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Retroperıtoneal Angıomyolıpoma Mımıckıng Adrenal Cortıcal Neoplasm. An Unusual Localızatıon

Angıomıolıpoma Retroperıtoneal que Sımula una Neoplasıa Cortıcal Suprarrenal. Una Ubıcación Inusual

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INTRODUCTION

Angiomyolipoma are relatively rare benign mesenchymal neoplasms that derive from perivascular epitheloid cells.[1] These tumors may occur in a sporadic form or as a part of systemic diseases especially with tuberosclerosis. [2] Compatible with its name, tumor consists of three components: smooth muscle cells, adipose tissue, and blood vessels. It is mostly seen in kidney, and rare extrarenal lesions mostly involve liver.[3] Extrarenal retroperitoneal angiomyolipomas (ERAML) are extremely rare. It poses a diagnostic problem by mimicking other retroperitoneal tumors.

In our current study, we reported an AML located adjacent to left adrenal gland mimicking adrenal neoplasm.

CASE HISTORY

A 53-year-old female patient was referred to our outpatient clinic with an incidentally diagnosed left adrenal mass. Her medical history revealed breast carcinoma, dysritmia and hypertension. She underwent mastectomy 9 years ago and was on follow-up with no recurrences. She was not also taking any medication for breast cancer. Initially she was admitted to the chest disease clinic with dyspnea and cough and her chest tomography revealed a left 3 cm solid adrenal mass. A full adrenal biochemical workup was performed (24 hour VMA: 2,85 mg/day; 24 hour normetanephrine 187mg/day; 24 hour metanephrine: 91.24 mg/day; noradrenaline: 26.56 mg/day; plasma renin activity: 3.06; cortisol dexamethasone 0.533; DHEA-S: 142mg/dL; aldosterone: 5.91 ng/dL). There was no sign for a hormonally active adrenal lesion. Adrenal MRI, and 18F-fluoro-deoxyglucose (FDG) PET/CT were performed. Adrenal MRI revealed a 35X28 mm lesion that was suggestive of an adrenocortical carcinoma or a metastatic primary tumor. **(Figures 1 y 2)**

FDG PET/CT demonstrated an abnormal FDG accumulation within the mass which was in the left adrenal region. Maximum standardized uptake value [SUVmax] 2.6 g/mL.

Due to her previous history of breast cancer and imaging properties, the initial diagnosis was thought to be metastatic adrenal tumor or primary adrenal carcinoma. After written consent taken from the patient, she underwent left sided laparoscopic adrenalectomy. A well- shaped spherical encapsulated ~3.5 cm solid adrenal mass separate from the left kidney was resected. **(Figure 3)**

The lesion was surgically seemed to be related with adrenal gland and did not worry us for a non-adrenal lesion. On the other hand, macroscopic evaluation of the specimen revealed 2.5 cm well shaped mass adjacent to the adrenal gland. The whole specimen was nearly 6x3x2.5 cm. The histopathological examination revealed that the lesion was angiomyolipoma and consisted of mature adipose tissue, thick-walled blood vessels and smooth muscle in different proportions **(Figures 4 y 5)**. HMB-45 and Desmin antibodies were also used

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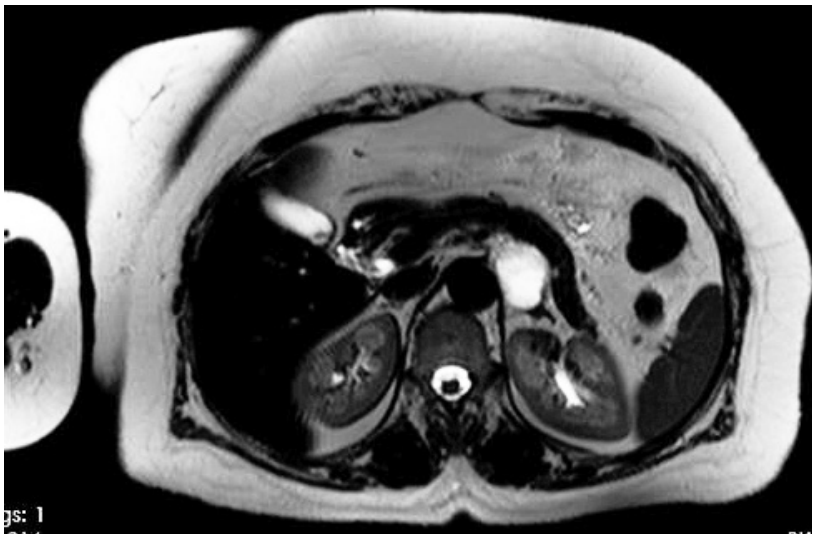

