

Myelolipoma of Kidney: A Rare Extra-Adrenal Tumor in an Unusual Site

Abstract

Myelolipoma of the kidney is a very rare benign disease which is composed of mature adipose tissue admixed with mature hematopoietic elements in varying proportions. The most common location of myelolipoma is in the adrenal glands, but extra-adrenal myelolipomas in other locations such as the retroperitoneum, presacral region, pelvis, and mediastinum has also been reported. In this case report, we present a case of huge extra-adrenal myelolipoma in renal parenchyma which is rare site for its origin. We report a case of extra-adrenal myelolipoma occurring in the left kidney of a 46-year-old Indian man. We describe the radiological and clinicopathologic features of this unusual tumor with a review of the literature. This case is noteworthy because the tumor was very enlarged and its site was unusual. It is generally impossible to distinguish extra-adrenal myelolipoma from other retroperitoneal tumors by radiological imaging given the rarity of tumor more cases need to be reported to elucidate its biological behavior.

Keywords: *Extra-adrenal, kidney, myelolipoma*

Introduction

Myelolipoma of the kidney is a very rare benign disease which may resemble other renal lesion majority of clinicians may be unaware of its entity. It is composed of mature adipose tissue admixed with mature hematopoietic elements in varying proportions. The most common site of involvement is the adrenal gland. Its incidence in extra-adrenal sites is 0.4% at autopsy.^[1] Extra-adrenal myelolipomas have reported in the retroperitoneum, pelvis, thorax, and mediastinum.^[2] Worldwide myelolipoma in the kidney or around the kidney has been reported in <10 cases. Here, we present a case of very large renal myelolipoma along with its radiological and clinicopathological discussion.

Case Report

A 46-year-old Indian man, presented in our hospital with left flank pain and left loin swelling 1 year back. Physical examination revealed a well-defined lump in left hypochondrium and lumbar region which was firm in consistency. [Figure 1] Ultrasonography (US) of his abdomen showed a huge mass of mixed echogenicity in the left renal fossa. Computed tomography (CT) of his

abdomen and pelvis a large heterogeneous predominantly fat density lesion of 15.2 cm × 20.2 cm × 26 cm arising from left kidney occupying nearly entire left lumbar and iliac fossa region showing discrete enhancing soft-tissue density areas (<20 Hounsfield units) within. Multiple radio-dense calculi noted in renal pelvis and upper, mid and lower calyces of left kidney the largest calculus in renal pelvis measures 26.5 mm × 12.5 mm × 22 mm. [Figure 2] His adrenal glands were reported normal and no lymphadenopathy was detected. Laparotomy through chevron incision was performed. Intraoperatively, lesion was well circumscribed, surgical planes with surrounding structures were maintained and the mass was removed.

On gross examination, the left kidney along with mass removed from the patient was irregular in shape and weighed 4.2 kg. Cut surface showed very small area of residual renal parenchyma with staghorn calculus. The pelvic calyceal system is lined with yellowish white flakes and a large tumor composed of fatty areas with foci of hemorrhage.

Microscopic examination of the specimen showed changes of chronic pyelonephritis with focal xanthomatous changes and tumor composed of adipocytes. The intervening septa show hematopoietic tissue

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: WKHLRPMedknow_reprints@wolterskluwer.com

How to cite this article: Agrawal S, Patil A, Singh AG, Ganpule AP, Sabnis RB, Desai MR. Myelolipoma of Kidney: A rare extra-adrenal tumor in an unusual site. *Int J App Basic Med Res* 2021;11:60-2.

**Shashank Agrawal,
Abhijit Patil,
Abhishek Gajendra
Singh,
Arvind Prakash
Ganpule,
Ravindra
Bhalchandra
Sabnis,
Mahesh Ramanlal
Desai**

*Departments of Urology,
Muljibhai Patel Urological
Hospital, Nadiad, Gujarat, India*

Submitted: 29-May-2020

Revised: 28-Jul-2020

Accepted: 19-Dec-2020

Published : 26-Jan-2021

Address for correspondence:

*Dr. Shashank Agrawal,
Department of Urology,
Muljibhai Patel Urological
Hospital, Dr Virendra desai
Road, Nadiad - 387 991,
Gujarat, India.
E-mail: shank789agrawal@
gmail.com*

Access this article online

Website:
www.ijabmr.org

DOI:
10.4103/ijabmr.IJABMR_341_20

Quick Response Code:



with cells of all three hematopoietic lineages (myeloid, erythroid, and megakaryocytes). No adrenal rest were found. Based on these findings, renal myelolipoma was diagnosed. Postoperative course of patient was uneventful. At follow-up of 1 year, he was asymptomatic. [Figure 3]

Discussion

Myelolipoma are benign lesions that comprise of adipose tissue and hematopoietic elements and commonly have been reported in the adrenal gland in comparison with in other organs.^[3] Myelolipoma was first described in 1905 by Gierke since then myelolipoma have been reported sporadically in various organs.^[4] The pathogenesis of myelolipoma is not exactly unknown. One theory postulated that it is hamartoma or groups of choristomatous hematopoietic stem cells that transmigrated into the others sites during intrauterine life, while other theory found clonal cytogenetic abnormality, which suggested it was a renal tumor origin.^[5-7]

Patients with either adrenal or extra-adrenal myelolipoma are typically asymptomatic and are diagnosed incidentally based upon radiological imaging for a different condition based on ultrasound scan, CT scan, or magnetic resonance imaging scan.

