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A case of adrenal myelolipoma mimicking pheocromocytoma

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Summary
A 34 year-old hypertensive para 2 + 0 (1 alive) woman, was found to have right suprarenal mass during routine ante-natal investigation. The suprarenal mass, which was initially diagnosed as phaeochromocytoma was excised and was found at histology to be myelolipoma! The radiological investigations and appearances (compared with phaeochromocytoma where necessary) have been discussed. The complications, differential diagnoses and treatment were also mentioned.

Keywords: *Adrenalmyelolipoma, pheocromocytoma, hypertension.*

Résumé
On a decouvert chez une femme hypertendue de 34 ans par 2 + 0 (1 vivant) une masse droite suprarenale lors des investigations pre-natalles de routine. La masse suprarenale, qui avait ete initiallement diagnostique comme pheochromocytome, était excise et on a deconvert ā l'hytologie que cétait la myelolipome. Les investigations et apparences radiologiques (compares au phecochromocytomes dés que necessoure) ont été discutées. Les complication, diagnostiques differentials et traitement ont aussi été mentionés.

Introduction

Myeoliopoma is a rare, benign, and biochemical nonfunctioning tumor which consists of mature fat interspersed with haemopoietic elements resembling bone marrow. Most patients are asymptomatic and the condition is discovered incidentally at autopsy [1]. Albeit possibly coincidental, there is frequent association with obesity, hypertension, and /or diabetic mellitus [2].

Wide experience about this tumour is difficult to obtain in one institution because the tumor is extremely rare, and a search through the medical literature revealed few case reports, which were discussed in relation to other nonfunctional tumours of the adrenals [3]. The case here presented posed a diagnostic riddle to the medical team managing the patient including the radiologists as it was initially thought that the patient had pheochromocytoma and was treated as such until the post adrenalectomy histology report gave the final daignosis of MYELOLIPOMA!

The rarity, coupled with the diagnostic challenge posed by this disease entity has prompted the authors to report this case to serve as a reminder to radiologists to include myelolipoma as a differential of an adrenal mass.

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Case report

A 34-year-old female banker presented with a two and a half-year history of elevated blood pressure, which was incidentally discovered during her first pregnancy. She had pre-eclampsia and had to be delivered by an emergency caesarian section at gestational age of 32 weeks. However the baby did not survive.

The patient was placed on antihypertensive medication, which she voluntarily stopped taking 3 months later. An initial ultrasound was requested because of the elevated blood pressure and this revealed a right suprarenal mass but the patient refused further investigation and treatment.

She again presented during her second pregnancy with elevated blood pressure and had to be placed on Aldomet and occasional doses of Hydralazine. She was delivered of a live baby at 34-week gestation. During this pregnancy there was no history of headaches, paroxysmal hot flushes or sweating, nor was there any history of palpitations or fainting attacks. She was not a known diabetic or asthmatic. Her family and social history revealed that she did not smoke or drink alcohol, but her mother was on treatment for hypertension.

On examination, she was a healthy looking, but grossly obese young woman. The pulse rate was 70/min, regular and of full volume. Blood pressure was 160/100mmHg and the heart sounds were normal. Review of the other systems was essentially normal. The electrolyte and urea, urinalysis, full blood count, packed cell volume the vanylmandelic acid (VMA) level and ECG were within normal limits. A working diagnosis of an adrenal mass? Pheochromocytoma of the right adrenal gland was made, and the patient was co-managed by a combined team of urologists and physicians.

The abdominal ultrasound scan examination showed a well-defined rounded echogenic mass superior to the right kidney and displacing the kidney infero-medially. The mass measured 9.2cm x 7.1cm (Fig. 1).

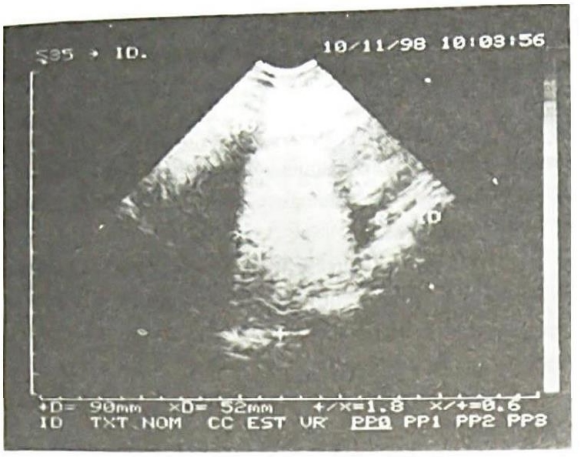

Fig 1: Ultrasound scan showed a well-defined echogenic right sura renal mass

The kidneys, the liver, and the spleen were however within normal limits. The intravenous urography showed prompt and satisfactory excretion of contrast bilaterally. There was slight displacement of the right kidney medially, and the pelvicalyceal system were minimally dilated. The left kidney and its pelvicalyceal system, and also the urinary bladder were within normal limits.

A pre-and post-contrast abdominal computerized tomographic scan done at 5mm intervals showed a hyperdense mass with cystic areas measuring 7.4 x 7.5 x 7.2cm superior to the right kidney and displacing the kidney inferiorly (Fig. 2).

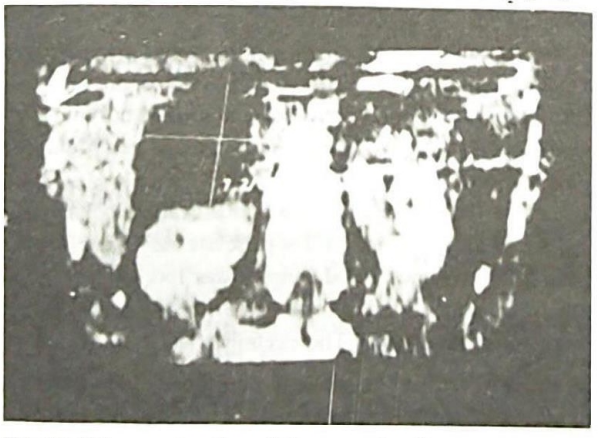

Fig 2: CT reconstruction of the mass showing a well-defined hypodense right supral renal mass. Both kidneys showed normal function, and the right displaced downwards

Both kidneys showed normal excretion and pelvicalyceal systems. The liver, spleen and pancreas were also normal. A diagnosis of a right suprarenal cyst was made.

At surgery a huge cystic yellowish right adrenal mass measuring 12cm x 10cm was found and a right trans-abdominal adrenalectomy was done.

She made a steady and satisfactory recovery post surgery. The blood pressure was still fluctuating when she was discharged home on the twelfth day post operative on Aldomet, and moduretics to be followed up in the out-patient clinic.

The histological findings as contained in the report: "A well-capsulated firm yellow brown mass that weighs 213g and measures 10.5 x 9 x 4.5cm was received. Section through the adrenal gland show compressed cortical tissue by mass that is composed of mature adipose tissue with interspersing haemopoietic cells and many megakaryocytes. The lesion has completely replaced the adrenal medulla. The compressed adrenal cortex contains normal vacuolated cells. The features are consistent with myelolipoma of the right adrenal gland"

Discussion

Non-functioning adrenal tumours constitute a broad category that is made up of both cystic and solid lesions, which include adenomas, carcinomas, metastases and myelolipomas.

Myelolipomas are rare benign tumours of the adrenal gland with varying proportion of adipose tissue and haemopoietic elements resembling bone marrow [4]. The tumour usually remains small and asymptomatic, and the condition is discovered incidentally at autopsy. Occasionally however they may reach massive proportions and then become symptomatic [1,5].

Myelolipoma manifests in four distinct clinicopathological patterns: isolated adrenal myelolipoma, adrenal myelolipoma with haemorrhage, extra adrenal myelolipoma and myelolipoma associated with other adrenal disease, where the patient may be having other adrenal mass, [4]. The case here presented appears to fit into the first category. It has now become obvious, that the patient's elevated blood pressure could not have been caused by the adrenal mass, which was initially thought to be phaeochromocytoma more so that the patients blood pressure remained high, post operatively.

Being a non-functional mass, patients with myelolipoma hardly present with any symptoms except when there are other associated adrenal masses, hence early diagnosis of this disease has posed a challenge to medical researchers in the recent past.

Myelolipoma is difficult or impossible to detect at plain radiography unless the lesion is large [4]. Before the development of CT, nephrotomography followed by arteriography was the accepted imaging approach in the evaluation of myelolipoma and indeed of adrenal mass be it functional or not.

The high resolution of newer sonographic equipment now affords a confident depiction of normal and pathologic adrenal gland. On ultrasound, myelolipoma is visualised as a hyperechoeic mass [6], and indeed this was also the ultrasound finding in the case here reported (Fig.1). However this appearance is not pathognomonic of myelolipoma as other adrenal masses can present in this way, particularly phaeochromocytoma [7]. This may have accounted for the initial misdiagnosis in this case.

Retrospectively, the diagnosis of phaeochromocytoma should not have been a first choice more so that the vanylmandelic acid (VMA) level was within normal limits. The level of VMA is very crucial in differentiating pheochromocytoma from other adrenal tumours [3,7].

Phaeochromocytoma is a functioning tumour while myelolipoma is a non-functioning tumour and a search through the literature did not reveal a single case of myelolipoma coexisting with hypertension therefore making this case a distinct form of presentation [1,3,6].

In the more recent past computerised Tomography (CT) has replaced earlier methods of diagnosis of adrenal tumours. CT frequently demonstrates large amounts of fat with areas of interspersed higher attenuation tissue [4,6] (Fig.2). The CT appearances of pheochromocytoma according to reviews of different articles and literature on adrenal mass by [7], vary considerably, while some produce attenuation coefficients similar to that of the renal parenchyma, others had low attenuation value. Whereas some were homogenous others were not, but have variable densities within the lesion. In other words the CT appearance of pheochromocytoma is not specific and therefore based on the CT finding alone pheochromocytoma could easily be confused with myelolipoma.

At magnetic resonance imaging, predominantly fatty areas usually have increased signal intensity on T1-weighted images and moderate hyperintensity complicated by marrow like elements in the corresponding regions on T2 weighted although the imaging appearance of myelolipoma is altered by the presence of haemorrhage [4].

Other differential diagnoses of myelolipoma include carcinomas, metastases and adrenocortical adenoma especially if it contains fat.

The major complication of myelolipoma is haemorrhage, when the mass reaches massive proportions they may become symptomatic. It has been advocated that surgical excision of myelolipoma is unnecessary unless the diagnosis is unclear or the lesion is symptomatic, and that asymptomatic nonhemorrhagic myelolipomas do not require therapy [4,5,8].

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