A rare case of unilateral giant adrenal myelolipoma

Adrenal myelolipoma is a rare, nonfunctional, benign neoplasm of the adrenal gland which is composed of fat and trilineage hematopoietic tissue. In the past, adrenal myelolipoma was detected at autopsy; however, at present, with increased use of radiological imaging such as ultrasonography, computed tomography, and magnetic resonance imaging, the incidental detection has become more common. Most of the adrenal myelolipomas are small (<5 cm). Larger myelolipomas, known as giant myelolipoma, are rare. We report a case of unilateral giant adrenal myelolipoma in a 52-year-old male which was managed by open adrenalectomy.

Key words: Adrenal, Incidentaloma, Laparoscopy, Myelolipoma.

INTRODUCTION

Adrenal myelolipoma is a tumour-like lesion which is composed of variable amounts of bone marrow elements and mature adipose tissue. However, it does not serve as a source for haemtopoiesis. It was first described by Giercke in 1905 and the term was coined by Oberling 24 years later. It is a rare benign lesion known to affect men and women equally in their fifth to seventh decades. The true incidence is unknown. They are usually unilateral and small; though bilateral lesions have also been reported. Although benign, these lesions can be a cause of dilemma for the treating urologist. We describe a case of unilateral giant adrenal myelolipoma in a 52-year male.

CASE REPORT

A 52-year-old male was admitted with complaints of the right-sided abdominal pain on and off for 6 years. There was no history suggestive of urinary tract infection, lower urinary tract symptoms, and any comorbid illness. At admission, his vitals were stable and abdomen was soft; there was no palpable lump. His complete blood count, coagulation profile, and liver and renal function tests were normal. Serum levels of metanephrine (20.8 pg/ml), normetanephrine (49.9 pg/ml), cortisol (10.67 µg/dl), aldosterone (42.2 pg/ml), plasma aldosterone-renin ratio (0.3), and plasma renin activity (0.14) were in the normal range. Abdominal ultrasound and computed tomography (CT) scan suggested a well-defined 10 cm × 5 cm lobulated mass arising from medial limb of the right adrenal gland, anterosuperiorly in contact with inferior surface of the liver and inferiorly in contact with anterior surface of the right kidney. Bilateral kidneys were normal (Figure 1a and 1b). Open right adrenalectomy (because of large tumor size) was done (Figure 2). Histopathology revealed adrenal myelolipoma.
Sharma, et al.: Myelolipoma of right adrenal gland

Figure 1: (a and b) Computed tomography scan showing the right suprarenal mass.

Figure 2: Cut section of the excised specimen.

DISCUSSION

Adrenal myelolipoma was first described by Giercke in 1905, but the term “myelolipoma” was coined by Oberling some 24 years later in 1929.[1,2] It is a lesion in the adrenal gland composed of variable amounts of mature adipose tissue and all three hematopoietic lineages of the bone marrow, i.e., white blood cells, red blood cells, and megakaryocytes.[3] However, it does not serve as a hematopoietic source.[4] Most are small (<5 cm), but giant myelolipomas, although rare, have also been reported. Small myelolipomas do not require surgery and can be followed annually with imaging. Larger myelolipomas present a risk of adrenal hemorrhage and require surgical excision.

Patients with adrenal masses can present in four different scenarios[4]

i. Patients who present with endocrinological symptoms, such as virilization or Cushing’s syndrome as seen in selected adrenocortical adenomas and carcinomas. Patients with pheochromocytomas or Conn’s syndrome present with hypertension, flushes, and headaches

ii. Patients who present with nonspecific symptoms such as pain, abdominal lump, fatigue, and weight loss, which on investigation, turn out to be an adrenal mass

iii. During workup of other malignancies, adrenal metastases may be detected

iv. Adrenal mass which is detected incidentally, the so-called adrenal incidentaloma.

The finding of an adrenal mass should prompt a diagnostic workup to differentiate between benign and malignant lesions.[8] The differential diagnosis of adrenal incidentaloma is nonfunctioning adenoma, functioning adenoma, pheochromocytoma, adrenocortical carcinoma, and metastasis.[9] Rare diagnosis is myelolipoma, lymphoma, liposarcoma, schwannoma, ganglioneuroma, hematoma, cavernous hemangioma, and various cysts of adrenal gland.[10]

The majority of adrenal myelolipoma cases are found either incidentally during workup for other reasons or at autopsies.[5] Systematic analysis of the true incidence of adrenal myelolipoma has not been done.[5] The incidence of detection of adrenal myelolipoma at autopsy ranges from 0.08% to 0.4% as per Olsson et al.[5,6] As per the recent series, adrenal myelolipomas account for about 10% to 15% of incidentalomas.[5] It usually presents between 50 and 70 years of age and there is no sex predominance.[9] Adrenal myelolipomas are benign tumors and are often small and asymptomatic; however, larger tumors may present with nonspecific abdominal pain and even with spontaneous retroperitoneal hemorrhage.[5]

Adrenal gland is the most common site of adrenal myelolipoma,[11] however, it can also occur in extra-adrenal sites such as presacral space, retroperitoneum, or even in the liver[9] and lungs.[11] Although unilateral cases are more common, bilateral involvement[8] and even presentation with renal cell carcinomas have been reported.[9] The size of the tumor is usually <5 cm, but giant myelolipomas, although rare, have also been reported.[11,9]

The most widely accepted theory regarding the etiology for adrenal myelolipoma is that it occurs due to metaplastic changes in the reticuloendothelial cells of blood capillaries in response to stimuli such as necrosis, infection, or stress.[1,10] Another etiology sited is prolonged stimulation with high levels of adrenocorticotrophic hormone or adrenal androgen.[10] Although adrenal myelolipoma is itself nonfunctional, its occurrence with Cushing syndrome, congenital adrenal hyperplasia, and adrenal ganglioneuroma has also been reported.[1,11]

Diagnostic modalities such as ultrasonography, CT, and magnetic resonance imaging are effective in detecting more than 90% of adrenal myelolipomas;[11] CT was most sensitive as it can identify fat in the adrenal gland.[11] On CT imaging, myelolipomas are seen as heterogeneous masses with low-density mature fat interspersed with more dense myeloid tissue.[11]

Being a benign condition, small adrenal myelolipomas which are <5 cm do not require surgery. These are usually monitored with imaging annually.[11] The indications for adrenalectomy are abdominal or flank pain, tumor diameter >8 cm, and uncertain diagnosis.[8] Minimal access surgeries such as laparoscopy and robotic surgery offer shorter operative time, less blood loss, less pain, and shorter hospital stay.[12] The reasons of conversion to open procedure are large tumor size, bleeding, and inadvertent injury to adjacent organs.[12] The outcome of surgery is good.
Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES