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SHORT COMMUNICATION

Uncommon benign lesions of the adrenal glands mimicking sinister pathologies: report of 8 cases

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Summary

Purpose: *To present series of patients with large rare primary lesions of the adrenal glands that were operated in our department. The initial indication for surgery was decided based on their impressive similarity to other more sinister adrenal pathologies.*

Methods: *The clinical records of the department and histopathology reports, covering the 1986-2015 period were assessed. Rare adrenal pathologies that preoperatively were clinically mimicking other adrenal tumors were included.*

Results: *In total, 8 patients (age range 20-75 years) with rare tumors of the adrenal glands were found. Seven patients had a preoperative indication for adrenalectomy due to the possibility of malignancy. In one patient surgery was undertaken due to the possible diagnosis of adrenal hemorrhagic cyst. Among these patients 4 had a histopathologic diagnosis of hemangiomas, one of a lymphangiomatous cyst and 3 of myelolipomas.*

Conclusion: *Rare benign tumors of the adrenal glands can present as very large lesions that can be either diagnosed incidentally or due to atypical symptoms. Though unusual, they should be considered in the differential diagnostic approach of adrenal lesions, because they share common clinical and radiological characteristics with more sinister and frequent lesions such as malignant tumors and also hematomas.*

Key words: *adrenal gland, hemangioma, lymphangioma, myelolipoma, uncommon adrenal tumors*

Introduction

Primary adrenal tumors comprise a group of lesions characterized by significant heterogeneity [1]. This group may include malignant neoplasms with major clinical impact, such as cortical adenocarcinomas and pheochromocytomas, as well as more subclinical benign lesions, such as non-secreting cortical adenomas, pseudocysts, endothelial or vascular cysts and other mesenchymal conditions. Thus, careful investigation and proper management are crucial [2].

Although in most of the cases the preoperative diagnosis is rather conclusive, in some cases surgical excision and histological examination are mandatory in order to establish a definitive diagnosis, which often reveal unusual tumors [3,4]. Herein we present 8 cases of large and unusual lesions of the adrenal glands with their major clinical and histological characteristics.

Patient presentation and results

Data retrieved from the archives of the Pathology and Surgical Department of our hospital during the 1986-2013 period revealed 7 patients with large and uncommon lesions of the adrenal glands that were managed in our hospital, whose preoperative diagnosis was thought to be rather

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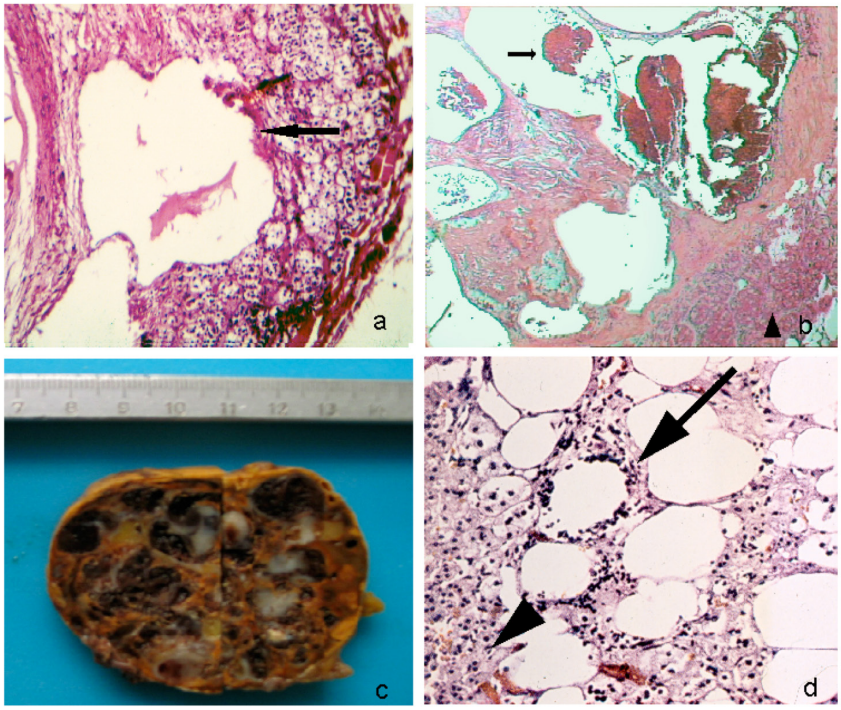

Figure 1. a. Histological section of adrenal cystic lymphangioma showing vascular spaces with lymph (arrows) (H&E, x25). **b.** Histological section of adrenal hemangioma showing blood-filled vascular spaces (arrow) surrounded by adrenal gland tissue (arrowhead) (H&E, x25). **c.** Gross view of adrenal hemangioma showing blood-filled vascular spaces surrounded by adrenal tissue. **d.** Histological section of adrenal mass showing bone marrow elements (arrow) surrounded by normal adrenal tissue (arrowhead) (H&E, x120p).

sinister as they were mimicking malignant lesions.

Seven patients had a preoperative indication for adrenalectomy due to high likelihood for malignancy, since preoperative cross-sectional imaging (i.e. computed tomography/CT and magnetic resonance imaging/MRI) could not exclude malignancy. In one patient surgery was undertaken due to the possible diagnosis of adrenal hemorrhagic cyst, based on her past history of a traffic accident.

The mean size of the tumors was 9.6 cm, ranging from 4 to 17 cm. The mean patient age was 49.5 years (range 20-75). There were 4 female and 4 male patients, among which 4 had a histopathologic diagnosis of hemangiomas, 1 of a lymphangiomatous cyst and 3 of myelolipomas. Table 1 summarizes the data from our series.

The first case concerned a 54-year-old male patient who was subjected to CT scan during the work up for atypical dyspeptic symptoms. A round, cystic lesion of 10 cm maximum diameter was recognized on the left adrenal gland, and the patient underwent open left adrenalectomy. Histopathology of the specimen revealed a cyst containing a thick, milky fluid, with a fibrous capsule with calcified areas, which was diagnosed as a lymphangiomatous cyst based on positive staining for VIII factor along with the aforementioned features (Figure 1a).

The second case concerned a 20-year-old woman who was subjected to CT due to persisting left flank pain. The CT scan demonstrated a large irregular lesion, consistent with a hematoma, and the patient underwent exploratory laparotomy. A lesion of the left adrenal gland was recognized and was removed. Histologic examination revealed a cystic lesion with 17 cm maximum diameter, consistent with a cavernous hemangioma.

The third case concerned a 50-year-old female patient, with known macronodular adrenal hyperplasia. On follow-up CT, a 4 cm lesion was detected on the left adrenal gland, and the patient was subjected to left adrenalectomy. The pathological examination showed a cavernous hemangioma (Figure 1b).

The fourth case concerned a 75-year-old female patient admitted in our department for management of a left breast tumor. The staging abdominal ultrasound revealed a left adrenal mass. A CT was performed which revealed a well-described, heterogeneous mass of 8 cm and a MRI scan followed which demonstrated irregular pe-

Table 1. Details of patients

Case	*Age (years)-Sex*	*Radiologic method*	*Size (Max. diameter-cm)*	*Symptoms*	*Operation*	*Histopathologic diagnosis*
1	54-Male	CT	10	Atypical dyspeptic symptoms	Open left adrenalectomy	Lymphangiomatous adrenal cyst
2	20-Female	CT	17	Persisting left flank pain. Previous history of traffic accident	Open left adrenalectomy	Cavernous adrenal hemangioma
3	50-Female	CT	4	Incidental finding on CT follow up for nodular adrenal hyperplasia	Laparoscopic left adrenalectomy	Cavernous adrenal hemangioma
4	75-Male	CT and MRI	8	Staging imaging for breast cancer	Open left adrenalectomy	Cavernous adrenal hemangioma with extra-medullary hemopoiesis areas
5	57-Male	CT	7	Atypical abdominal pain	Open right adrenalectomy	Adrenal myelolipoma
6	40-Female	US and CT	15	Investigation of splenomegaly and anemia	Open right adrenalectomy	Adrenal myelolipoma
7	45-Female	CT	10	Hypertension and increased urinary metanephrines	Open right adrenalectomy	Adrenal myelolipoma (No pheochromocytoma found)
8	55-Male	CT, MRI and FDG-PET scan	6	Incidental finding during work up of chronic kidney disease and mediastinal lymphadenopathy	Laparoscopic right adrenalectomy	Cavernous adrenal hemangioma

CT : computerized tomography, MRI : magnetic resonance imaging, US : ultrasound, PET: positron emission tomography

ripheral enhancement. As metastases could not be excluded, fine needle aspiration cytology was performed which was not diagnostic. The patient underwent modified radical left mastectomy and left adrenalectomy. The breast lesion was diagnosed as adenocarcinoma, and the adrenal lesion as cavernous hemangioma, characterized by dilated vascular spaces with thin endothelial lining (Figure 1c). Interestingly, areas of extramedullary hemopoiesis were also found, making this case unique in the relevant literature.

The fifth case concerned a 57-year-old male patient, who was subjected to a CT scan of the abdomen as part of atypical abdominal pain investigation. Imaging demonstrated a right retroperitoneal tumor of 7 cm, characterized by heterogeneous density and enhancement. Due to size and symptoms the patient underwent open right adrenalectomy and the pathology examination revealed a mass consisting of mature adipose tissue with nests of bone marrow cells, consistent with myelolipoma (Figure 1d).

