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# Clinicopathological and radiological spectrum of myelolipoma in a tertiary care center

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**Abstract:**

Myelolipomas are uncommon benign nonfunctional tumors most commonly involving the adrenal gland. However extra adrenal presentations are also observed. They are usually asymptomatic, unilateral and endocrinologically nonfunctional neoplasms. These tumors are usually incidentally detected during radiological investigations on suspicion of other diagnosis. Large tumors are usually misdiagnosed as liposarcoma. We report our experience based on a single institute surgically treated cases of myelolipoma over a period of 6 years and literature review. Our results were in concordance with the literature review.

**Key Words:** Adrenal Myelolipoma, Incidentaloma, Nonfunctional tumors,

## Introduction:

Adrenal myelolipoma is a rare, nonfunctioning, and benign tumor. It was first described by Gierke in 1905 and named by Oberling in 1929 [(1)]. In the past, these lesions were detected at autopsy or in conditions where massive growth of the tumor produced symptoms. In recent days due to the advent of imaging techniques, these tumors are detected incidentally; it is designated as "Incidentalomas". These tumors are usually incidentally detected during radiological investigations on suspicion of other diagnosis.. Large tumors are usually misdiagnosed as liposarcoma. Large tumors have areas of hemorrhagic, necrosis,cyst formation , calcification and ossification. They are usually asymptomatic, unilateral and endocrinologically non functional neoplasms. Though they are usually unilateral, some bilateral cases are also reported in the literature .They may also develop in extra adrenal sites like the retroperitoneum, thorax, and pelvis. Typical Myelolipoma are nonencapsulated, well circumscribed tumors. They are seen in association with adrenal cortical adenoma or cortical hyperplasia. Microscopy shows mature adipocytes and extra medullary trilineage hematopoietic cells with full maturation, often there is a marked increase in megakaryocytes. While myelolipomas are mostly incidentally detected , they may become symptomatic causing flank pain and abdominal discomfort by causing pressure of surrounding structures and may even present with necrosis, rupture, hemorrhage, or hemorrhagic shock [(2)]. In this study we reviewed the clinicopathological spectrum of myelolipoma presented to our department from the year 2018 to 2023.

## Materials and methods:

A retrospective study of all the cases of Adrenal reported at the department of Pathology in SRIHER from 2018 to 2023 were reviewed. Out of which 12 were myelolipomas, included in the study. Clinical and radiological details were recorded from the archives. The variables of the spectrum include age, sex, presentation of symptoms, medical history, side and size of the tumor , clinical and radiological provisional diagnosis. All of them underwent surgical excision and a definitive diagnosis of myelolipoma was made on histopathological examination.

## Results:

| Sno | Age | Sex | Presentations | Medical History | Clinical Diagnosis | Side | Size (cm) | Image Study | Radiological diagnosis |
|---|---|---|---|---|---|---|---|---|---|
| 1 | 31 | F | Right loin pain-1 month | Nil | Right adrenal myelolipoma | R | 5x4.5x3 | CT | Right adrenal myelolipoma |
| 2 | 37 | M | Left chest pain | HTN- 5 years | Suprarenal mass | L | 4.5x4x3 | USG | Left adrenal myelolipoma |
| 3 | 45 | M | Lower Back ache | Nil | Adrenal adenoma | R | 7.7x5.6x6.2 | CT | Adrenal adenoma |
| 4 | 46 | M | Incidental - Master health check up | Nil | Right adrenal myelolipoma | R | 10x9.6x7.9 | CT | Right adrenal myelolipoma |
| 5 | 46 | M | Right loin pain- 1 week | Nil | Right adrenal tumor | R | 7.6x7.5x6.5 | CT | Myelolipoma |
| 6 | 48 | F | Right loin pain-3 days | T2DM | Adrenal tumor | BL | 13.8x5x3.5 | CT | Myelolipoma |
| 7 | 50 | M | Abdominal pain-1 week with cholelithiasis | Hereditary spherocyto-sis | Left adrenal lesion | L | 5.2x4x3.5 | USG | Left adrenal lesion |
| 8 | 55 | M | Incidental - cholilithiasis | HTN | Right adrenal myelolipoma | R | 12x7x5 | CT | Right adrenal myelolipoma |
| 9 | 56 | M | Incidental - Master health check up | T2DM & HTN | Right adrenal tumor | R | 7x4.5x4 | USG | Right myelolipoma |
| 10 | 57 | F | Abdominal distension-3 months | DM & HTN | Right adrenal mass | R | 20x17x10 | CT | Well differentiated liposarcoma |

| | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|
| 11 | 57 | F | Abdominal distension-2 weeks | T2DM- 10 yrs | Retroperitoneal tumor | R | 16x14x12 | CT | Right myelolipoma |
| 12 | 61 | F | Right loin pain- 1 week | T2DM, HTN | Retroperitoneal tumor | R | 20.5x18.7x13.8 | CT | well differentiated liposarcoma |

F- Female HTN- Hypertension R- Right

M- Male T2DM- Type 2 diabetes mellitus L- Left

CT- Computed tomography scan BL- Bilateral

USG- Ultrasound sonography

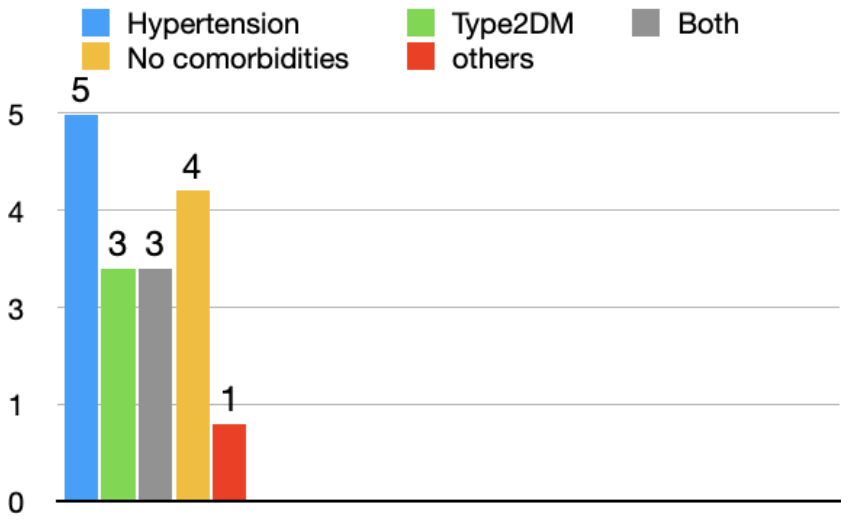


**Figure 1: The most common symptom in this study was hypertension**

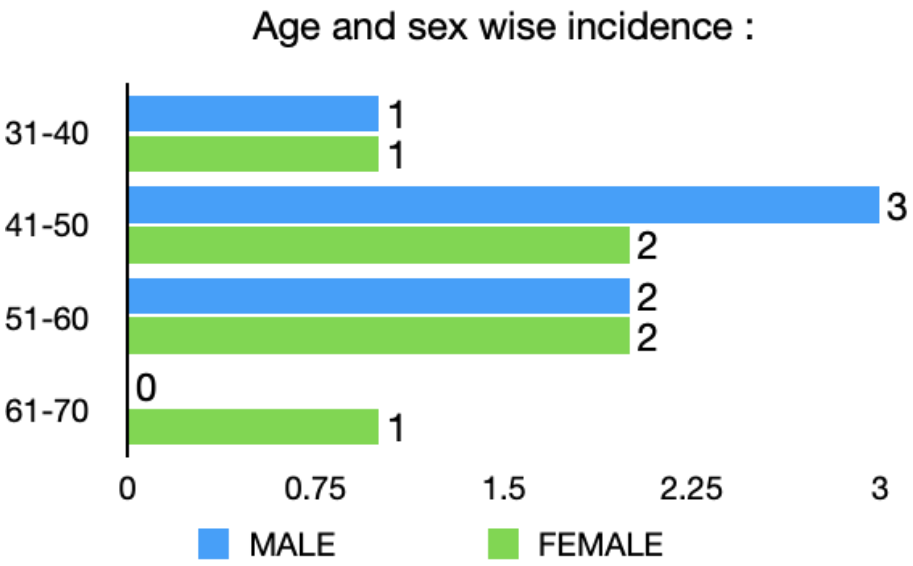


**Figure 2: The most common incidence was observed in age group 41-50 with male predilection**

All cases of adrenal tumors were analyzed out of which 12 cases were myelolipoma with the incidence of 17.9 %. The highest incidence of myelolipoma was reported in age groups ranging 31-65 with male predominance. Incidence of two cases in younger age groups has also been observed. The analysis showed the incidence of 9 cases involving the right adrenal gland and 2 cases involving the left adrenal gland. Adrenal myelolipomas are usually unilateral but bilateral involvement was seen in one case which is extremely rare. Clinical symptoms like abdominal / loin pain and abdominal distention was observed in 10 cases. While 2 cases were incidentalomas on master health check up. Eight cases were associated with comorbidities like diabetes mellitus, hypertension and hereditary spherocytosis . None of the cases were associated with any endocrinopathies. All patients received image study by CT and two cases by USG. CT showed well circumscribed , mixed density lesion. USG findings were predominantly well defined hyperechoic mass lesion with areas of punctate calcification within the lesion. Among the 12 cases, two cases were radiologically diagnosed as retroperitoneal mass while the others were radiologically reported as adrenal mass. Largest tumor was measuring 20.5 cm and weighing 1.5kgs presented as abdominal pain with the radiological diagnosis of well differentiated liposarcoma, considering the large bulk of the tumor. Histopathological and radiological

correlation was observed in 69.2% of the tumors, while others were radiologically diagnosed as various other adrenal lesions like adenomas and liposarcomas. The gross external surface of the tumor was glistening with variegated appearance. Cut surface of the lesion appeared yellowish to dark brown in color with soft consistency. None of the large sized tumors in our study were associated with rupture or any other complications. The yearly incidence of the cases has increased from one case reported in 2018 to 5 cases reported in 2022 and 6 cases until May 2023.

## Discussion:

It is vital to understand the tumorigenesis of myelolipoma. Several studies have proposed different theories in context to the tumorigenesis of myelolipoma . The most acceptable hypothesis states that the fat components are derived by the mesenchymal stem cells of stromal fat of adrenal cortex under certain stimuli. Mature adipocytes begin to accumulate and become inflammatory stimulating neighboring adrenal cortex tissue to release possibly G-CSF to recruit circulating haematopoietic progenitors [(3)]. As the tumor proliferates the hematopoietic stem cells obtain energy by burning the surrounding fat until they are fully differentiated and division stops. The central part undergoes necrosis and calcification while the cells in the periphery continue to produce newly derived adipocytes and hematopoietic progenitors. The necrosis or calcification of the tumour the inflammation persists and the tumour generates a self-growing signaling loop, entailing a continuous growth even without further stimuli [(3)]. Sz-Wen Hsu et al performed a case a case study, the analysis on which showed patient mean age was 47.7 ± 2.94 years with the occurrence to be slightly more often in males than in females, which correlated with our case study with incidence most commonly seen in the fourth decade with mean age of 48.36 and higher incidence in males than in females [(1)].The largest myelolipoma ever reported was by Boudreaux et al. which was 34 cm and weighed 5900 grams [(4)]. The largest myelolipoma observed in our study was 20.5cm and weighed 1500 grams. Duarte Regalado CS et al reported a case in which the patient with history of laparoscopic cholecystectomy for cholelithiasis presented left flank pain, and CT image suggesting left adrenal myelolipoma [(5)]. Small lesions measuring < 5 cm and those who are asymptomatic are usually monitored via imaging over a period of 1–2 years [(6)]. On CT due to their lipid content, myelolipomas present with attenuation less than 0 HU, sometimes less than −50 HU on pre-contrast CT images. Due to the appearance of

hematopoietic tissue , the attenuation of myelolipomas is mildly higher than that of ambient fat space [(7)]. contrasted enhancement of the hematopoietic tissue is noted on contrast-enhanced CT images. MRI, shows high signal on both T1- and T2-weighted images and there is a loss of signal on fat-saturated MRI. Several studies has revealed the strong association between myelolipoma and conditions such as Cushing's disease, pheochromocytoma, obesity, and diabetes. The prognosis of adrenal myelolipoma is remarkable with recurrence-free survival rates of up to 12 years [(8)]. Sakamoto A et all in his study reported that extra-adrenal myelolipomas have been reported to be accompanied by diabetes mellitus, cancer, and steroid use in 21.6%, 18.9%, and 16.2% of patients, respectively [(9)]. Definitive diagnosis of adrenal myelolipoma depends on cytological or histological evaluation [(10)].

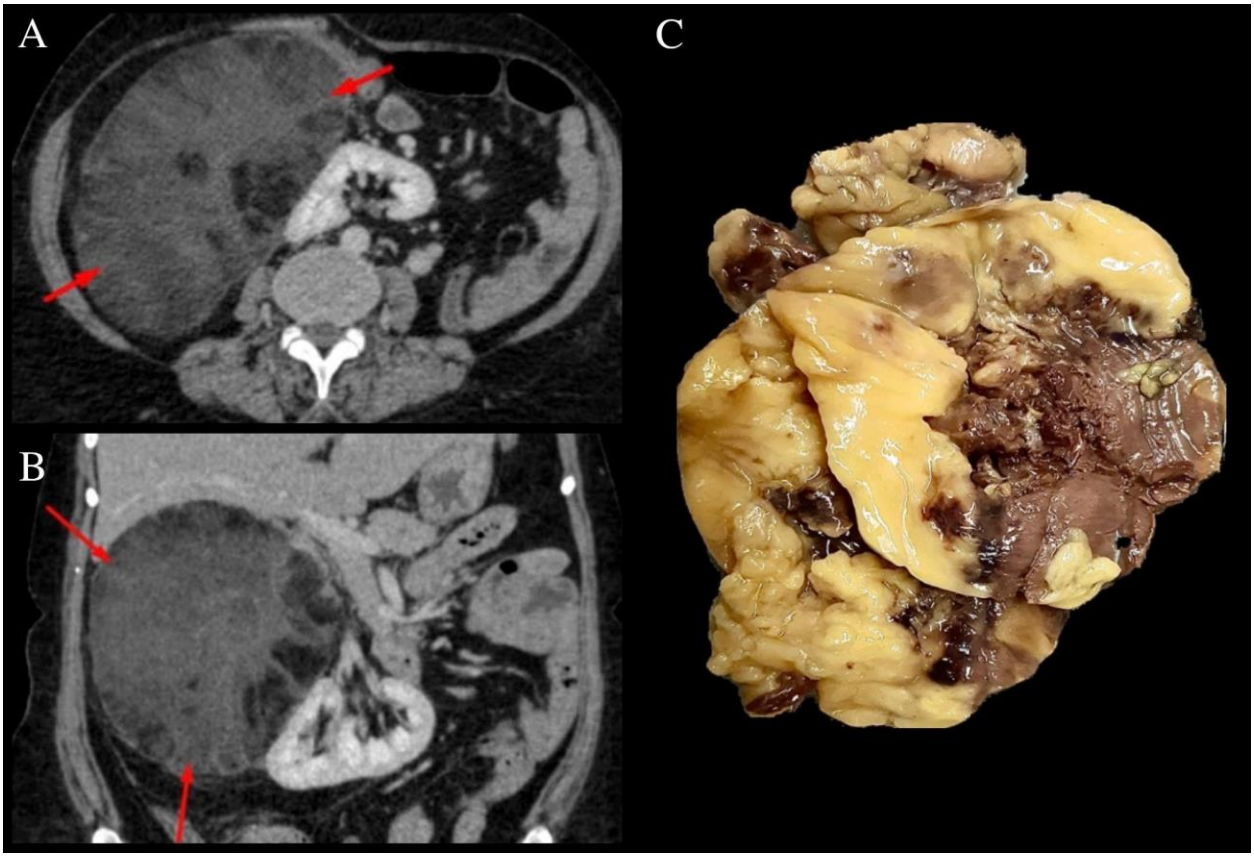

Figure 3: (A+B) CT image showing well circumscribed , mixed density lesion.
(C) Gross appearance of the tumor showing a glistening, bright yellow and variegated surface.

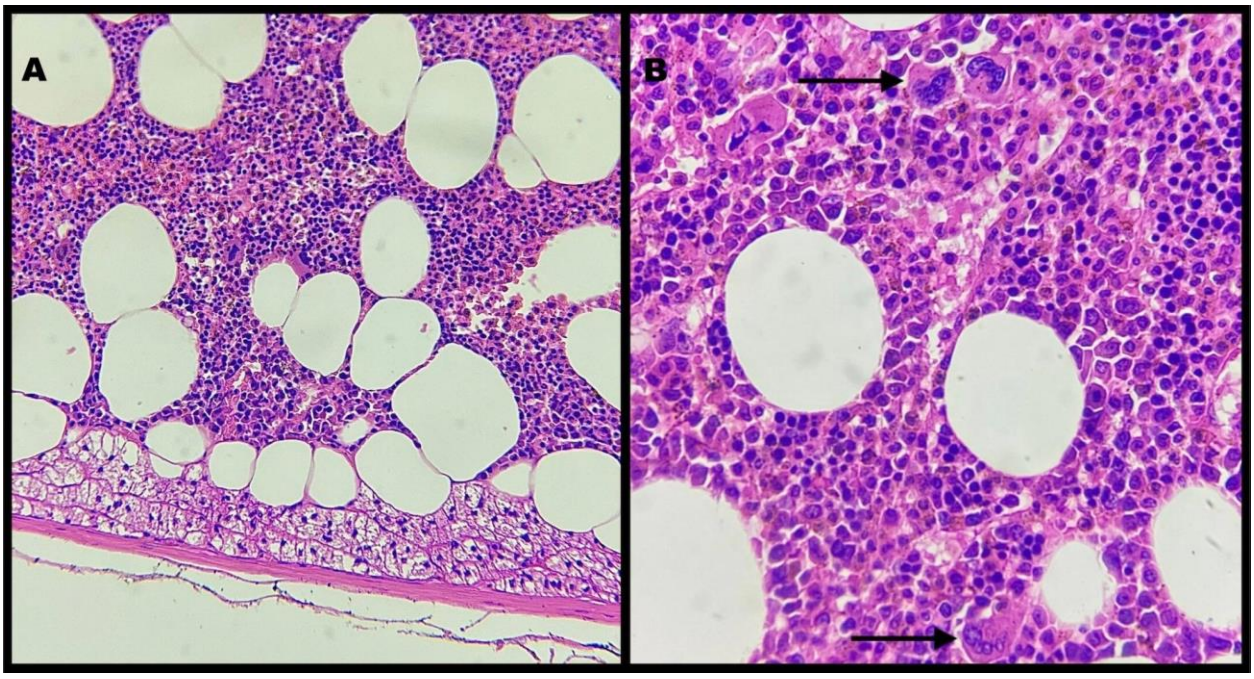


Figure 4: (A) Mixture of mature adipocytes with extramedullary trilineage hematopoietic cells and megakaryocytes with a rim of compressed adrenal cortex at the periphery.
(B)Increase in megakaryocytes (black arrow) noted intermingled between extramedullary trilineage hematopoietic cells and mature adipocytes.

## Conclusion:

Myelolipomas are non-functional tumors, hence small tumors are managed by medical and imaging control, ideally with CT to objectify the stability or changes in size. Large tumors compress the neighboring organs or manifest complications like rupture or hemorrhage, hence surgical removal is advocated. This is to avoid the risk of abdominal pain or life-threatening rupture and hemorrhage. Studies suggest that the laparoscopic approach is superior to the open method as it can lead to lower morbidity especially on surgical site infection and lung complications, faster recovery and hospital discharge. Open method is preferred for masses larger than 10 cm or with adhesions and infiltration of the surrounding structures .The increase in incidence of adrenal Myelolipomas over the years is attributed to increase in the advancement of

imaging modalities and with accurate diagnosis by histopathological findings. Hence the patient is medically or surgically managed based on the case presentation.

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