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BILATERAL GIANT ADRENAL MYELOLIPOMA: A RARE CASE OCCURRENCE

Pathology

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ABSTRACT

Myelolipoma is a combined lesion involving mature adipose tissue and bone marrow elements. Adrenal gland is the commonest site of myelolipoma. Usually these lesions are asymptomatic, unilateral and small in size. Occasionally myelolipomas become enormous and symptomatic depending on the location. Here we describe a case of bilateral giant adrenal myelolipoma presenting with abdominal pain. The case was managed surgically without any post-operative complications.

KEYWORDS

Adrenal myelolipoma, Giant myelolipoma, Bilateral adrenal tumors

INTRODUCTION:

Adrenal myelolipomas (AML) are benign neoplasms, composed of a variable proportion of hematopoietic tissue and mature adipose tissue.Generally,AML were accidentally discovered at autopsy with a low incidence varying from 0.08% to 0.4%.[1]Majority of adrenal myelolipomas are small (less than 4 cm), unilateraland asymptomatic (70%). Rarely they can present with hormonal disorders such as Cushing syndrome, congenital adrenal hyperplasia, or aldosterone-producing adenoma.[2] Adrenal though is the commonest site of myelolipomas, several extra-adrenal sites such as pelvis, retroperitoneum, and thorax were described in literature. Here we are presenting one case of bilateral giant adrenal myelolipoma presenting with dull aching abdominal pain.

Case report:

A 45 year old female presented to our institution with dragging abdominal pain localised to right flank for past 9 months. The patient was diabetic but otherwise healthy and without any hormonal symptoms. There was no associated history of hypertension.

Clinical examination of abdomen revealed a huge ill-defined firm, non-tender, ballotable mass in the right lumbar and hypocondrium region. The mass was moving with respiratory drive. On careful examination another small ballotable mass involving the left lumbar region identified. The mass was painless and had nodular surface.CT scan abdomen showed bilateral, heterogenous lesion with extensive intra-lesional fat attenuation involving both the adrenal fossae. The right side lesion measured 13x 8 x 6 cm, with central necrosis, and the left side lesion measured 8 x 7 x 4 cm. No evidence of local invasion or distant metastases was found.

Routine biochemical and haematological tests were normal. Serum cortisol and urinarymetanephrine levels were not raised and dexamethasone suppression test was normal.

The tumours were excised after proper anaesthetic check-up and gross examination of right adrenal tumour revealed an encapsulated 12 x 10 x6 cm globular fibro-fatty mass. Cut section showed yellow-greyish to black areas with central area of necrosis. Left adrenal tumour had nodular surface with greatest dimension of 7 cm, cut section showed similar areas.

Figure 1: (a) Gross photograph showing bilateral adrenal gland tumour and left adrenal tumour showing nodularity (b) Cut surface was brown-black with mixed yellow areas. Central portion of right adrenal tumour showed necrosis.

Microscopic examinationof both the adrenal tumour showe dhistological features of a well circumscribed tumour consisting of mature fat elements and haematopoietic elements mixed in different proportions. Many scattered megakaryocytes were identified in marrow elements. The main tumour is rimmed by normal adrenal cortical cells. Areas of necrosis identified in right adrenal tumour. No evidence of atypia or capsular breach identified. On the basis of above findings, diagnosis of bilateral adrenal myelolipoma was made.There was no post-operative complication and patient was discharged on 5th post-operative day.

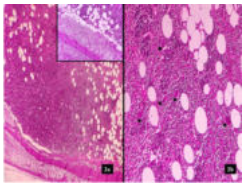

Figure 2: (a) Low magnification of tumour showing variable proportion of mature adipose tissue and haematopoietic tissue, the tumour is rimmed by adrenal cortical cells (100x, H & E); inset showing high power view of the same (400x, H & E) (b) High magnification showing many dispersed megakaryocytes (marked with*)in haematopoietic areas (400x, H & E)

DISCUSSION:

Myelolipoma is a rare, benign neoplasm of adrenal gland composed of mature adipocytes and hematopoietic tissue. In1905, Gierkefirst described it as a lesion consisting of mature fat tissue and bone marrow elements.[3]Later, in 1929 Oberling gave the name “formations myelolipomatoses”.[4] Previously AML used to be diagnosed at autopsy, but now a days, frequent use of CT scan and MRI have increased the frequency of detection.

The incidence of myelolipoma increases with age and mean age group is 50-70 years with equal sex distribution.[5] Some publications reported that myelolipoma is commoner on the right side than on the left. Majority of cases are confined to unilateral adrenal gland but other areas like pelvis, retroperitoneum, thorax has been documented in literature.

Myelolipomas are relatively slow growing tumors and more than 10cm in diameter are known as giant myelolipomas.[5] Bigtumors can become symptomatic. Nonspecific abdominal pain, nausea, vomitingare the usual symptoms but hematuria, or renovascular hypertension can occur because of compression of peritumoral tissueor intratumoral haemorrhage.[6]

The pathogenesis of myelolipoma development is not clear and several hypothesis have been proposed. The most accepted hypothesis stated that both altered mesenchymal stemcell functioning and hormonal stimuli act together in formation ofbilateral myelolipoma.[7]

Occasionally, AML can be associated with Cushing syndrome, congenital adrenal hyperplasia, and rarely with pheochromocytoma and adrenal insufficiency.[8]

The ultrasound ofmyelolipoma shows a hyperechoic mass intermixed with hypoechoic myeloid elements. However, diagnosis through imaging alone is difficult because of the tumor's non-uniform architecture and different proportion of fat content.[9] Associated calcification and haemorrhage also makes diagnosis difficult. CT scan and MRI are helpful in difficult cases and loss of signal of fat under fat suppression is a diagnostic clue in imaging. When larger in size and tumours with low fat content, involving the bilateral adrenal glands must be differentiated from pheochromocytoma.

Grossly, myelolipomas are well circumscribed and deep brown to yellow in colour depending on the proportion of fat and marrow elements. The surrounding adrenal gland is usually compressed. Microscopically, AML is composed of a variable proprtion of mature fat and hematopoietic elements. Differential diagnoses include lipoma, liposarcoma, angiomyolipoma, extramedullary hematopoiesis, and lipoadenoma. Extramedullary hematopoiesis (EMH) is ruled out with the absence of haematological and skeletal abnormality. In EMH, fat is usually present in the periphery of poorly circumscribed tumorwhereas in myelolipoma, fat is a component of the tumor itself.[8]

Management of AML is not standardised. However, small and asymptomatic myelolipomas (<5 cm) are usually managed conservatively with yearly CT follow up. Large (>5 cm) or symptomatic lesions are surgically resected to minimise the risk of spontaneous rupture leading to retroperitoneal bleed and malignant transformation. Giant AML generally need open approach rather than laparoscopic surgery.[10]

CONCLUSION:

Adrenal myelolipoma is an uncommon case and bilateral tumours are found in only 10% of cases. Here we are presenting a case of AML because of it's giant size and bilaterality. The post-operative period was uneventful. Extramedullary haematopoiesis was the closest differential.

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