EDORIUM Journals

Int J Case Rep Images 2024;15(2):34–37.
www.ijcasereportsandimages.com

CASE REPORT PEER REVIEWED | OPEN ACCESS

A case of adrenal myelolipoma: A rare encounter requiring specific approach in diagnosis and management

Kamel El-Reshaid, Sundus Hussein, Samer Abou-Deeb

ABSTRACT

Introduction: Myelolipoma is a rare, benign and non-functional tumor that predominantly develops in the adrenal gland.

Case Report: A 71-year-old man presented with large and symptomatic myelolipoma that indicated surgery for symptomatic relieve and definite diagnosis. Hormonal testing was negative for functioning adrenal tumors. Since the patient had moderate renal failure; contrast studies were avoided. Computed tomography (CT) scan showed 6×6 cm well circumscribed left adrenal mass with adipose tissue (very low CT density) and foci of calcifications. 18F-FDG-positron-emission tomography (PET)/CT scanning showed hypermetabolic activity limited to the tumor rim. Macroscopic examination showed foci of hemorrhage, necrosis and cystic degeneration with calcifications. Microscopic examination showed mature adipose tissue admixture with an extramedullary trilinear hematopoiesis without atypia of sarcomata and smooth muscle elements akin to angiomyolipomata.

Conclusion: The diagnosis of myelolipoma requires hormonal testing, CT/magnetic resonance imaging (MRI) scans and PET study with individualized management tailored to each case.

Keywords: Adrenal, Incidentaloma, Metaplasia, Myelolipoma

Kamel El-Reshaid[1], Sundus Hussein[2], Samer Abou-Deeb[3]

Affiliations: [1]Department of Medicine, Faculty of Medicine, Kuwait University, Kuwait City, Kuwait; [2]Department of Pathology, Mubarak Al-Kabeer Hospital, Ministry of Health, Kuwait City, Kuwait; [3]IMAGES Diagnostic Center, Kuwait City, Kuwait.

Corresponding Author: Dr. Kamel El-Reshaid, Professor, Department of Medicine, Faculty of Medicine, Kuwait University, Kuwait City, Kuwait, P O Box 24923, 13110 Safat, Kuwait; Email: kamel@hsc.edu.kw

Received: 18 May 2024
Accepted: 03 July 2024
Published: 23 August 2024

How to cite this article

El-Reshaid K, Hussein S, Abou-Deeb S. A case of adrenal myelolipoma: A rare encounter requiring specific approach in diagnosis and management. Int J Case Rep Images 2024;15(2):34–37.

Article ID: 101466Z01KE2024

****

doi: 10.5348/101466Z01KE2024CR

INTRODUCTION

Myelolipoma is a benign and non-functional tumor that predominantly develops in the adrenal gland. It was first described by Gierke in 1905 [1]. It was considered rare and an incidental autopsy finding with reports of only 300 cases by 2000 [2]. With computed tomography (CT), and magnetic resonance imaging (MRI); it became more common, constituting up to 10% of 4% incidental adrenal masses (incidentaloma) [3]. It is a form of metaplasia, in the gland, that leads to formation of mature adipose tissue and scattered islands of hematopoietic elements, resembling bone marrow [4]. The tumor is usually small and asymptomatic. However, its thin-walled vascular channels lined by a single layer of endothelial cells may exhibit bleeding in some cases that leads to loin pain and rapid enlargement that may culminate with catastrophic intraperitoneal rupture and shock [5]. Elective surgery can prevent a more severe symptom presentation and life-threatening progression and allows accurate diagnosis in patients with tumors larger than 6 cm [6]. In this case report, we present a patient with symptomatic and large myelolipoma that indicated surgery for symptomatic relieve and definite diagnosis.

