SAS Journal of Surgery
Abbreviated Key Title: SAS J Surg
ISSN 2454-5104
Journal homepage: https://www.saspublishers.com

OPEN ACCESS

Pathology

Adrenal Myelolipoma: Case Report

Dounia Benaich[1,2*], Ihssan Hadj Hsain[2,3], A. Jahid[1,2], F. Zouaidia[1,2], K. Znati[1,2], Z. Bernoussi[1,2]

[1]Department of Pathology, Ibn Sina University Hospital, Rabat, Morocco
[2]Faculty of Medicine and Pharmacy of Rabat, Mohamed V University, 10100, Rabat, Morocco
[3]Department of Radiology, Ibn Sina University Hospital, Rabat, Morocco

DOI: 10.36347/sasjs.2024.v10i02.003 | **Received:** 17.12.2023 | **Accepted:** 23.01.2024 | **Published:** 10.02.2024

***Corresponding author:** Dounia Benaich
Department of Pathology, Ibn Sina University Hospital, Rabat, Morocco

Abstract **Original Research Article**

Introduction*:** Myelolipomas were first described in 1905 as an adrenal tumor composed of mature fat mixed with myeloid and erythroid cells. Adrenal myelolipomas are the second most common benign type of tumor in the adrenals, following after adrenocortical adenomas. It is mostly discovered incidentally on imaging of abdomen done for non-adrenal related reasons or at autopsy. Symptomatic tumors, growing tumors or tumors larger than 10 cm should be excised surgically. We report the case of 1 male patient presenting with flank pain and upper pole renal masse, treated surgically with adrenalectomy. Myelolipoma was confirmed on histology. ***Case presentation*:** a 46-years-old male, with a history of blood hypertension, presented with persistent abdominal discomfort and occasional pain in the right flank region. Physical examination revealed a non-tender palpable mass in the right upper quadrant of the abdomen. The abdominal CT scan revealed a large well-defined retroperitoneal tumor, which compressed and displaced the right kidney downward, the mass was heterogeneous with areas of soft-tissue attenuation. The patient underwent open left adrenalectomy. The pathological study confirmed the diagnosis of adrenal myelolipoma. The differential diagnosis of fat-containing retroperitoneal masses would include retroperitoneal lipoma or liposarcoma and renal angiomyolipoma. ***Discussion: Adrenal myelolipomas are the second most common adrenal incidentalomas comprising 6 to 16% of adrenal incidentalomas. The size of adrenal myelolipomas is variable and can range from a few millimeters to greater than 10 cm. These are called giant adrenal myelolipomas. The largest adrenal myelolipoma reported to date weighed 6 kg. Extra-adrenal sites for myelolipomas include the retroperitoneum, thorax, and pelvis. Usually unilateral however they can also involve both adrenals. The most common symptoms observed are abdominal discomfort/pain, hypochondrial pain, and flank pain. Ultrasonography (USG), CT, and MRI are all highly beneficial in the diagnosis of adrenal myelolipoma, with CT scan being the most sensitive for the identification of fat within the lesions. Histological assessment can be made on the resected specimen. Thorough gross and microscopic evaluation is needed for confirmatory diagnosis and to exclude malignancy. Surgery is indicated in symptomatic tumors, rapidly growing lesions, or tumors more than 6 cm in size. The prognosis of adrenal myelolipoma is remarkable with recurrence-free survival rates of up to 12 years. ***Conclusion*:** Adrenal myelolipoma is commonly benign, asymptomatic, and hormonal inactivity. A surgical strategy is suggested for high-complication-risk patients. Radiological findings help in early suspicion of this asymptomatic tumor. The laparoscopic approach is safe and effective with an obvious advantage over open procedures. Although histopathological examination remains the gold standard for definitive diagnosis of adrenal myelolipoma.

Keywords: Arenal myelolipoma, adrenal incidentaloma, case report, adrenalectomy, laparoscopic surgery.

