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## Case Report

# Multifocal extra-adrenal myelolipomas with bilateral perirenal and retroperitoneal nodal involvement—Computed tomography features <sup>☆</sup>

Waylon Wai Chung Tsui, MBBS\*, Man Hon So, MBBS

Department of Diagnostic &amp; Interventional Radiology, Kwong Wah Hospital, 25 Waterloo Road, Yaumatei, Kowloon, Hong Kong

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## ABSTRACT

Extra-adrenal myelolipomas are exceedingly rare benign tumors composed of adipose and myeloid tissues, which have been reported to occur in various sites including the retroperitoneum, pelvis, and thorax. Myelolipomas are more commonly encountered in the adrenal glands. We illustrate a case of a 72-year-old woman with surgically proven bilateral perirenal and para-aortic lymph nodal myelolipomas detected incidentally by computed tomography. Extra-adrenal myelolipomas can be difficult to distinguish from other fat-containing lesions particularly liposarcomas, which are more commonly encountered in the retroperitoneum. This case highlights the unusual multifocal involvement of extra-adrenal myelolipomas and despite its rare occurrence, should be included in the differential diagnosis of retroperitoneal lipomatous lesions.

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## Introduction

Myelolipomas are uncommon non-functional benign tumors of mesenchymal origin which consists of mature adipose tissue intermixed with mature hematopoietic elements [1]. The tumor predominantly involves the adrenal glands with a reported incidence rate ranging from 0.08% to 0.4% discovered at autopsy [2]. Involvement in extra-adrenal sites is rare, with an incidence of 14% of all myelolipomas [3].

Sites involved are varied, including the presacral, pelvis, retroperitoneum, thorax, and mediastinum.

In the literature, there have only been about 10 reported cases of perirenal myelolipomas, with bilateral cases being much rarer in occurrence. To the best of our knowledge, this is the first case of bilateral perirenal myelolipomas with retroperitoneal lymph node involvement reported.

## Case report

A 72-year-old woman with a history of chronic renal failure on continuous ambulatory peritoneal dialysis was

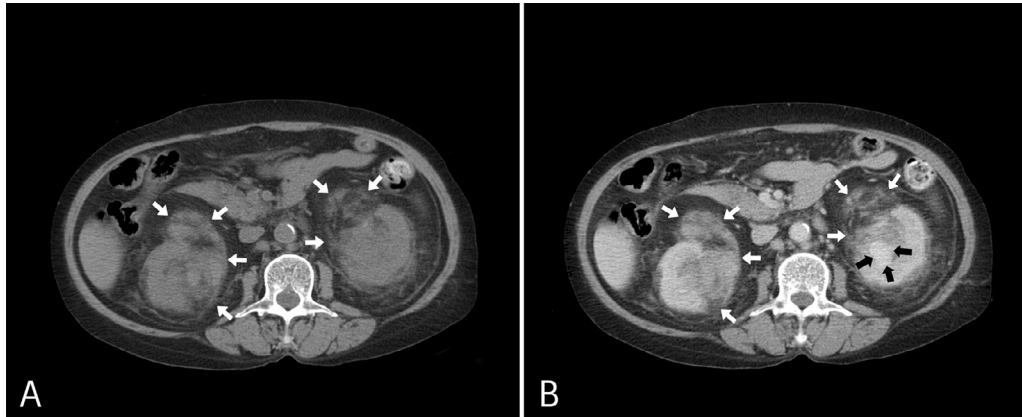
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\* Corresponding author.