CASE REPORT

A 71-year-old man presented with dull-aching left loin pain for two months. Past history was significant

for diabetes mellitus and hypertension for 15 years. Moreover, he had mild renal impairment for four years. His medications included: Alpha methyl-dopa, Amlodipine, insulin, statin, and Furosemide. The patient was short and obese with weight at 100 kg. Blood pressure was normal and he did not have peripheral edema. Systemic examination did not show abnormality. Laboratory investigations showed elevated serum creatinine at 240 umol/L and low albumin at 32 g/L with proteinuria. Thyroid function and serum (TSH), serum electrolytes, and liver function tests were normal. Hemoglobin was low at 110 g/L with normal transferrin saturation % and vitamin B12. Ultrasound of the abdomen revealed a large heterogenous mass at the upper pole of the left kidney. Computed tomography scan of the abdomen showed it to be well-defined lesion with adipose tissue (CT density—16 Hounsfield units) and foci of calcifications. It measured 65, 6×6, and 6×6 cm in anteroposterior (AP), transverse (TV), and craniocaudal (CC) dimensions, respectively (Figure 1). ^{18}F-FDG-PET/CT scanning form vertex to mid-thigh showed hypermetabolic activity only at the rim of the mass (SUV_{max} 4.0) with non-avid hypodense center (Figure 2). Hormonal tests showed normal AM Cortisol, adrenocorticotropic hormone (ACTH), and low-dose Dexamethasone suppression tests. Serum as well as 24-hour-urine for catecholamines, renin-aldosterone ratio, dehydroepiandrosterone sulfate (DHEAS), and testosterone. The latter findings were not in favor of (a) functioning tumor and (b) malignancy. However, left adrenalectomy was advised since the patient was symptomatic and medically stable as well as for definite diagnosis and cure. Conventional extraperitoneal approach was used since the tumor was large and surgery went without complications. On gross appearance, the left adrenal mass was partially covered with fibrofatty tissue, weighing 153 g and measuring 110×75×42 mm (Figure 3A). On sectioning, a pale yellow to brown encapsulated and well-circumscribed tumor was seen measuring 60×52×35 mm with foci of hemorrhage, necrosis, and cystic degeneration. The tumor was seen surrounded by a thin rim of normal-appearing golden yellow adrenal cortical tissue (Figure 3B). Microscopic examination showed a well-circumscribed lesion with a thin rim of normal appearing adrenal cortical tissue. It was mostly of organizing hematoma and in parts of variably vascular channels lined by a single layer of endothelial cells filled with blood and surrounded by fibrous tissue, islands of mature adipose tissue admixed with bone marrow elements including megakaryocytes, foci of osseous metaplasia, and dystrophic calcification (Figure 4A–C). There was no evidence of high-grade nuclear changes and increased mitotic activity. Moreover, there were smooth muscle elements akin to angiomyolipomata. Hence, the final diagnosis was consistent with large myelolipoma.

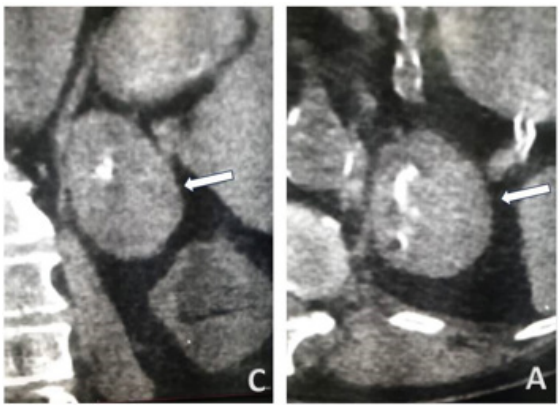

Figure 1: Coronal (C) and axial (A) views of left adrenal solid mass lesion (arrows) with coarse calcifications and small areas of fat density without necrosis and peripheral fat smudging.

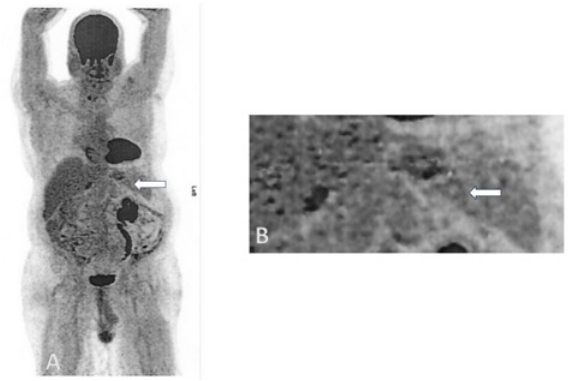

Figure 2: PET scanning form vertex to mid-thigh showed hypermetabolic activity only at the rim of the mass (SUV_{max} 4.0) with non-avid hypodense center (A) and magnified area (B).