Figure 1. Heterogeneous mass is seen that is of high signal on T2 sequences

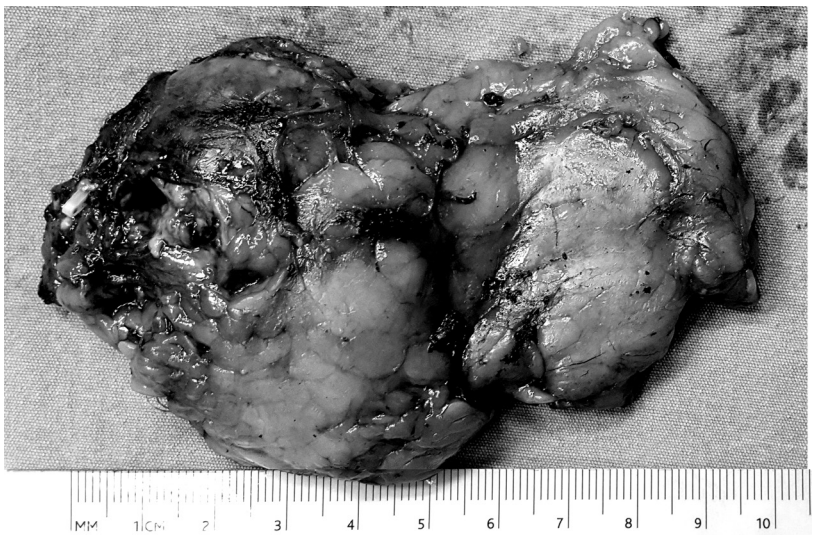

Figure 2. Heterogeneous circumferential contrast enhancement is seen with administration of gadolinium. There was no macroscopic fat within the lesion

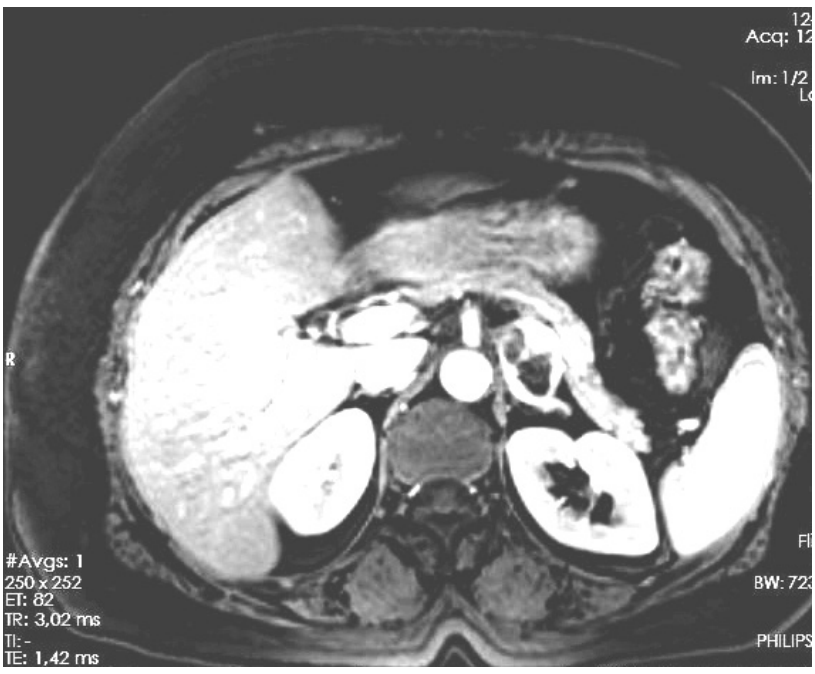

Figure 3. Macroscopic view of the resected specimen

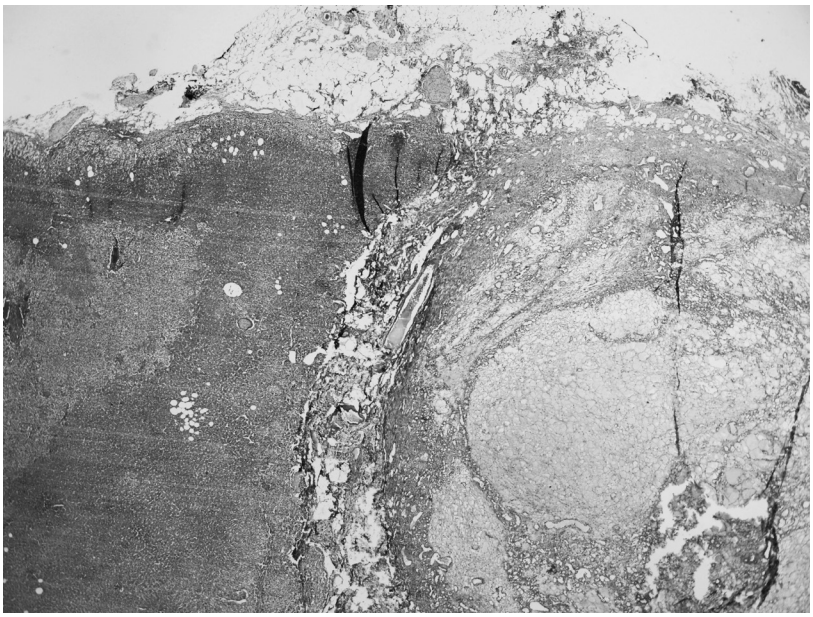

Figure 4. Histopathological examination demonstrating the distinction between tumor (on the right side) and adrenal gland (on the left side) (x40)

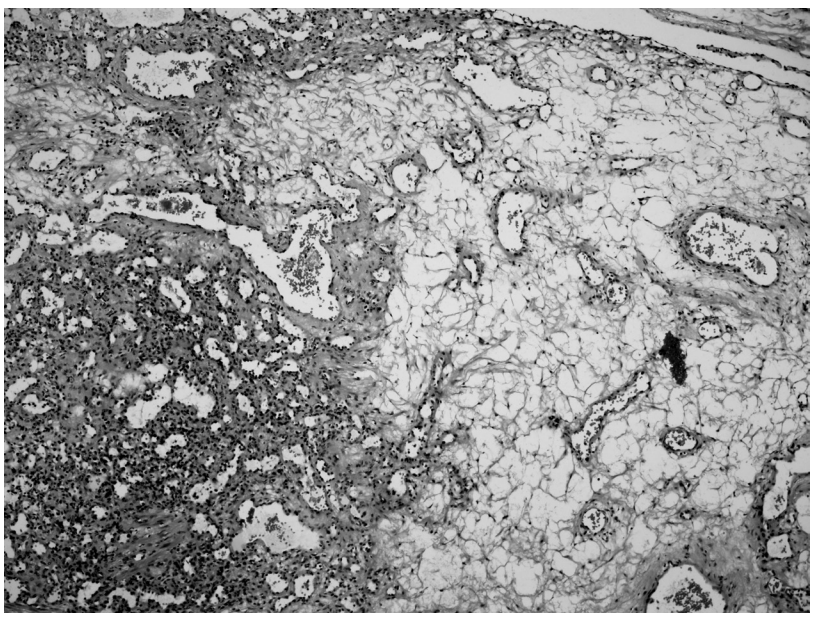

Figure 5. Histopathological view of the adipose structures and thick walled blood vessels (x100)

immunohistochemically to confirm the diagnosis.

Postoperative period was uneventful, and the patient discharged on postoperative 2nd day. Her postoperative 1st year of follow-up was also insignificant.

DISCUSSION

AMLs are uncommon lesions of perivascular epithelioid cells. Its incidence in general population is 0.013%.[4] Dysmorphic blood vessels, epithelioid smooth muscle and adipose cells are three components of the tumor. The diagnosis is based on its radiological characteristics and histopathological properties. Immunohistochemical stains with HMB-45 and MAR-1/Mela A antibodies are also helpful.[4] AMLs mostly occur in kidney but occasionally can be found in extrarenal sites. To date 30 cases of retroperitoneal origin has been reported but none of them included periadrenal adjacent area.[5] Most of the reported cases were originated from the perinephric region of the retroperitoneal area. As in our case most patients reported in the literature were female. Out of 30 cases reported in the literature 25 (84%) cases were female.[5] Extrarenal retroperitoneal AMLs (ERAML) may present in a variety of clinical

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symptoms and signs. Most of them are related to the place and organ involved. In our case the patient had no symptoms or signs. Her chest clinic visit due to dyspnea and cough revealed an incidental adrenal mass. Imaging modalities for such adrenal lesions are crucial for preoperative diagnosis and guiding for surgery. Computed tomography and especially MRI are the most preferred imaging modalities for evaluating these lesions[5]. Although imaging modalities guide surgeon for preoperative evaluation, the value of histopathological examination is undeniable. In our case complete workup including biochemical workup and imaging modalities (adrenal MRI and FDG PET Ct scan) directed us for a metastatic adrenal tumor or an adrenocortical carcinoma. There was no sign for a non-adrenal lesion. So, we did not consider performing a percutaneous image-guided biopsy. The lesion was removed laparoscopically for histopathological examination.

CONCLUSION

ERAMLs are rare tumors and retroperitoneal region is an extremely rare location. The diagnosis can be challenging despite complete biochemical and imaging workup. Histopathological examination and complete resection preserving the adjacent organs is crucial in the management of suspicious lesions.

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