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Clinicopathological spectrum of adrenal myelolipomas.

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Introduction

Adrenal myelolipomas are common benign adrenal tumour, being second most common tumour after adrenal adenomas. They make up to 6–16% of all adrenal tumours. It is composed of elements of adipose tissue and extramedullary hematopoiesis. The most recent World Health Organization update on endocrine tumours places myelolipomas in the category of mesenchymal and stromal tumours of the adrenal cortex. [1] Their average age at presentation is 50 years old and are typically discovered in late adulthood. Men and women appear to be equally affected. Although certain bilateral tumours have been reported in the literature, these lesions are mostly unilateral and asymptomatic. (2) When first found, myelolipomas frequently have a diameter of less than 4 cm, but they have the potential to grow to enormous sizes. (3) Despite occasionally coexisting with primary aldosteronism and congenital adrenal hyper plasia, they are hormonally inactive. Adrenal myelolipoma is clinically relevant despite its benign behaviour because it may make it difficult to differentiate between adrenal tumours.

Patients and methods

All patients presenting to SGPGI from January 2014 to January 2020 were reviewed. Thirty-seven patients with histopathological confirmation of adrenal myolipomas medical records could be retrieved and were included in the present study. The histopathological confirmation was done at our department of pathology.29 cases presented with chief complain of intermittent dull aching pain in abdomen and rest were incidentally detected. They underwent radiology as routine investigation. The clinical factors (age, sex, symptoms, and duration of symptoms, histological features), therapeutic factors

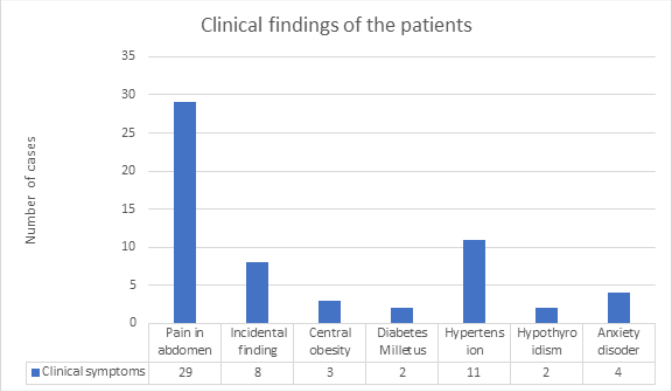
(treatment modality) and his to logical features were recorded and evaluated appropriately.

Results

Their age mean was 45.9 (22 to 74 years) with M: F ratio of 14:23. They presented with complain of intermittent dull aching pain in abdomen in 29 cases, wherever 8 were incidentally detected. Serum level of cortisol (overnight) <27.6 mcg/dl, urinary metamachine level < 400 mcg /24 hour and normetanephrine level were in range of 75-375 mcg/24 hour for all patient. Aldosterone level was done only for three patient which ranges from 0.2-0.54 mcg/24 hours. Plasma renin ranges from 0.2-3.4 ng/ml/hour for all patients.

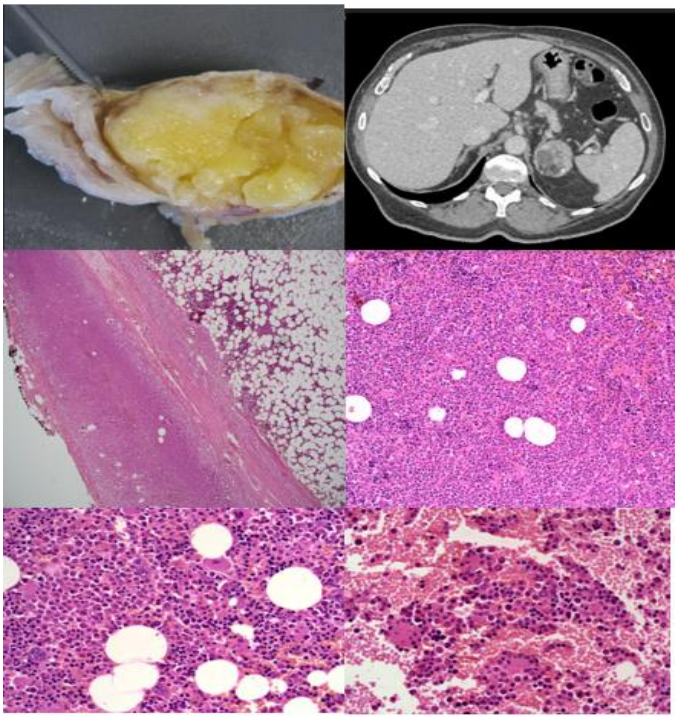
14 cases had comorbities. 3 cases had central obesity; 2 cases had history of diabetes whereas 2 cases had history of hypothyroidism. 10 cases presented with history of hypertension. One diagnosed case of chronic pyelonephritis with nephrolithiasis showed accelerated hypertension. Only four had a history of anxiety disorder.33 cases had no history of fracture, renal calculi, syncopal attack, or neuropsychiatric disorder.

