Adrenal Myelolipoma: Report of Three Symptomatic Cases: A Case Report

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Abstract:

Myelolipoma is a rare benign tumour composed of mature adipose tissue and normal haemopoietic elements. Although it can occur in extra-adrenal sites, it typically occurs in the adrenal glands. Being small and asymptomatic in most cases, myelolipomas are incidentally discovered at autopsies and now-a-days, by imaging. The tumour becomes symptomatic when it attains a large size, when it ruptures, or when it is associated with endocrine abnormalities of the adrenals. We report three cases of adrenal myelolipoma with symptoms like abdominal pain and discomfort, obesity and hypertension. One of the cases also had a co-incidental Hydatid cyst of liver. Diagnosis of myelolipoma was suspected on imaging (computed tomography and ultrasound scan) and was confirmed by histopathology. The patients were followed up for one year after surgery. One patient was found well and symptomless. One sustained hypertension but was otherwise healthy. The remaining patient, who had hypertension and renal complications previously, developed fluctuating hypertension and chronic renal failure about six months after operation.

Key words: Adrenal myelolipoma, extra-adrenal myelolipoma.

Introduction:

Myelolipoma is a rare benign tumour composed of mature adipose tissue and haemopoietic elements. The tumour occurs typically in the adrenal glands¹,²,³,⁴,⁵,⁶,⁷, though it has been reported to occur in the renal hila²,³,⁴, liver⁵,⁶, spleen⁵,⁶, stomach⁵,⁶, lungs⁵,⁶,⁷, mediastinum⁵,⁶,⁷, retroperitoneum⁷, pre-sacral region⁷, mesentery¹² and even in the nasal cavity¹⁹. It is usually unilateral; bilaterality is rare²,¹⁰. The tumour is seen to occur in obese adults and older⁸. Males and females are almost equally affected¹³.¹¹. Vast majority are asymptomatic and are incidentally discovered on imaging or at autopsy¹,²,³,⁴,⁹,¹¹. The tumour becomes symptomatic when it attains a large size, when it ruptures, or when it is associated with endocrine abnormalities of the adrenals⁶. Symptomatic patients may present with abdominal pain¹,⁷,¹¹,¹², heaviness¹⁵, haematuria³, Cushing’s syndrome⁸ or hypertension⁸,¹²,¹⁸,²⁵. Myelolipomas are now known to have a clonal origin and to show association with some cytogenetic abnormalities⁸,¹⁴,¹⁵.

Case reports:

We report three cases of adrenal myelolipoma, all of which were symptomatic.

Case 1:

A 50-year-old male patient presented with pain in the right hypochondrium and heaviness in the abdomen. Ultrasonogram and computed tomography scan of the abdomen revealed a right-sided adrenal mass and a large cyst in the liver with calcified wall. Both ultrasonogram and CT scan reports suggested lipoma of the adrenal gland and Hydatid cyst of the liver. Both the lesions were surgically removed. Grossly the adrenal mass was 10x7x3 cm in size weighing 105 gm. The cut surfaces were fatty with grey brown and haemorrhagic areas. Histopathology revealed a benign tumour composed of mature adipose tissue admixed with bone marrow elements containing cells of all three lineages. The hepatic cyst was grossly and microscopically confirmed as a Hydatid cyst. The patient was followed up for one year after surgery. He was found well with no complaints.
Case 2:

A 37-year old male patient presented with hypertension and numbness in the extremities. He also gave history of frequent urinary tract infection since childhood. Abdominal ultrasonography revealed a mass in the right adrenal gland. Report of CT scan-guided FNAC (fine needle aspiration cytology) of the mass suggested angiomyolipoma. The patient’s blood biochemistry revealed increased serum creatinine level (2mg/dl). Serum cortisol, catecholamines and urinary vanillyl mandelic acid (VMA) were normal. The surgically removed adrenal mass was a yellowish nodule measuring 5.5x4x3.8 cm and weighing 25 gm. The cut surfaces were fatty with red brown areas. Microscopically mature adipose tissue admixed with haemopoietic cells rimmed by a thin layer of adrenocortical cells was seen typical of myelolipoma. The patient was well till six months after operation. Then he gradually developed fluctuating hypertension, chronic renal failure and anaemia. His serum uric acid level was also raised. He is now being given symptomatic treatment. Renal biopsy has not been done.