No definitive radiological criteria exist for diagnosing extra-adrenal myelolipoma however the characteristics of adrenal myelolipoma on imaging can be exploited to identify these structures which present in a variety of locations.^[8] Extra-adrenal myelolipoma features on imaging can mimic with several malignancies such as retroperitoneal liposarcoma, a renal or adrenal myelolipoma, renal angiomyolipoma or a retroperitoneal teratoma.

Preoperatively, an attempt for definite diagnosis can be made with a fine-needle biopsy under US or CT guidance which would then reveal fat tissue lesion admixed with variable amount of hematopoietic tissue and no malignant cells. Differential diagnosis after biopsy can extramedullary hematopoiesis and extra-adrenal myelolipoma, which can be distinguished from latter by the presence of splenomegaly or other organomegaly, chronic anemia, and marked hyperplasia of the bone marrow.^[9] In our case, after evaluating the CT scan which contained low density (<20 HU) soft-tissue lesion arising from the kidney angiomyolipoma, myelolipoma of kidney, liposarcoma were kept as differential diagnosis. Preoperative biopsy was not done as patient was planned for left nephrectomy in view of large mass effect causing pain to patient.

Grossly, extra-adrenal myelolipoma is a solitary circumscribed mass ranging in size from a few centimeters to 27 cm.^[10] It is a well-circumscribed, nonencapsulated, solid mass in the kidney. The cut surface of myelolipoma would tend to reveal areas of soft yellow fatty tissue together with admixed with irregular areas of brownish friable tissue.^[11] Microscopically, myelolipoma is a mixture

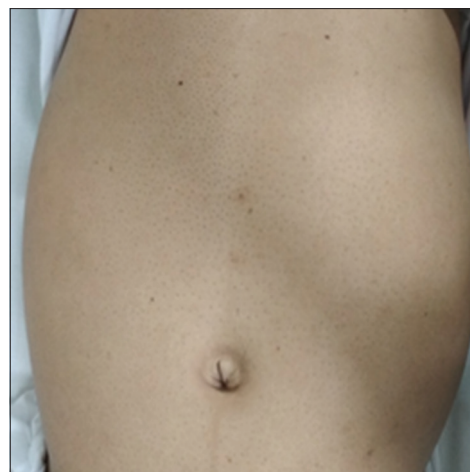


Figure 1: Lump in left hypochondrium and loin visible

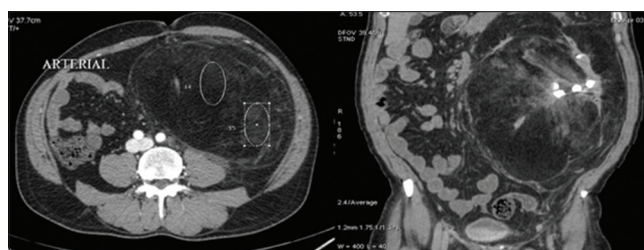


Figure 2: Computed tomography of abdomen showing a large heterogeneous predominantly fat density lesion is noted arising from left kidney occupying nearly entire left lumbar and iliac fossa region showing discrete enhancing soft-tissue density areas within

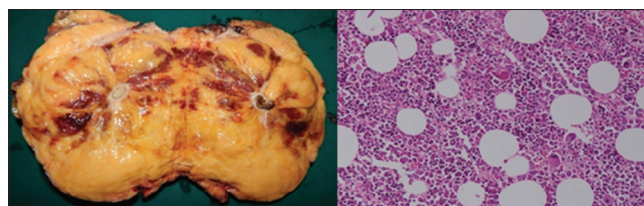


Figure 3: Gross photograph showing a large tumor of fatty areas with staghorn calculi with foci of hemorrhage. Higher power magnification (x200) showing normal hematopoietic elements, including megakaryocytic, erythropoietic and granulopoietic cell lineages interspersed within adipose tissue

of adipose tissue and normal hematopoietic cells with all the three hematopoietic cell lineages (granulocytic, erythroid, as well as megakaryocytic). Kidney myelolipoma does not tend to have connection to medullary cavity of any bone.

Some of the differential diagnoses of myelolipoma of the kidney include: angiomyolipoma, sarcoma including liposarcoma, extramedullary hematopoietic tumors, renal cell carcinoma.

