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Adrenal Myelolipoma - An Enigma

Amjad NM[1]*, Kassian J[1], Baharudin S[2] and Kalavathy R[2]

[1]Department of Surgery, International Islamic University Malaysia, Kuantan, Pahang, Malaysia
[2]Hospital Tengku Ampuan Afzan, Kuantan, Pahang, Malaysia

Abstract

Adrenal myelolipomas are rare neoplasms. They are benign in nature comprising mainly of adipose and myeloid tissues. Majority of these adrenal gland tumors are hormonally inactive. In view of their rarity, they are usually detected incidentally or misdiagnosed. Misdiagnosis is quite common and can lead to difficulty in management. We wish to report a case of adrenal myelolipoma that was misdiagnosed which fortunately did not result in a major issue. With the availability of advanced imaging techniques and minimally invasive procedures, these situations can easily be avoided. The main reason for highlighting this patient is to create awareness among both the surgeons and the radiologists.

Keywords: Duodenal tumour; Metastasis; Adrenal gland; Myelolipoma

Introduction

Adrenal myelolipomas are rare benign neoplasms composed of adipose tissues and myeloid tissue, as in normal bone marrow. They are mostly hormonally inactive tumors of the adrenal gland with a reported incidence of about 0.03% at autopsy [1]. In view of their rarity, they are usually detected incidentally or misdiagnosed. It is estimated that the frequency of Adrenal myelolipoma detected incidentally on imaging varies from 7% to 15% [2]. Misdiagnosis is quite common and can lead to difficulty in management. We wish to highlight a case of myelolipoma that presented a management dilemma in our unit.

Review Report

A 40-year old lady presented to our upper GI unit of the department of surgery with a 4-month history suggestive of gastric outlet obstruction. She had no complains of abdominal pain, early satiety or haematemesis. She did complain of loosing two kilograms in weight during this period, which she attributed to reduced intake due to her vomiting. There was no history of any significant medical illnesses in the past.

On examination she looked well, no pallor or signs of any nutritional deficiencies. Examination of the abdomen was unremarkable.

Upper GI endoscopy revealed a deformed D1 D2 junction with inability to pass the scope beyond this. No mucosal abnormality was detected. A biopsy was taken from this narrowed area, which was reported as normal duodenal mucosa.

Barium meal studies confirmed our endoscopy findings as shown in Figures 1 and 2.

As we were of the opinion that it was a tumor arising from the duodenum, we proceeded to carry out a CT scan without requesting for an ultrasound scan of the abdomen. The CT scan of the abdomen showed a growth at D1D2 junction with additional features suggestive of a secondary deposit in the right adrenal gland as shown in figures 3 and 4.

Decision was made to proceed with a laparotomy. At surgery the duodenal deformity did not appear to be a malignant lesion. It was mainly fibrotic thickening involving the antero-medial aspect of D1D2 abutting the head of the pancreas. There were few small lymph nodes less than 1 cm diameter in the suprapyloric region (Station 5).

As reported in the CT scan, the right adrenal gland was replaced by a localized encapsulated tumor.

We did not have facilities to carry out a frozen section examination on that particular day.

Considering the patient's age and the ambiguous nature of the duodenal lesion, a metastasis from the duodenal growth seemed a less likely diagnosis. In view of this, a decision was made not to proceed

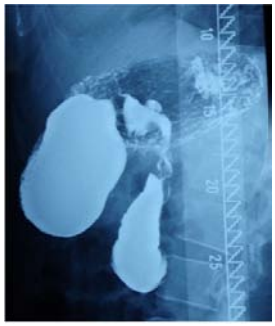

Figure 1: An eccentric irregular filling defect of about 3cm in length seen at the D1D2 junction with partial obstruction.

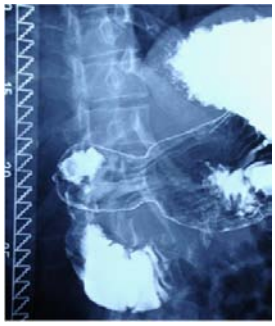

Figure 2: The irregular filling defect at the D1D2 junction with partial obstruction.

***Corresponding author:** Nasser Amjad, Department of Surgery, IIUM, Kuantan 25710, Pahang, Malaysia, E-mail: safah90@yahoo.com

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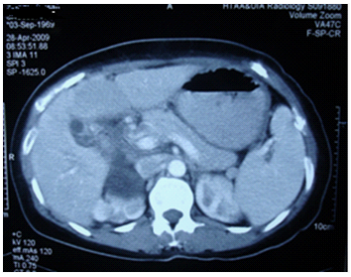

Figure 3: D2 mass with portal nodes. Shows a heterogeneous non-enhancing hypodense mass measuring 4 cm x 3.3 cm arising from the right adrenal gland. Features are of a duodenal growth with possible metastasis to the adrenal gland.

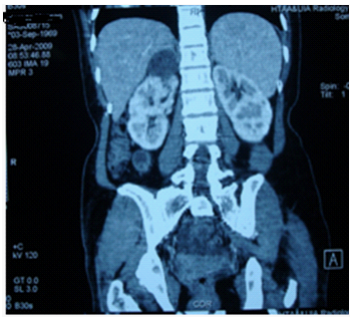

Figure 4: Sagittal view of possible metastasis to the adrenal gland.

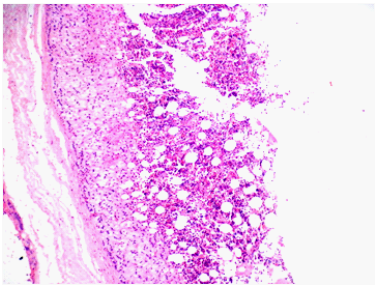

Figure 5: A bit of normal adrenal gland on the left and mature adipose tissue admixed with haemopoietic cells on the right (H&E stain, X40 magnification).

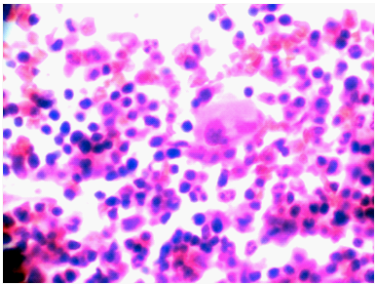

Figure 6: Haemopoietic cells of erythroid and myeloid precursors including megakaryocytes (H&E stain, X400 magnification).

with a radical operation but to carry out a gasto-jejunostomy, obtain biopsies of the lesion, lymph nodes and to remove the right adrenal mass. Post-operative recovery was uneventful.

The duodenal biopsy was reported as chronic inflammation, the lymph node as reactive hyperplasia and the adrenal mass as adrenal myelolipoma.

Figures 5 and 6 shows the histologyslides of the adrenal mass.

Discussion

Gierke first described a lesion with features of Adrenal Myelolipomasin in 1905 [3], but it was only in 1929 that it was recognized as a distinct lesion called myelolipoma by Oberling [4].

The aetiology of these tumors remains unclear. Theories include autonomous proliferation of bone marrow cells transferred during embryogenesis, degeneration of epithelial tissue of the adrenal cortex, metaplasia of mesenchymal cells and bone marrow emboli [5]. The most widely accepted theory is that is a differential proliferation of undifferentiated mesenchymal stem cells into myeloid and adipose tissue occurring in response to stimuli such as necrosis, infection or stress [6].

They are usually unilateral and asymptomatic. Right side lesions like in our patient are commoner than the left. Majority of these are nonfunctional but may present with symptoms either due to retroperitoneal hemorrhage or compression of local structures due to large bulky tumors. Endocrine disorders like Cushing Syndrome and hyperaldosteronism has been very rarely reported in a few cases [7]. Myelolipoma are benign and no malignant transformation has been identified [8].

Adrenal myelolipoma have been associated with common medical conditions like obesity (25%), hypertension (26%) and diabetes mellitus (26%). They have also been associated with chronic inflammation; burn injuries, and various cancers like renal cell carcinoma and uterine cervical carcinoma [6,9,10].

Benign adrenal myelolipomas were earlier detected only by chance at autopsy with an incidence of 0.08 to 0.2% [11] but advances in imaging modalities has resulted in a dramatic shift in the detection and management of these strange tumors.

The radiological appearance of a myelolipoma depends on the histological components of the tumor. On ultrasound examination it appears hyperechoic if it contains predominantly fatty components and heterogeneous or hypoechoic if the myeloid cells predominate. In our patient an ultrasound scan was not done. As noted ultrasound scan features of myelolipoma are not as specific as CT scan or MRI and as such one of them should be obtained for further evaluation.

Fat component of myelolipomas appear low attenuating on CT, and hyper-intense in both T1-weighted and T2-weighted MRI. The diagnosis can be made confidently when CT scan density measurements confirm discrete regions of fat attenuation (30 to100 HU) within an adrenal mass [12]. Presence of gross fat (attenuation <30 HU) is diagnostic [7]. Calcifications are rarely present and can be related to previous haemorrhage. Presence of large amounts of hematopoietic tissue only, with hardly any adipose tissue make adrenal myelolipomas difficult to distinguish from other adrenal tumors or metastatic deposits on imaging as was seen in our patient. Minimally invasive endoscopic ultrasound guided or percutaneous biopsy techniques could be utilized to establish a diagnosis.

Chemical shift MR imaging is used to differentiate adrenal adenomas from metastases. MRI may demonstrate signal intensity similar to fat on all sequences without significant loss of signal on opposed phase MRI. As myelolipomas contain abundance of adipose

tissue, the signal loss does not occur on out-of-phase images, but any signal loss noted is comparable to fat or fat-suppressed images [1].

Myelolipomas has been categorized into three types based on their MRI features [13]. First type is the predominantly fat containing tumors that appear homogeneous. The second type are the ones which appear as localized enhancing lesions due to presence of mainly myeloid cells and the third type has a heterogeneous appearance when both fatty and myeloid elements present.

Current imaging modalities like PET scans can be used to distinguish adrenocortical neoplasms from non-adrenocortical tumors including metastasis to the adrenal with high specificity (89%) and sensitivity (96%) [14].

The differential diagnosis of adrenal myelolipoma includes retroperitoneal lipoma, teratoma, liposarcoma, exophytic renal angiomyolipoma, adrenal adenoma, primary adrenal malignancy and adrenal metastasis as in our patient [14].

Elective surgery for myelolipoma is carried out more frequently now due to the ability to diagnose these lesions preoperatively utilizing the new imaging modalities and minimally invasive biopsy techniques.

An endocrine evaluation is recommended in all patients with adrenal masses. This should include plasma cortisone, Adreno Cortico Tropic Hormone (ACTH) and aldosterone levels. In addition plasma-renin activity too should be assessed. These measures are required to exclude any endocrine activity prior to embarking on intervention.

Asymptomatic cases with characteristic features of myelolipomas on imaging can be managed conservatively with serial scans to detect any changes that would warrant surgical intervention. Surgical resection is the treatment of choice when the diagnosis in doubt or the patient is symptomatic.

Conclusion

Adrenal myelolipomas are rare benign mostly hormonally inactive neoplasms. Clinical awareness of this lesion can prevent misdiagnosis and inappropriate treatment as nearly happened to our patient. Recent advances in imaging modalities have greatly improved the chances of making a positive diagnosis preoperatively.

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