INTRODUCTION

Myelolipomas were first described in 1905 as an adrenal tumor composed of mature fat mixed with myeloid and erythroid cells [6]. Myelolipomas are not extra medullary hematopoiesis (which does not contain fat) and are not associated with any specific hematologic disorder [7]. Adrenal myelolipomas are the second most common benign type of tumor in the adrenals, following after adrenocortical adenomas [6]. It is mostly discovered incidentally on imaging of abdomen done for non-adrenal related reasons or at autopsy. Usually asymptomatic, but has been reported to present with symptoms such as flank pain resulting from tumor bulk, necrosis or spontaneous retroperitoneal haemorrhage. Symptomatic tumors, growing tumors or tumors larger than 10 cm should be excised surgically. We report the case of 1 male patient presenting with flank pain and upper pole renal masse, treated surgically with adrenalectomy. Myelolipoma was confirmed on histology [1].

CASE REPORT

We present the case of a 46-years-old male who presented with persistent abdominal discomfort and occasional pain in the right flank region. The patient has underlying hypertension with one anti-hypertensive drug. Physical examination revealed a non-tender palpable mass in the right upper quadrant of the abdomen. The abdominal CT scan revealed a large well-defined retroperitoneal tumor, which compressed and displaced the right kidney downward, the mass was heterogeneous with areas of soft-tissue attenuation [Fig. 1]. The approximate dimensions of the lesion were 12,7 x 11 x12,5 cm. In light of these findings and after a thorough preoperative workup, the patient was operated for left adrenalectomy and the specimen was sent for histopathological examination.

Grossly, the tumor was firm with a lobulated surface measuring 8 mm × 40 mm and weighing 140 grammes was partly covered by fatty tissue with an intact fibrous capsule [Figure 2A]. Serial sections showed a well-circumscribed tumor with heterogeneous yellowish to a greyish cut surface. Few foci of small haemorrhagic spots were seen. No necrosis identified. There is a remnant of adrenal gland attached to the lesion measuring 20 mm × 7 mm [Figure 2B]

Histologically, it is a well-circumscribed tumor composed of lobules of mature univacuolated adipocytes separated by fibrous septae and rimmed by a thin capsule. There were intervening blood vessels and bone marrow elements containing trilineage hematopoietic cells with an increase in the number of megakaryocytes. Foci of haemorrhages were seen within the adipose component [Figure 3A]. No lipoblast or atypical stromal cell seen. No necrosis or calcification identified. There was an attenuated adrenal cortical tissue seen at the periphery. No evidence of malignancy seen. [Figure 3B]

The final histopathology was reported as myelolipoma of the adrenal gland.

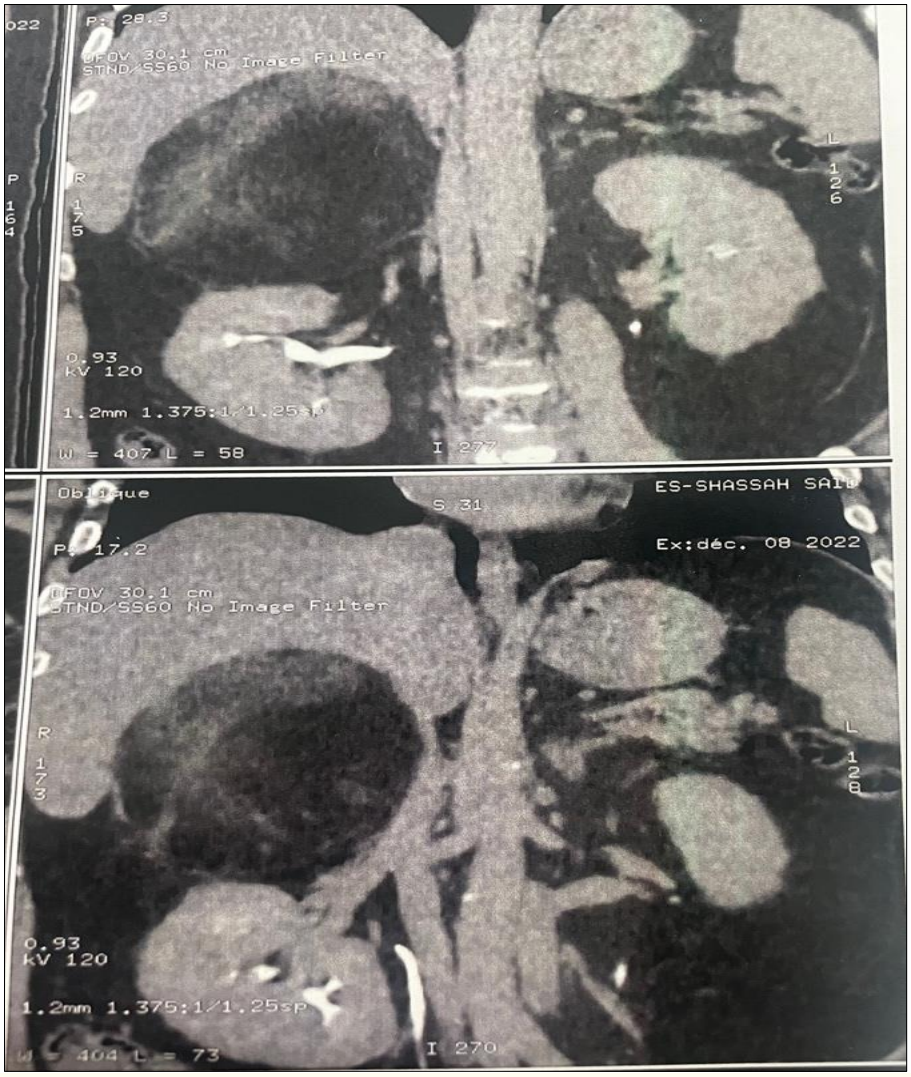


Figure 1: CT-scan revealed a right adrenal mass measuring 12.7 x 11 x 12.5 cm with characteristics suggestive of a myelolipoma

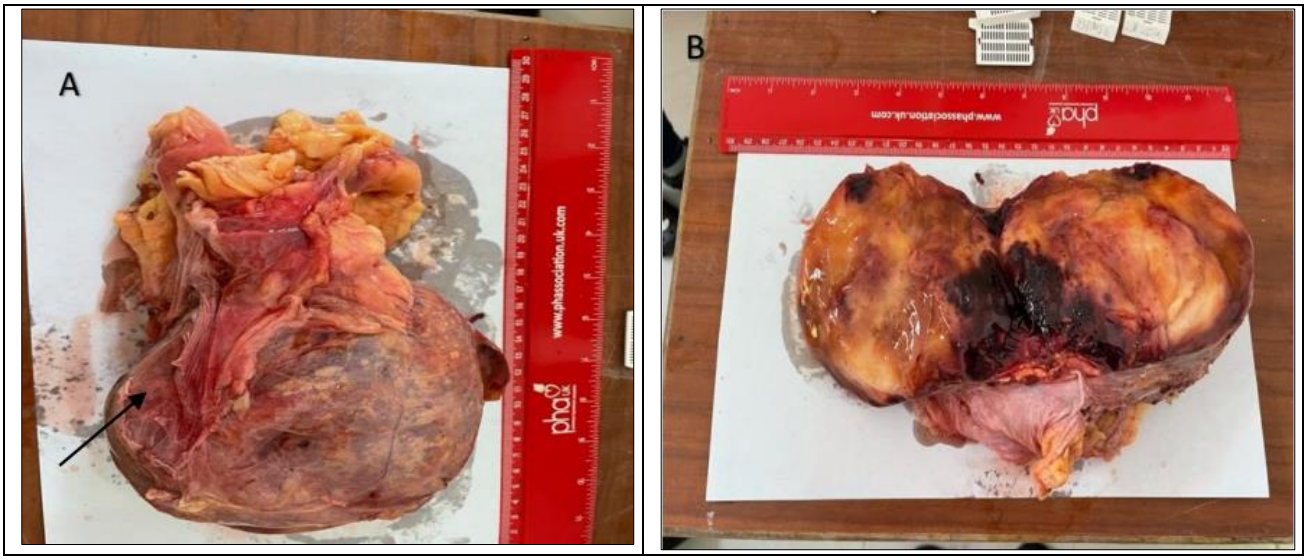


Figure 2: Macroscopic adrenalectomy specimen (formalin-fixated). A: (surface view). Round tumor with adherent perirenal fat and right adrenal gland (black arrowhead). B: (tumor cut open) Yellowish cut surface (adipose tissue) with haemorrhagic spots (myeloid tissue).