Angiomyolipoma represents one of the most common benign renal lesions.^[12] Microscopically, typical angiomyolipoma is a triphasic tumor which carries varying amounts of dysplastic blood vessels, spindle and epithelioid smooth muscle cells and mature adipose elements.

Fat-predominant angiomyolipoma mimic other lesions such as liposarcoma and myelolipoma.

Liposarcoma involves middle-aged adults with a peak incidence in the 6th decade, with no sex predominance. Retroperitoneal liposarcoma are often asymptomatic until the tumor has become big in size. Very few cases of liposarcoma located in renal parenchyma has been reported.^[13] On gross examination it consists usually of a large, well-circumscribed, lobulated mass with variable color from yellow to white depending on the proportion of adipocytes, fibrous and/or myxoid areas. Microscopically, it is composed either entirely or in part of a mature adipocytes proliferation showing a significant varying number of lipoblasts. In our case, the tumor contained, in addition to mature adipose tissue, nests of hematopoietic elements were seen.

Extra medullary hematopoietic tumors are characterized by anemia, frequent hepatosplenomegaly, and abnormal peripheral blood smears. Extra-adrenal myelolipoma are well circumscribed but extra medullary hematopoietic tumors lack circumscription and are ill defined. Microscopically, extra medullary hematopoietic tumors have a predominance of hematopoietic elements, with erythroid hyperplasia. Fat is not an enlarged component of the process.^[14]

If myelolipoma is diagnosed is established prehand, then subsequent management depends upon its size and symptomatic status of patient. Small asymptomatic lesions may be managed expectantly. Radiological follow-up is recommended to look for growth and hemorrhage. In symptomatic patients, who possess an enlarging tumor mass, as in our patient, surgical intervention is warranted. Long-term prognosis is excellent.

Conclusion

Our case was different because the location of giant extra-adrenal myelolipoma in renal parenchyma is extremely rare. It is generally impossible to distinguish extra-adrenal myelolipoma from other retroperitoneal tumors by radiological imaging. Hence, the final diagnosis should depend on the histopathological features of surgically removed specimen. Acknowledging the rarity of tumor, more cases need to be reported for elucidation of biological behavior of the disease.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and

other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Prahlow JA, Loggie BW, Cappellari JO, Scharhng JO, Teot A, Iskandar SS: Extra-adrenal myelolipoma: Report of two cases. *South Med J* 1995;88:639-43.
2. Xuefeng T, Rui C, Jianping X, Mingu Y: Myelolipoma of the kidney: A seldom site for a rare extra-adrenal tumor. *J Med Coll PLA* 2010;25:317-20.
3. Venyo AKG. Myelolipoma of the kidney: A review and update. *J Kidney Treat Diagn*. 2018;1:8-15.
4. Gierke E. Unusual myeloid tissue in the adrenal gland. *Beitr Pathol Anat* 1905;7:311-25.
5. Chang KC, Chen PI, Huang ZH, Lin YM, Kuo PL. Adrenal myelolipoma with translocation (3;21)(q25;p11). *Cancer Genet Cytogenet* 2002;134:77-80.
6. Bishop E, Eble JN, Cheng L, Wang M, Chase DR, Orazi A, *et al.* Adrenal myelolipomas show non-random X-chromosome inactivation in hematopoietic elements and fat: Support for a clonal origin of myelolipomas. *Am J Surg Pathol* 2006;30:838-43.
7. Amin MB, Tickoo SK, Schultz D. Myelolipoma of the renal sinus. An unusual site for a rare extra- adrenal lesion. *Arch Pathol Lab Med* 1999;123:631-4.
8. Nguyen B: Retroperitoneal extraadrenal myelolipoma: Technitium-99m sulfur colloid scintigraphy and CT imaging. *Clin Nucl Med* 2007;32:135-8.
9. Fowler MR, Williams RB, Alba JM, Byrd CR. Extra-adrenal myelolipomas compared with extramedullary hematopoietic tumors: A case of presacral myelolipoma. *Am J Surg Pathol* 1982;6:363-74.
10. Shapiro JL, Goldblum JR, Dobrow DA, Ratliff NB: Giant bilateral extraadrenal myelolipoma. *Arch Pathol Lab Med* 1995;119:283-5.
11. Ghaouti M, Znati K, Jahid A, Zouaidia F, Bernoussi Z, Mahassini N, *et al.* Renal myelolipoma a rare extra-adrenal tumor in a rare site: A case report and review of the literature. *J Med Case Rep* 2013;7:92.
12. Armah BH, Yin M, Rao NM, Parwani VA: Angiomyolipoma with epithelial cysts (AMLEC): A rare but distinct variant of angiomyolipoma. *Diagn Pathol* 2007;2:11.
13. Bader AL, Peres AB, Bader SL: Renal liposarcoma. *Int Braz J Urol* 2004;30:214-5.
14. George SA, Manipadam MT, Thomas R. Primary myelolipoma presenting as a nasal cavity polyp: A case report and review of the literature. *J Med Case Rep* 2012;6:127.