The sixth case concerned a 40-year-old woman, who was subjected to abdominal CT scan, during the investigation of anemia. A 15 cm heterogeneous mass of the right adrenal gland was detected and the patient underwent open right adrenalectomy. Histology revealed a typical myelolipoma.

The seventh patient was a 45-year-old female, with increased levels of urine metanephrines, detected during investigation for severe hypertension. A CT scan of the adrenals followed which demonstrated a 10 cm mass on the right adrenal gland. The patient underwent open right adrenalectomy with a preoperative diagnosis of pheochromocytoma, yet the pathology revealed a typical myelolipoma. Interestingly, after the operation the urine metanephrine levels returned to normal and blood pressure decreased.

Finally, the last case was a 55-year-old male who was incidentaly diagnosed with a right adrenal mass following an ultrasound scan for the investigation of newly diagnosed chronic kidney disease. A CT scan revealed a 8-cm mass in the right adrenal with radiologic characteristics of

malignancy. This was in addition to mediastinal and paraaortic lymphadenopathy. A MRI scan also showed the same findings. The patient did not have any clinical or biochemical finding of hormonal hypersecretion. A PET/CT scan showed no uptake in the adrenal lesion or the lymph nodes. Following this detailed workup the patient was subjected to right laparoscopic adrenalectomy and pathology revealed a cavernous hemangioma of the adrenal gland.

Discussion

Adrenal glands are sites of origin of many different neoplasms. Apart from the more clinically frequent adenomas, pheochromocytomas, adrenocortical carcinomas and metastases from other organs, many other types of tumors may be detected. Advances in imaging methods have increased the incidence of more rare lesions incidentally found in the adrenal glands. Lesions that belong to this category are ganglioneuromas, adrenal cysts, myelolipomas, benign mesenchymal tumors, adenomatoid tumors, primary melanomas, hematolymphoid neoplasms and others [1,2].

It is very important to be able to differentiate these benign conditions from malignant adrenal tumors (i.e. adrenocortical carcinomas and malignant pheochromocytomas), as their management can be more conservative. However, their characteristics can prove to be quite misleading [2,5,6].

Adrenal cysts are divided into 4 subgroups. Pseudocysts comprise the majority of adrenal cystic lesions and are considered as post-injury vascular anomalies, characterized by hemorrhage, fibrosis and hemosiderin deposition. They lack cellular lining in their wall, and patients may present with vague symptoms due to mass effect or hypertension when the renal artery is compressed [1,4]. Endothelial cysts differ from pseudocysts only by the presence of smooth, flat endothelial lining. They are further subdivided into lymphangiomatous and hemangiomatous cysts. True adrenal cysts are characterized by true endothelial or mesothelial lining, they are usually filled with serous fluid and are subcategorized in retention cysts, cystic adenomas and embryonal cysts. The fourth subgroup includes parasitic, mainly hydatid cysts. MRI is the most accurate imaging method for investigating such lesions which have to be differentiated from malignant adrenal tumors with cystic degeneration [4].

Myelolipomas are benign tumors characterized by the presence of hematopoietic elements in a substrate of adipose tissue [3]. They are thought to derive from bone marrow emboli to the small vessels of adrenal medulla, or metaplastic change of the reticuloendothelial cells of blood capillaries. They are usually asymptomatic but may manifest with atypical abdominal pain. When they are large and symptomatic they should be removed, as there is a potential for rupture and hemorrhage [7]. CT is the imaging study of choice, due to their characteristic fatty element. MRI may be needed in order to delineate their nature and confirm that their origin is the adrenal gland and not some other tissue of the retroperitoneum [1]. Finally, there are reports [8,9] of these tumors occurring in combination with syndromes of cortisol or catecholamine hypersecretion.

Hemangiomas are rare neoplasms, which are usually asymptomatic and are typically cavernous or capillary. Their incidence is about 0.01% and they affect predominantly women [1]. As in the case of cystic lesions, MRI is the most accurate imaging method, although they are usually found in CT scans and due to size and the presence of calcifications, malignancy cannot be excluded [10].

The cases presented herein were diagnosed over a long period of time in our hospital and as a result the investigations that were performed had the limitations of their time. In our days adrenal CT scan is much accurate in differentiating adrenal tumors. In addition, the performance of FDG-PET scan has revolutionized the diagnosis of malignant tumors. Nevertheless, even with the use of these modalities, rare adrenal tumors can still be misdiagnosed. One should, however, be aware of the incidence of these lesions, as they can appear as big-sized abnormal tumors that can mimic malignant tumors. The fact that they can be asymptomatic, or diagnosed incidentally poses a real clinical dilemma concerning their management. Asymptomatic tumors could be followed-up, if their benign nature can be confirmed radiologically. Evolution of imaging modalities can aid in the differential diagnosis of these tumors, contributing thus to their management, always considering the possibility of malignancy, primary or metastatic.

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