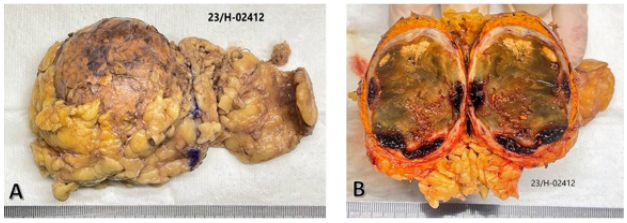

Figure 3: Left adrenal mass partially covered with fibrofatty tissue (A). On sectioning, it is well-circumscribed with foci of hemorrhage, necrosis, and cystic degeneration, and surrounded by a thin rim of normal-appearing golden yellow adrenal cortical tissue (B).

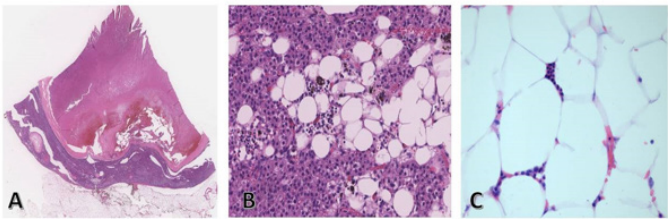

Figure 4: Histopathological picture of the adrenal mass showing hematoma associated with blood filled channels with admixture hemopoietic cells and fat lining a compressed gland rim (A), mature adipose tissue with trilinear hemopoietic elements (B), mature fate with hematopoietic cells including erythroid precursors and increased megakaryocytes (C) [H&E ×40, 200, 400, respectively].

DISCUSSION

Adrenal tumors (masses) are common with 10% prevalence in autopsy cases. They are subdivided into: (a) benign tumors, viz. adenoma, myelolipoma, neurofibroma, and hyperplasia; (b) malignant tumors, viz. adrenal carcinoma and neuroblastoma; and (c) metastatic tumors. Only 10% of pheochromocytoma are malignant. Adrenocortical adenoma is the most common lesion (52%) followed by carcinoma (12%), pheochromocytoma (11%), myelolipoma (8%), and metastasis from lung, breast, and gut (2%). A minority (<15%) of both benign and malignant lesions are functional, meaning they overproduce their respective hormones. These include cortisol, aldosterone, and DHES from adenomas and carcinomas, as well as catecholamines from medullary tumors such as pheochromocytomas and neuroblastomas [7]. Management of adrenal tumors requires close coordination among professionals from various medical fields. The majority (75%) of adrenal tumors are non-functioning, benign adenomas, and often do not require surgical intervention [8]. On the other hand, surgery is indicated in functioning benign and/or malignant adrenal tumors and metastases [9]. Functioning tumors overproduce cortisol (Cushing syndrome), aldosterone (Conn's disease), catecholamines (pheochromocytoma), and sex-steroid hormones (hyperandrogenism) [10]. For decision-making in management, three steps were indicated: (a) exclusion of functioning tumor, (b) exclusion of benign incidentaloma by abnormal CT findings, and (c) assessment for hypermetabolism in the mass indicative of malignancy. In this patient, with moderate renal disease, CT scan with contrast was not done to avoid contrast-induced nephropathy. Moreover, MRI with gadolinium-based dye was not done to avoid, in this patient with severe renal failure, nephrogenic systemic fibrosis. Enhancement of masses, following with CT and MRI dyes, is a valuable sign of malignancy. Hence, such third step was replaced by PET scanning. The existence and distribution of hypermetabolism in the masses is an alternative sign of malignancy in absence of infections. In our patient, hypermetabolism was limited to the rim only. Based on the above-mentioned data, the patient had (a) well-defined tumor borders, (b) foci of calcifications, and (c) rim hypermetabolism, which were more in favor of bleeding in a large myelolipoma rather than necrotizing malignant neoplasm [11]. Foci of calcifications, due to previous degeneration or differentiation of choristomatous primitive mesenchymal cells, have been reported in 25% of myelolipomata [12, 13]. Notably, though reported, fine needle aspiration is associated with inadequate tissue for diagnosis and core one with bleeding and seeding [14]. However, since the (a) tumor was large, (b) patient was acutely symptomatic (from bleeding), and (c) patient was not high-risk for surgery; surgery was advised to establish definite diagnosis and permeant cure. Histological examination ruled out malignancy, viz. liposarcoma, myxoid liposarcoma, and myofibrosarcoma as well as angiomyolipoma [15].

CONCLUSION

Myelolipoma diagnosis requires hormonal testing, CT/MRI scans, and PET study with individualized management tailored to each case.

REFERENCES

1. Gierke E. Uber knochenmarksgewebe in der Nebenniere. Beitr Z Pathol Anat 1905;37:311–25.
2. Nabi J, Rafiq D, Authoy FN, Sofi GN. Incidental detection of adrenal myelolipoma: A case report and review of literature. Case Rep Urol 2013;2013:789481.
3. Low G, Dhliwayo H, Lomas DJ. Adrenal neoplasms. Clin Radiol 2012;67(10):988–1000.
4. Feng C, Jiang H, Ding Q, Wen H. Adrenal myelolipoma: A mingle of progenitor cells? Med Hypotheses 2013;80(6):819–22.
5. Kim DS, Lee JW, Lee SH. Spontaneous rupture of adrenal myelolipoma as a cause of acute flank pain: A case report. World J Clin Cases 2021;9(22):6552–6.
6. Wani NA, Kosar T, Rawa IA, Qayum A. Giant adrenal myelolipoma: Incidentaloma with a rare incidental association. Urol Ann 2010;2(3):130–3.
7. Lam AKY. Update on adrenal tumours in 2017 World Health Organization (WHO) of endocrine tumours. Endocr Pathol 2017;28(3):213–27.
8. Bovio S, Cataldi A, Reimondo G, et al. Prevalence of adrenal incidentaloma in a contemporary computerized tomography series. J Endocrinol Invest 2006;29(4):298–302.
9. Gimm O, Duh QY. Challenges of training in adrenal surgery. Gland Surg 2019;8(Suppl 1):S3–9.
10. Mongiat-Artus P, Miquel C, Meria P, Hernigou A, Duclos JM. Adrenocortical secretory tumors. [Article in French]. Ann Urol (Paris) 2004;38(4):148–72.
11. Mayo-Smith WW, Song JH, Boland GL, et al. Management of incidental adrenal masses: A white paper of the ACR incidental findings committee. J Am Coll Radiol 2017;14(8):1038–44.
12. Rossi A, Incensati R. Bone tissue in adrenal myelolipoma: A case report. Tumori 1998;84(1):90–3.
13. Lee JKT, Sagel SS, Stanley RJ, Heiken JP. Computed Body Tomography with MRI Correlation. 4ed. Philadelphia: Lippincott Williams & Wilkins; 2006.
14. Hsu SW, Shu K, Lee WC, Cheng YT, Chiang PH. Adrenal myelolipoma: A 10-year single-center experience and literature review. Kaohsiung J Med Sci 2012;28(7):377–82.
15. Lam KY, Lo CY. Adrenal lipomatous tumours: A 30 year clinicopathological experience at a single institution. J Clin Pathol 2001;54(9):707–12.

****

Author Contributions

Kamel El-Reshaid – Conception of the work, Design of the work, Acquisition of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Sundus Hussein – Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Samer Abou-Deeb – Acquisition of data, Analysis of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission

The corresponding author is the guarantor of submission.

Source of Support

None.

Consent Statement

Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.