Case 3:

Our third case was also a male person aged 65 years who presented with abdominal pain. The patient was obese and hypertensive. Clinically a mass was palpated in the right hypochondrium. Ultrasonogram of the abdomen revealed a right adrenal mass compressing the right kidney. CT scan of the upper abdomen suggested a myelolipoma of the right adrenal gland. The patient’s biochemical reports of blood including blood glucose, urea, creatinine were normal. Serum hormone levels were not done. The 24-hour urinary vanillyl mandelic acid (VMA) was slightly raised. The tumour was removed surgically. Grossly the mass was capsulated measuring 9x7.5x5 cm and weighing 100 gm. Sectioning revealed fatty cut surfaces with small foci of haemorrhage. Microscopic examination revealed a myelolipoma. The patient was followed up for six months after surgery. He is still hyper-

Discussion:

The rare benign tumour myelolipoma was first described by Gierke in 1905, and later nomenclatured by Oberling
tissue and bone marrow elements. The fatty tissue is mature and the bone marrow component is normal with active trilineage haematopoiesis. Patients do not have any associated haematopoietic disease. The pathogenesis of the lesion has been hypothesized as metaplasia of the mesenchymal cells of the affected organ and migration of haemopoietic stem cells. Bishop E, et al. (2006) showed a non-random X chromosome inactivation in both myeloid and fatty tissue elements of adrenal myelolipoma, suggesting a clonal origin of the tumour. Earlier, Chang KC, et al. (2002) reported a case that showed balanced translocation between chromosomes 3 and 21 ([3;21](q25:p11)) in conventional cytogenetic study.

Being small and asymptomatic in most cases, myelolipomas have been incidentally discovered at autopsies (0.08%-0.4%)\(^{1,11,18}\). Incidental diagnosis or suspicion of a myelolipoma is being made now-a-days with improved image studies like computed tomography scan (CT scan), magnetic resonance imaging (MRI) and also ultrasound scan\(^{11,13}\). However, it is difficult to distinguish the lesion radiologically from other fat-containing tumours like angiomyolipoma, lipoma and well differentiated liposarcoma\(^7,18\). Symptomatic adrenal myelolipomas usually present with abdominal pain and discomfort\(^{17,11,12}\), back or flank pain\(^3\), haematuria\(^3\), hypertension and endocrine abnormalities\(^{8,18,25}\). These are attributed to mass effects\(^{13,5,7}\), rupture of the tumour\(^8,13\) and associated other adrenal pathologies like adrenal cortical hyperplasia, cortical adenoma or adrenocortical carcinoma\(^8,12,18,20\). Spontaneous rupture with retroperitoneal haemorrhage is a well-recognized complication of adrenal myelolipoma\(^1,12\). Patients may also have multiple endocrine neoplasia (MEN) syndrome\(^12\).

All three of our patients were adult males. They were symptomatic having right-sided adrenal mass. They all had abdominal pain and heaviness because of large size of the tumours. Two of them had hypertension and one was obese. Case 2 had numbness in the extremities which was probably due to pressure effect of the tumour on the spinal nerve roots. This patient also had urinary symptoms which he claimed to have from the childhood. He also had raised serum uric acid level and later developed chronic renal failure. But these events are assumed to be unrelated to adrenal myelolipoma, as no such link has been reported so far. Rather, studies have shown a pathogenic link between hyperuricemia, hypertension and progressive renal failure\(^{21,22,23}\). Our case 1 had simultaneous hydatid cyst in the liver. Hydatid cyst is infectious in origin and unrelated to myelolipoma.

Although myelolipomas can be diagnosed by image studies and fine needle aspiration cytology (FNAC), the diagnosis is confirmed by histopathology. FNAC usually reveals fragments of mature adipose tissue and trilineage haemopoietic cells in various stages of maturation in a bloody background\(^6,24\). The key histopathological picture of myelolipoma is unique consisting of mature lipid and myeloid elements in variable proportions. We also observed a thin rim of normal adrenal tissue adjacent to the tumour in our cases. Areas of haemorrhage, necrosis and calcification are commonly found in larger lesions\(^7,9,11\).

Prognosis of myelolipoma is generally excellent\(^11\). Patients with small tumours are symptomless and often remain undiagnosed. Patients with large ones are usually well after removal of the tumour. However, symptoms like obesity, hypertension and other chronic and vague abnormalities have been reported to persist in some patients\(^25\).

**Conclusion:**

Report of our three cases points to the possible occurrence of myelolipoma with big adrenal masses and the very benignity of the tumour. Being a tumour with good prognosis, adrenal myelolipoma must be accurately diagnosed to avoid unnecessary and hazardous therapy. As an uncommon tumour, it can be easily confused with other fat-containing and aggressive retroperitoneal neoplasms. Extra-adrenal myelolipomas should be differentiated from foci of extramedullary haemopoiesis. Any adrenal mass must be categorized by histopathology.
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References:


