A Case of Retroperitoneal Liposarcoma Mimicking an Adrenocortical Carcinoma

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Abstract

An adrenal mass can be a diagnostic challenge as it is not easy to differentiate the adrenal glands from other adrenal pseudotumours with only radio-imaging. We report a 28-year-old patient who was diagnosed radiologically as an adrenal cortical carcinoma after he presented with abdominal pain and fullness. Biochemically, he demonstrated secondary hyperaldosteronism. Intra-operatively there was a huge mass, inferior to a normal right adrenal, which was histopathologically proven to be a dedifferentiated liposarcoma.

Key words: adrenal pseudotumour, dedifferentiated liposarcoma, histopathology

INTRODUCTION

The investigations of an adrenal mass include assessing its functionality and its potential to be malignant. Often large adrenal tumours (LATs) point to an adrenal cortical carcinoma, especially if patients present with features suggestive of hormonal excess. Preoperative investigations, such as radio-imaging, is integral in establishing a preliminary diagnosis, as well as to provide essential information in formulating a management plan. However, as the adrenals are bordered by various anatomical structures, at times adrenal pseudotumours may be misinterpreted as adrenal pathologies. A large adrenal pseudotumour >4 cm, might be interpreted as an adrenocortical carcinoma if the patient is hypertensive or exhibits hypercortisolism.

CASE

A 28-year-old male who was recently diagnosed as hypertensive for the past 1 year but not on treatment, presented with 1-month history of abdominal pain and fullness associated with nausea, vomiting, and significant weight loss in the preceding three months. Clinical examination revealed blood pressure ranging from 130-140/80-90 mmHg, with presence of a vague mass at the right lumbar region. There were no features suggestive of Cushing’s syndrome or phaeochromocytoma. Abdomen ultrasound demonstrated a suprarenal mass measuring 13 cm x 12.5 cm x 14 cm. This was confirmed by a CT scan, which showed a right suprarenal mass, likely of adrenal origin, measuring 14 cm x 12 cm x 15 cm with compression of the inferior vena cava, right renal vein and right renal artery. Biochemically, there was evidence of secondary hyperaldosteronism with raised plasma renin activity and serum aldosterone, possibly due to compression of the renal vasculature by the mass. His serum electrolytes, DHEA-Sulphate, urine catecholamines and steroid profiles were normal (Table 1).

A month later, he presented with abdominal pain and fullness, suggesting the possibility of an enlarging adrenal mass. Adrenal CT revealed an enlarged mass measuring 15.8 cm x 14.4 cm x 17.9 cm with local infiltration to the right kidney. There were hypodense areas within the tumour, representing areas of necrosis (Figure 1A and 1B).

Due to the rapid progression of the size of the tumour, a right adrenalectomy was performed. However, intraoperatively, a huge peritoneal mass (16 cm x 14 cm x 11 cm) was noted inferior to the normal right adrenal gland, with a normal-looking right kidney (i.e., no evidence of tumour invasion). Both the tumour and right adrenal were removed (Figure 2A and 2B).

Histopathological examination of the tumour revealed a FNCLCC (Fédération Nationale des Centres de Lutte Le Cancer) grade 2 dedifferentiated liposarcoma (Figure 3A and 3B). Sections of the tumour show a

Table 1. Biochemical investigations results of the patient

<table>
<thead>
<tr>
<th>Parameters</th>
<th>Results</th>
<th>Normal Range</th>
</tr>
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<tbody>
<tr>
<td>Plasma renin activity</td>
<td>3.08</td>
<td>0.30 – 1.90</td>
</tr>
<tr>
<td>Serum aldosterone</td>
<td>325.1</td>
<td>41.71-208.9 pg/ml</td>
</tr>
<tr>
<td></td>
<td></td>
<td>(supine) 67.40 – 335.1 pg/ml (upright)</td>
</tr>
<tr>
<td>DHEA-sulphate</td>
<td>10.44</td>
<td>0.44 – 13.4 µmol/L</td>
</tr>
<tr>
<td>24-hour urinary free epinephrine</td>
<td>19</td>
<td>&lt;21 mcg/24 hours</td>
</tr>
<tr>
<td>24-hour urinary free norepinephrine</td>
<td>136</td>
<td>15-80 mcg/24 hours</td>
</tr>
<tr>
<td>24-hour urinary free dopamine</td>
<td>451</td>
<td>65-400 mcg/24 hours</td>
</tr>
</tbody>
</table>

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Figure 1. (A) Suprarenal mass with hypodense areas displacing the right kidney postero-inferiorly (CT Abdomen axial view); (B) CT Abdomen (coronal view).

Figure 2. (A) Huge mass measuring 16 cm x 14 cm x 11 cm, weighing 1610.6 g, comparing to the normal right adrenal gland (4.0 cm x 3.5 cm x 1.3 cm); (B) Normal right adrenal measuring 4.0 cm x 3.5 cm x 1.3 cm.

Figure 3. (A) Dedifferentiated area composed of diffuse sheets of pleomorphic cells displaying large irregular nuclei with vesicular chromatin, inconspicuous nucleoli and moderate eosinophilic cytoplasm. Numerous bizarre and multinucleated cells are seen (H&E, x40); (B) Fluorescence in situ hybridization (FISH) analysis for MDM2 gene using MDM2/CEP 12 probe (green signal) (VYSIS), shows many nuclei with amplified signals (red signal), i.e., consistent with MDM2 gene amplification.
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We report the case of a middle-aged male patient that presented with a tumour mass in the right retroperitoneum. The patient was referred to the endocrine team after an MRI of the abdomen revealed a mass measuring 6 × 4.5 cm. The tumour presented as a non-metastasizing tumour composed of matured adipocytes and lipoblasts, with a well-differentiated liposarcomas (WDLPS), usually found retroperitoneally, are the most frequent tissue sarcomas.6

Large adrenal tumours (LATs), defined as adrenal masses with the size of 6 cm or more, are often rare with the incidence of 8.6% to 38.6%.1–3 The discovery of a LATs often indicates malignancies unless proven otherwise.2 A study by Mege et al., in 2014 reported that 64% of their LATs patients had malignancies, with 44%, 27% and 21% of these patients having adrenocortical carcinomas, adrenal metastases and malignant phaeochromocytomas respectively.4

Histologically, DDLPS is characterized by the abrupt transition from WDLPS to a region of non-lipogenic sarcoma. Under the microscope, the dedifferentiated area appears as atypical non-lipogenic stromal cells with hyperchromatic nuclei scattered in fibrous septa. Ninety percent of DDLPS arises de novo, while 10% occurs in recurrence. In recurrent tumours, dedifferentiation occurs in almost 20% of first time recurrences and 44% of second-time local recurrences, implying acquisition of additional aberrations within WDLPS as it recurs.14

The risk of dedifferentiation is higher in deep-seated tumours, especially in the retroperitoneum and is probably a time-dependent phenomenon.10 Often, DDLPS can be diagnosed easily through adrenal radio-imaging such as CT or MRI, with features often described as heterogenous, non-lipogenic with a region of abnormal-appearing fat.15 In cases where histological examination is equivocal, immunohistochemical staining of MDM2 (sensitivity 95%, specificity 81%) and CDK4 (sensitivity 92%, specificity 95%), allows a definitive diagnosis of DDLPS.16 The detection of MDM2 amplification and overexpression of MDM2 genes (100% of cases) and CDK4 (90% of cases) using FISH or quantitative PCR is highly specific for the diagnosis of DDLPS.10

Treatment of primary retroperitoneal DDLPS is surgery. Systemic therapy with chemotherapy or targeted agents should be considered if a surgical margin is not feasible or there is recurrence. Targeted therapies aimed at MDM2 and CDK4 oncogenes are still in clinical trials.4 Prognosis is determined by local recurrences (40-60%), especially in the retroperitoneum, despite the low metastatic potential (15-20%). There is an overall rate of 41% of local recurrence. The overall mortality ranges from 28-40% at 5 years.10,15 Retroperitoneal lesions have 100% local recurrence rate and almost invariably lead to death.

CONCLUSION

Presentation of an adrenal mass can pose a diagnostic challenge as it is difficult to differentiate an adrenal mass from other retroperitoneal masses (lymphomas, liposarcomas, ganglioneuromas, etc.) by using radio-imaging modalities due to the close proximities of various organs in a tight retroperitoneal space. Surgical resection is often necessary if the mass exhibits features suggestive of malignancy while a histopathological examination will provide a definite diagnosis. Retroperitoneal liposarcomas are often aggressive and may present to the endocrinologist?
as an adrenocortical carcinoma. Identification of the MDM2 and CDK4 genes via immunohistochemical staining, qualitative PCR and FISH is diagnostic.

Ethical Consideration
Patient consent was obtained before submission of the manuscript.

Statement of Authorship
All authors certified fulfillment of ICMJE authorship criteria.

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