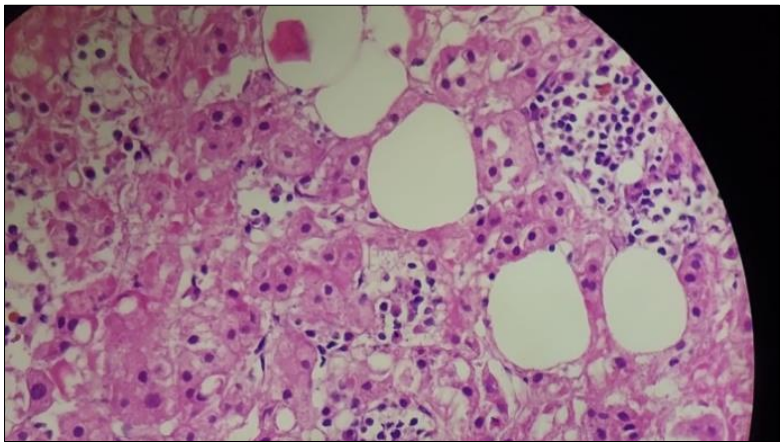
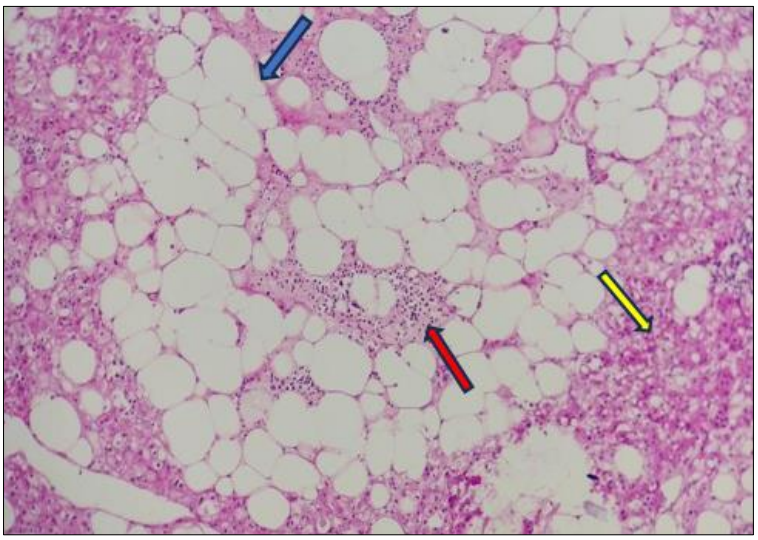


Figure 3: (A) Histopathologic picture of myelolipoma composed of mature fat cells mixed with hematopoietic elements.



B. Microscopic image showing mature hematopoietic tissue (Blue arrowhead) between mature fat cells (Red arrowhead) compatible with myelolipoma. In the bottom right corner (Yellow arrowhead) is visible.

DISCUSSION

Adrenal myelolipomas are the second most common adrenal incidentalomas comprising 6 to 16% of adrenal incidentalomas. They have an approximate autopsy prevalence of 0.08 to 0.2%. They are usually recognized in adulthood, with a median age of diagnosis around 51 years. There is no gender predilection. There may be a slight right-sided predilection, as reported by some studies [2].

The pathogenesis of myelolipoma is obscure. It is hypothesized that the primary event is a metaplastic change occurring in the reticuloendothelial cells of the blood capillaries due to stimuli like necrosis, infection or stress [5].

The size of adrenal myelolipomas is variable and can range from a few millimeters to greater than 10 cm. These are called giant adrenal myelolipomas. The largest adrenal myelolipoma reported to date weighed 6 kg. Extra-adrenal sites for myelolipomas include the retroperitoneum, thorax, and pelvis. Usually unilateral however they can also involve both adrenals [2].

The most common presenting symptoms for adrenal myelolipomas are as follows:

1. Abdominal pain--22.5%
2. Hypochondrial pain--13.9%
3. Flank pain--13.9%
4. Abdominal mass--5.2%

Rarely, dyspnea, back pain, fever, weight loss, and virilization can be the presenting symptoms of adrenal myelolipomas [2].

Ultrasonography (USG), CT, and MRI are all highly beneficial in the diagnosis of adrenal myelolipoma, with CT scan being the most sensitive for the identification of fat within the lesions. As these tumors are nonfunctional, endocrinological tests are not useful as these lesions are mostly functionally inert. The differential diagnosis of fat-containing retroperitoneal masses would include retroperitoneal lipoma or liposarcoma and renal angiomyolipoma [2].