Graph 1:



All 37 cases under went image study by CECT and 3 cases underwent USG.CECT showed heterogenous mass with predominant fatty component in most cases with 28 on right side, 8 on left side and 1 was bilateral. Laparoscopic adrenalectomy in 13 and open peritoneal adrenalectomy in 24 patients were performed. The differential diagnosis for the mass was hemangioma or myelolipoma. All tumors in our series were greater than 3 cm. The largest was of dimension 24x17x9 cm. On gross examination tumor were solid brown with Haemorrhagic and fatty areas. On microscopy all had lobules of adipocytes with foci of sheets of hematopoietic cells, including cells of myeloid, erythroid and mega karyocyte lineages along with unremarkable adrenal tissue. 9 cases showed areas of hemorrhages.

Figure 1:



[A] Gross specimen: Encapsulated adrenal tumour mass. Cut section showed brownish whitish and fatty areas, replacing entire adrenal parenchyma with rim of normal adrenal gland.

[B] CT Imaging: Lobulated well defined heterogeneously enhancing lesion in left suprarenal region. Multiple foci of fat density are seen within the lesion.

[C] Microscopy (low power): Sections from the adrenal shows a peripheral portion of normal adrenal tissue.

Central portion shows islands of hematopoietic tissue and mature fat.

[D, E] Microscopy (higher power): Foci of Hema to poietic tissue shows erythroid, myeloid and mega karyocytes.

[F] microscopy (higher power): Islands of hematopoietic tissue with areas of hemorrhage

Discussion

Adrenal myelolipomas are usually seen in one out of 500-1250 autopsy cases [4]. However, due to the huge proportion of asymptomatic patients and the benign nature of the tumour, it is very impossible to determine the precise clinical prevalence of the condition. AML is the second most prevalent main adrenal incidentaloma, accounting for 6-16% of adrenal incidentalomas after the adrenocortical adenoma, which accounts for 60-70% of adrenal incidentalomas [5–7].

Extra-adrenal myelolipoma refers to a rare occurrence of myelolipomas outside of the adrenal gland. Myelolipomas have been reported in a variety of places, including the mediastinum, spleen, kidney, bones, thorax, nasal cavity, ectopic adrenal cortex, and extra Dural regions. Biochemically, myelolipomas are mostly non-functional. The majority are asymptomatic, but a few may rarely present with stomach pain because of their size or from spontaneous bleeding. Although there are case reports of their relationship with excessive adrenal hormone synthesis, these tumours are typically hormonally inactive. Dehydroepi and roster one sulfate (DHEAS) over production, congenital adrenal hyperplasia brought on by a 21-hydroxylase deficiency, congenital adrenal 17-hydroxylase deficiency, Cushing disease, Conn syndrome, adrenal insufficiency, and pheochromocytoma have all been linked to myelolipomas. (8,9)

The majority of myelolipomas displayed non-random X-chromosome inactivation, according to Bishop et al., indicating that these cancers have a clonal origin [10]. The genesis of adrenal myelolipoma, according to another theory, is due to the interaction of two different kinds of progenitor cells. First, mesenchymal stem cells stored in the vascular wall of the stromal fat of the adrenal cortex give rise to adipose tissue in response to certain stimuli. Adipocytes undergo inflammatory changes as they develop and mature, which trigger the adrenal cortex to produce agents that draw circulating Hema to poietic progenitors to settle and undergo differentiation. [11] In most instances, surgical excision is curative.

Myelolipomas are well-defined heterogeneous masses that contain dense myeloid tissue interspersed with low-density mature fat (less than -30 Hounsfield Units [HU]). (Figure 1). (12)) Myelolipomas are shown as a well-defined tumour on ultrasonography with different degrees of hyperechoic (fatty tissue) and hypoechoic areas (myeloid tissue). (13) Since there are no precise imaging criteria to diagnose adrenal myelolipoma, accurate preoperative diagnosis may be challenging. In half of our patients, malignancy could not be completely ruled out. Cytological or histological analysis is required for the definitive diagnosis of an adrenal myelolipoma. [14]. In most instances, surgical excision is curative. Although other less common adrenal tumours containing fat, such as teratoma, lipoma, and liposarcoma should be taken into consideration, a fatty adrenal mass is essentially diagnostic of myelolipoma. Angiomyolipoma should be part of the differential diagnosis even though it is significantly more frequent in the kidneys. Although adrenal adenomas might have attenuation values less than 18 HU, they are denser and more homogeneous

than myelolipomas despite their appearance as low-density lesions. Our research showed that the size of the adrenal myelolipoma is connected with symptoms. Larger adrenal myelolipoma should present symptoms more clearly. Surgery does, at least in our circumstances, ease symptoms. Cytological or histological analysis is required for the definitive diagnosis of an adrenal myelolipoma. Surgery is typically not required if the diagnosis of an adrenal myelolipoma is clear-cut and there are no established symptoms or hormonal activity. However, elective surgery can stop the development of life-threatening symptoms and more severe symptom presentation.

Conclusion

The rate of tumour discovery is rising as awareness grows. Radiology and clinicopathological correlation are trustworthy diagnostic methods. In the group of patients whose tumour size was higher than 6 cm, elective surgery is also advised for the prevention of spontaneous rupture. Furthermore, less invasive laparoscopic surgery may motivate patients to undergo surgery in addition to providing an alternate therapeutic option. Despite being a benign tumour, surgery is nevertheless used in the management of adrenal myelolipoma. For the purpose of creating an accurate treatment plan, more research is required.

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