Case report

An unusual presentation of an adrenal tumour–a case report

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ABSTRACT

Introduction and importance: Adrenal myelolipomas are rare, benign tumours with an incidence of 0.08-0.2%. They present between the fifth and seventh decade of life [1].

Case presentation: Our patient presented with complaints of vomiting and left lumbar pain of four weeks duration. Blood work revealed dyselectrolytemia. Contrast enhanced computed tomography of the abdomen and pelvis confirmed the diagnosis and the patient was planned for an adrenalectomy. Histopathology report revealed the pathology. She is currently on routine follow up and is disease free. Written informed consent was obtained from the patient for publication of this case report and its accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request. This case report has been reported in line with the SCARE criteria [2].

Clinical discussion: With the increased use of imaging modalities of the abdomen, they are now considered to be the second most common cause of adrenal incidentalomas (6-16%) [3]. Most tumours are small, asymptomatic and often go undiagnosed. Large tumours can cause chronic pain and other nonspecific symptoms.

Conclusion: Though myelolipomas are identified on routine CT scans, on a background of dyselectrolytemia, a further evaluation is of utmost importance to rule out the possibility of a functioning tumour.

1. Case description

An elderly asthmatic and hypertensive lady walked in to the outpatient department with complaints of left sided lumbar pain which was intermittent and radiating to the back for four weeks. The pain was associated with three to four episodes of non-projectile, non-blood stained vomitus. Her comorbidities were well controlled on medication. No relevant drug, family or genetic history. General examination revealed mild dehydration. Abdominal examination revealed mild tenderness in the left hypochondrial and left lumbar regions. Systemic examination of the lungs and spine were unremarkable. Serum electrolytes revealed dyselectrolytemia (Na + = 128 mEq/L, K + = 2.8 mEq/L) and elevated urea (50 mmol/L) and creatinine (2.80 mg/dL) levels. Urine examination was found to be normal.

Ultrasonogram abdomen revealed a well-defined hyperechoic lesion (4.5 x 3 x 3.5 cm) arising from the left suprarenal gland. Serum cortisol (17.60 μg/dL), urinary VMA (11.4) and serum metanephrines (243.26 μg/dL) were within normal limits; the possibility of pheochromocytoma and Cushing syndrome were excluded from the differential diagnosis.

Contrast enhanced CT abdomen and pelvis confirmed the size and origin of the mass (Fig. 1).

The mass was abutting the upper pole of the left kidney, spleen and the left psoas muscle. Upon administration of contrast no focal enhancement seen with a mean HU of –28 to +10 in arterial, venous and delayed phases. There were no features of calcification/hemorrhage/necrosis within the mass and no fat stranding surrounding the mass.

The patient was hydrated adequately to normalize her electrolytes and renal function. With a pre-operative diagnosis of a non-functioning left adrenal mass, anaesthetic fitness was obtained and was planned for a left adrenalectomy under general with epidural anaesthesia. The surgical oncology team performed the two hour surgery.

Left subcostal incision made. Left adrenal vessels identified, ligated and the adrenal gland was removed in toto without spillage. Specimen was sent for histopathological examination (Fig. 2). Liver, peritoneum and omentum were normal and no lymph nodal enlargement noted. Post-operatively, patient was managed with antibiotics, analgesics and DVT prophylaxis. Patient discharged on POD five. Follow up advised at two weeks, one, three and six months.

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Histopathology report revealed an adrenal gland neoplasm composed of mature adipocytes admixed with areas of congestion and hematopoietic elements without evidence of dysplasia (Fig. 3). The report confirmed the mass to be an adrenal myelolipoma.

2. Introduction

Benign tumours of the adrenal glands are either, most commonly, adrenocortical adenomas or myelolipomas. Rarely a benign adrenal tumour produces excess amounts of endogenous hormones are said to be active or functioning tumours. Nonfunctioning tumours that do not secrete these hormones [4]. Because of their asymptomatic presentation, these tumours are referred to as incidentalomas. The incidence of adrenal myelolipomas are on the rise with the rising use of imaging modalities to guide clinicians. Tumours that are present extra-adrenally (thorax, pelvis, liver, spleen, stomach, mesentery) are termed as extra adrenal myelolipomas.

2.1. Epidemiology/etiology/pathogenesis

The exact etiology is unknown. Apart from stress and dietary habits, three theories have been proposed that explains the etiopathogenesis of the disease. Patients with obesity, hypertension, atherosclerosis and hereditary conditions (thalassemia) have adrenal myelolipomas [5]. One theory suggests that when the adrenal gland is exposed to a stimulus (necrosis or inflammation), the reticuloendothelial cells undergo metaplastic changes that lead to the development of the tumour. Another hypothesis states that adrenal cortical adipocytes become inflamed, which releases cytokines and causes recruitment of hematopoietic progenitor cells which deposits in the adrenal cortex [6]. The last theory suggests that ACTH has an effect on the formation of myelolipomas. Patients with Cushing syndrome, characterized by excess ACTH production, have adrenal myelolipomas, thereby validating the hypothesis [6].

2.2. Clinical presentation and diagnosis

Adrenal myelolipomas are asymptomatic and are usually incidentally diagnosed on imaging. They are symptomatic when the size of the tumour increases. Palpable abdominal mass, fever, left hypochondrial and flank pain are the most common presenting symptoms [6]. Tumours with a size greater than four cm and composed of more than 50% fat can result in retroperitoneal hemorrhage. Nausea, vomiting, hypotension and anemia are the common presenting complaints in these patients [7]. Functional tumours present with endocrine dysfunction syndromes like Cushing syndrome, virilization and primary hyperaldosteronism [6]. Extra-adrenal myelolipomas are most commonly found in the pelvis, perineum, intraspinal region, thorax, retroperitoneum, stomach, spleen and mesentery. Based on their location and size, tumours cause a mass effect on the surrounding structures [8].

Ultrasound, CT and MRI are the common radiological modalities used to diagnose adrenal myelolipomas. Fine needle aspiration cytology can be done when in a diagnostic dilemma [8]. Serum cortisol, aldosterone and androgen levels are indicated in functioning tumours. However, it must be done routinely before surgery. Imaging of the thorax rules out extra-adrenal myelolipomas [7]. Ultrasound of the abdomen will reveal a heterogenous mass that is made up of fat and myeloid tissue. Complicated myelolipomas reveals focal areas of necrosis, hemorrhage and calcification [6]. CT abdomen and pelvis reveals a well circumscribed heterogenous lesion (HU -90 to -129). Tumours with a high content of fat and myeloid tissue exhibits a “smoky” appearance (HU +10 to +20). This differentiates the tumour from the surrounding retroperitoneal fat on imaging. Owing to the negative attenuation values provided on CT scans, it is said to be effective in diagnosing adrenal myelolipomas [6]. MRI provides better delineation
of the mass and is considered as a part of surgical work-up [6]. Biopsy is indicated in cases to rule out the possibility of pheochromocytoma and inconclusive radiographic findings of a benign adrenal mass [9].

2.3. Treatment

Treatment depends on the size and the presenting symptoms. Small (less than 5 cm), asymptomatic and nonfunctioning lesions can be managed non-operatively. They are monitored with regular imaging for up to one to two years. Literature states that tumours more than seven cm should be operated on. Patients presenting with features of hypotension and cardiovascular instability must be treated as an emergency basis [6]. Conventional anterior transabdominal approach was considered for large tumours with compression of the surrounding structures but extraperitoneal approach aids in faster post-operative recovery. Recent studies state that laparoscopic adrenalectomy is the gold standard [10]. Adrenal sparing surgeries prevent complications of adrenal insufficiency and lifelong adrenal hormone supplementation [11]. Hand assisted techniques allow for better visualization, traction and securing hemostasis.

3. Conclusion

Though myelolipomas are identified on routine CT scans, on a background of dyselectrolytemia, a further evaluation is of utmost importance to rule out the possibility of a functioning tumour. Minimally invasive techniques are to be considered in dealing with these tumours regardless of size for better post-operative recovery of the patient.

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Ethical approval

Informed consent was obtained from the patient for publication of this case report.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

This has been mentioned in the manuscript under the heading “abstract”.

Research registration (for case reports detailing a new surgical technique or new equipment/technology)

Not applicable.
This is a case report.

Guarantor

Prof. Dr. A.C Senthilkumar

CRediT authorship contribution statement

Dr. A. C. Senthilkumar: Chief surgeon who operated on this case.
Dr. Sivaram Sridharan: 1st assistant, writing the paper.
Dr. Swetha Vennelaganti: revision of written work, spell check.

Declaration of competing interest

No financial/personal relationships/conflicts of interest.

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