cavernous hemangiomas difficult to differentiate from adrenal cortical cancer. On contrast series, the presence of peripheral enhancement followed by centripetal filling, and highly dense peripheral rim could suggest the diagnosis of cavernous hemangioma, these characteristics being typical for liver hemangiomas [4,7]. MRI may show a homogeneous adrenal mass with low intensity signal on T1-weighted imaging; peripheral focal hyperintensity is possible as a sign of hemorrhage. T2-weighted imaging may show a heterogeneous high intensity signal [7,8].

Indications for surgery are symptomatic lesions, complications such as hemorrhage and suspicion of malignancy. Size is a strong sign of possible malignancy. In literature, the risk for malignancy for adrenal lesions bigger than 6 cm ranges between 35% and 98% [9]. In our patient, because of the size of the mass, high attenuation values on non-contrast CT-scan, calcifications, heterogeneity, the presence of necrosis and the increase of tumor size over time, which could all be consistent with adrenal cortical cancer, we decided to perform a surgical resection by an open right subcostal incision.

In literature, 71% of the reported surgical resections for adrenal cavernous hemangiomas have been performed by an open approach, but it must be said that since the first published laparoscopic resection was performed in 2001, almost half of the surgery procedures have been performed by laparoscopy later on [1]. The main argument driving the choice of the surgical approach should be the size of the lesion and the possibility of malignancy and local invasion, as for other adrenal tumors [9,10]. Laparoscopy is safe for first exploration and vascular control of adrenal vein (s), but if the surgeon remarks local invasion the procedure should be converted to an open approach [9,11].

Histopathology leads to the definitive diagnosis. Microscopy usually shows a well-encapsulated lesion with large cystically dilated and interconnected vessels lined by non-atypical endothelial cells positive for endothelial markers, such as CD31 and ERG. Large necrotic areas are present [1].

Conclusion

In conclusion, a case of a 10-cm asymptomatic, non-functioning right adrenal cavernous hemangioma is presented. Although radiologic characterization is difficult, the possible diagnosis of adrenal cavernous hemangioma should be taken into account during the workup of an adrenal incidentaloma. Some radiological features may guide the preoperative diagnosis. However, adrenal cavernous hemangiomas are often large masses at the time of their discovery and surgical management is recommended to rule out malignancy, to confirm the diagnosis on histopathological examination, and to prevent further enlargement and risk of rupture with subsequent hemorrhage.

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