Cavernous hemangioma of the adrenal gland

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ABSTRACT

A case of cavernous hemangioma of the adrenal gland is described. Hemangiomas are rare adrenal tumors usually asymptomatic. The patients appear to develop late complaints from compression and displacement of adjacent organs.

Salient radiographic findings at CT are the presence of multiple calcifications with typical appearance of phleboliths and pooling of contrast material in the periphery of the mass. Because the imaging procedures lack specificity in the differentiation between malignity or benignity of the lesion surgical excision is needed.

Key words: adrenal gland neoplasm; cavernous hemangioma

Hemangiomas are benign vascular tumors consisting in dense collections of blood vessels. The term “cavernous” is used to describe the large vascular channels present in some of these neoplasms as opposed to the capillary type of hemangioma. Cavernous hemangiomas are frequently located in the skin and in the liver but they can also be present in the cerebellum and eye grounds as part of the von Hippel-Lindau syndrome in conjunction with cystic lesions of the pancreas. Hemangiomas of the adrenal gland are rare non functioning tumors. Usually asymptomatic they are often discovered incidentally on radiographic examinations or at autopsy. Only few cases come to clinical attention by a syndrome of abdominal mass with compression symptoms. The most of these tumors are cavernous. Their etiology is unknown although some physicians suggest a possible relationship with prior trauma (1-2) or displasia, typical of the old age (3). However, the increased use of US and CT for adrenal imaging may result in increasing recognition of these masses. The first surgically treated adrenal hemangioma was described in 1955 by Johnson and Jeppers (4).

A 76-year-old woman with a large mass in the left upper abdominal quadrant, discovered incidentally on radiographic examination, was hospitalized at our Department. The mass presented increase in size during the last six-ten months prior to admission, and the patient suffered moderate upper abdominal discomfort, slight weakness, but no major illnesses.
CT scans showed noncystic, rounded, large left adrenal mass (12 cm in diameter) in the upper left abdominal quadrant displacing anteriorly the stomach and inferiorly the left kidney (FIGURE 1). The central area of the mass was hypodense with focal loci of necrosis and hemorrhagic material. Marked marginal enhancement was also present and the periphery of the tumor was thick, irregular with calcifications. No clear cleavage between the mass and the kidney was seen (FIGURE 2).

The increment in volume of the mass during the time, the symptoms of compression on the contiguous structures and the risk of neoplastic degeneration were the predominant indications at surgery, despite the advanced age of the patient.

The patient underwent surgical operation through a bilateral transverse subcostal laparotomy. A large, left upper quadrant tumor, extremely adherent to the tail of the pancreas, to the ilus of the spleen and with macroscopic suspected invasion of the upper pole of the kidney was found (FIGURE 3).

En bloc left adrenalectomy, distal pancreatectomy with splenectomy and left radical nephrectomy were performed (FIGURE 4).

Several enlarged regional and lombo-postoperatively aortic lymph nodes were also discovered and dissected. The patient recovered uneventfully and was discharged from the hospital 14 days after surgery.

Pathologic examination of the specimen showed a large ovoid mass, approximately 12x13x9 cm, adherent to the spleen and to the kidney.

The mass was well circumscribed but without a clear plane of dissection inferiorly between itself and the kidney in correlation to the inflammatory response. A thick, firm fibrous wall surrounded the mass. On sectioning the

FIGURE 1. Contrast-enhanced CT scan shows a very large heterogeneous left adrenal mass with peripheral calcifications. Intense marginal enhancement after contrast medium injection

FIGURE 2. Not evident cleavage between the mass and the kidney

FIGURE 3. Intraoperative aspect of the adrenal mass including the spleen and the left kidney

FIGURE 4. Macroscopic aspect of the specimen
tumor showed a central replacement by reddish-yellow-brown granular materials and clots, prominent cavernous vessels and diffuse areas of hemorrhage, focal calcifications and a thin peripheral rim ochre-yellow related to the adrenal (FIGURE 5).

Histologically the mass was composed of numerous dilated vessels and cavernous spaces filled with red blood cells, and with an endothelium-line positive for factor VIII to immunoperoxidase staining. Normal adrenal glands and cells were identified at the periphery of the mass (FIGURE 6). There was no histologic evidence of malignancy (FIGURE 7). The spleen, the pancreas, the kidney and the lymph nodes showed a moderate grade of inflammation and sinuses histiocytosis. The diagnosis was “cavernous hemangioma of the adrenal medulla with diffuse thromboses”.

DISCUSSION

Patient age ranges between fifth and eighth decade of life with the vast majority of them in the sixth or seventh decade. Reported cases show a clear predilection for women.

Cavernous hemangioma is asymptomatic. Symptoms from adrenal medullary or cortical hyperfunction have not been described with adrenal hemangioma in contrast with other hormonally active adrenal tumors.

Also in our patient we did not found endocrinologic alterations. For this reason the mass comes often to the clinical attention as incidental finding during the course of radiographic imaging for non-urologic symptomatology. Hemangiomas may contain calcifications that are related to multiple phleboliths located in the dilated vascular spaces of the tumor. The calcifications can be noted on radiographic images but they are not specific of hemangioma because they occur in many other adrenal diseases such as carcinoma, cysts, tuberculosis, neuroblastoma and spontaneous hemorrhage. In some cases cavernous hemangioma come to the clinical attention because of its size. When the tumor becomes very large (masses) the patients may have non-specific symptoms like tenderness or abdominal vague discomfort and painless abdominal palpable mass. Gastrointestinal symptoms usually result from compression and anterior displacement of the stomach and the splenic flexure or transverse colon by a painless palpable abdominal mass (5,6).
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CLINICAL IMAGES

The presence of calcifications with the typical appearance of phleboliths or round, smooth calcified bodies with lucent center, is the usual radiographic finding.

Both CT and US can image adrenal space-occupying lesions. Computed tomography is superior to US and it is commonly considered the imaging procedure of choice when the presence of a suprarenal mass is suspected. The US and CT findings of adrenal hemangioma have been described in literature (7-10). Ultrasound examination shows a mass of moderate-high hyperreflectivity with a variety of non-specific structural patterns: heterogeneous, solid, cystic. In CT images the tumor appears as a low attenuated, heterogeneous and well-delineated mass. After bolus injection of contrast agent there is a peripheral inhomogeneous papillary enhancement of the lesion. These findings are similar to those found on angiography: multiple pools of contrast material at the periphery of the tumor. Although pooling of contrast material persists on late imaging as prolongation of this abnormal stain well into the venous phase of the study, the filling-in-phenomenon, frequently described in cases of liver hemangiomas, is seldom seen in adrenal hemangiomas for the presence of necrosis, fibrosis and thrombosis in the core of the tumor. The CT images are not specific of the tumor histopathology but they help to differentiate between benign and malignant lesions. In particular, the results of contrast enhanced CT seem characteristic enough to suggest a preoperative diagnosis of adrenal hemangiomas, at least in patients with large lesions. This may allow surgeons to plan more accurately the therapeutic approach to the patient and to avoid potentially dangerous fine-needle aspiration/or biopsy of the mass.

MRI reports of adrenal hemangiomas are very rare (1,11-13). The described findings are ambiguous, consisting of multiple areas of various signals related to aspecific hemorrhagic and necrotic foci.

PATHOLOGIC FINDINGS

Cavernous haemangioma usually consists of a well-circumscribed, encapsulated, spheroidal mass with a fibrous capsule enclosing a thin rim of yellow soft tissue, probably residual adrenal gland tissue. The cut surface shows that most of the mass is red with areas of necrosis and hemorrhage.

The histology demonstrates marked degenerative changes in vessel walls, some of which are obliterated. Moreover empty lacunae lined by fibrous tissue in the presence of blood and containing hemosiderin deposits are revealed. The blood filled lacunae are lined by benign endothelial cells. At the periphery of the tumor are present normal tissues and cells characteristic of the normal adjacent adrenal gland (1,9).

Despite the benign nature of adrenal hemangioma, surgical excision is recommended because a significant number of patients appears to develop local complications due to the large size of these lesions and because the risk of spontaneous hemorrhage or bleeding induced from minimal trauma.

CONCLUSION

Adrenal haemangioma are rare tumors of unknown ethiology. The symptoms are not characteristic. The usual complaints are abdominal discomfort, vague gastrointestinal symptoms and the presence of a palpable mass. The necessity of endocrinologic studies must be considered, because it is always possible to find some expression of hormonal activity in incidental adrenal mass. Because imaging procedures lack specificity, all adrenal masses should be considered malignant. Therefore the treatment of choice is surgical excision. Adrenalectomy or en bloc resection of involved adjacent organs is the correct surgical approach.
REFERENCES