Histological assessment can be made on the resected specimen. Thorough gross and microscopic evaluation is needed for confirmatory diagnosis and to exclude malignancy. Specimens should be sampled 1 cm apart for microscopic analysis in order not to miss any malignancy [3]. The characteristic microscopic appearance of myelolipoma is usually a dense adipose tissue, which can occasionally vary. The other constant component is trilineage hematopoietic tissue with the abundance of erythroid and granulocytic/lymphoid elements as well as megakaryocytes. Rarely, myelolipomatous foci were found in adrenocortical adenomas or ganglioneuroma. Other findings at histopathological examination in association with myelolipoma were osseous metaplasia, adrenal nodular hyperplasia, calcification, black pigmented adenoma, and hibernoma [5].

It's very important to differentiate between myelolipomas and other lipomatous adrenal tumors. These masses include adrenocortical adenoma, adrenocortical carcinoma, retroperitoneal liposarcoma, exophytic renal angiomyolipoma and adrenal lipoma [5].

Management of adrenal myelolipoma should be per individual basis. In a lesion of less than 4 cm in size, conservative treatment can be applied with an imaging technique. Surgery is indicated in symptomatic tumors, rapidly growing lesions, or tumors more than 6 cm in size. This is to avoid the risk of abdominal pain or life-threatening rupture and haemorrhage. The laparoscopic approach is more 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. However, laparoscopic adrenalectomy is not warranted for masses larger than 10 cm or with adhesions and infiltration of the surrounding structures. The prognosis of adrenal myelolipoma is remarkable with recurrence-free survival rates of up to 12 years [3].

CONCLUSION

Adrenal myelolipoma is commonly benign, asymptomatic, and hormonal inactivity. A surgical strategy is suggested for high-complication-risk patients [8]. Radiological findings help in early suspicion of this asymptomatic tumor [4].

The laparoscopic approach is safe and effective with an obvious advantage over open procedures [8], although histopathological examination remains the gold standard for definitive diagnosis of adrenal myelolipoma [4].

REFERENCES

1. Muzaffar, N., Hashmi, A., Mubarak, M., Mohsin, R., Shazad, A., Sultan, G., & Rizvi, A. H. (2009). Adrenal myelolipoma: a report of three cases and review of literature. *JPMA. The Journal of the Pakistan Medical Association*, *59*(7), 491-493. PMID: 19579745.
2. Bokhari, M. R., Zulfiqar, H., & Garla, V. V. (2023). Adrenal Myelolipoma. 2023 Mar 20. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing. PMID: 28613782.
3. Azizan, N., Myint, O., Wynn, A. A., Thein, T. T., Hayati, F., & Lah, N. A. S. N. (2020). A clinically silent tumour of adrenal myelolipoma: a case report. *International journal of surgery case reports*, *72*, 63-65. doi: 10.1016/j.ijscr.2020.05.056. Epub 2020 Jun 1. PMID: 32506033; PMCID: PMC7283099.
4. Tayade, S., Bondge, A. R., Buch, A., & Kumar, H. (2020). Adrenal myelolipoma: A rare

entity. *Medical Journal of Dr. DY Patil University*, *13*(1), 82-84. DOI: 10.4103/mjdrdypu.mjdrdypu_109_19.

5. Decmann, Á., Perge, P., Tóth, M., & Igaz, P. (2018). Adrenal myelolipoma: a comprehensive review. *Endocrine*, *59*, 7-15. doi: 10.1007/s12020-017-1473-4. Epub 2017 Nov 21. PMID: 29164520.
6. Calissendorff, J., Juhlin, C. C., Sundin, A., Bancos, I., & Falhammar, H. (2021). Adrenal myelolipomas. *The lancet Diabetes & endocrinology*, *9*(11), 767-775. doi: 10.1016/S2213-8587.
7. Cyran, K. M., Kenney, P. J., Memel, D. S., & Yacoub, I. (1996). Adrenal myelolipoma. *AJR. American journal of roentgenology*, *166*(2), 395-400. doi: 10.2214/ajr.166.2.8553954. PMID: 8553954.
8. Feng, Q., Li, H., Chen, X., Feng, X., & Li, J. (2022). Case report: Adrenal myelolipoma resected by laparoscopic surgery. *Frontiers in Oncology*, *12*, 1058211. doi: 10.3389/fonc.2022.1058211. PMID: 36544699; PMCID: PMC9760819.