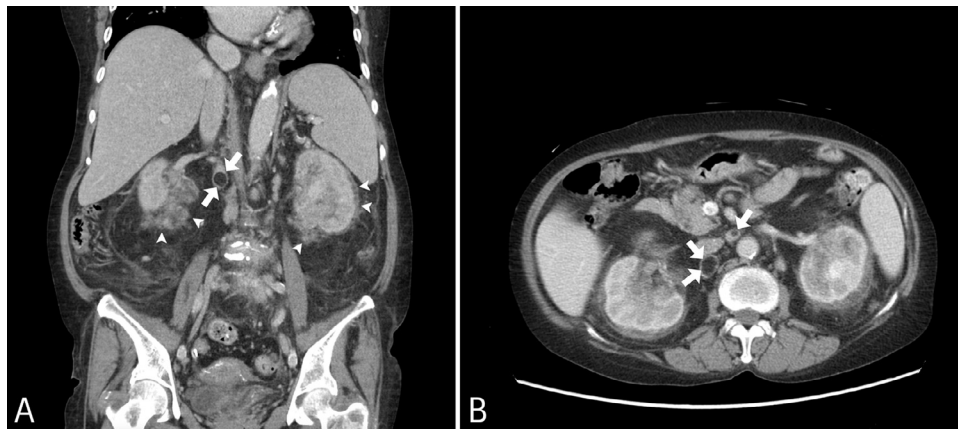
E-mail address: [waylont@gmail.com](mailto:waylont@gmail.com) (W.W.C. Tsui).

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**Fig. 1 – Noncontrast (A) and contrast-enhanced (B) axial CT images of the abdomen showing mixed fat and soft tissue attenuation lesions involving bilateral perirenal regions and renal sinuses (white arrows). A prominent enhancing soft tissue nodule is detected in the left renal sinus (black arrows). No evidence of renal invasion is seen.**



**Fig. 2 – Contrast-enhanced coronal (A) and axial (B) CT images of the abdomen showing low attenuation components in a non-enlarged aortocaval lymph node and an enlarged retrocaval lymph node (arrows). Bilateral perirenal mixed fat and soft tissue lesions are also present (arrowheads).**

admitted for acute abdominal pain. She developed low-grade fever upon admission. Physical examination revealed generalized abdominal tenderness. Dialysis fluid was turbid in appearance. Blood tests showed a mildly elevated white cell count. An urgent CT scan of the abdomen and pelvis was performed for assessment of possible peritoneal dialysis-associated peritonitis.

CT scan showed no evidence of peritonitis or intra-abdominal sepsis. Incidentally, there were bilateral perirenal mixed fat and soft tissue containing lesions seen surrounding the kidneys and more extensively involving the bilateral renal sinuses. Prominent enhancing soft tissue components were seen bilaterally which are larger in the left kidney (Fig. 1). Multiple enlarged retroperitoneal lymph nodes involving the aortocaval and retrocaval regions with a few nodes containing macroscopic fat components were detected (Fig. 2). No evidence of renal vasculature involvement was seen.

Given the presence of prominent solid enhancing soft tissue components in the left kidney, there was concern for malignancy. Due to the high suspicion of renal cell carcinoma in

this patient with chronic kidney disease, a review of the case in the urological cancer multidisciplinary meeting decided for surgical treatment with left radical nephrectomy together with open biopsy of the right kidney subsequently performed. Pathology revealed the presence of mature adipose tissue admixed with hematopoietic cells from erythrocytic, granulocytic, and megakaryocytic lineages. Lymphoplasmacytic infiltrate was found rimming the renal parenchyma. No evidence of malignancy was present. Pathologic evaluation confirmed the diagnosis of extra-adrenal myelolipomas with bilateral perirenal and retroperitoneal lymph nodes involvement.

## Discussion

Extra-adrenal myelolipomas are exceedingly rare tumors. They tend to occur more frequently in women and in middle-aged to older patients [1]. These lesions are usually asymptomatic and are thus detected incidentally. Occasionally,

myelolipomas may present as abdominal pain reflecting the development of complications such as intra-tumoral hemorrhage, tumor infarction, or mechanical compression [1].

The pathogenesis remains unclear, although there are hypotheses postulating chromosomal translocations. However, no genetic predisposition has been reported in the literature. Other hypothesized pathogenesis of extra-adrenal myelolipoma include distant embolization of bone marrow tissue or the embryonic connective tissue reactivation as a response to stimulus such as sepsis or endocrine dysfunction [3]. Malignant transformation of myelolipomas has never been reported in the literature [4].

The adrenal gland is the most common site for myelolipoma involvement. Extra-adrenal myelolipomas tend to occur in the presacral region, accounting for half of all cases [3]. Other sites include the retroperitoneum, thorax, and mediastinum [5]. In contrast to adrenal myelolipomas, extra-adrenal lesions tend to have a lower proportion of fat (<50%) compared to adrenal ones (50%–90%) [6].

Myelolipomas show variable appearance depending on its composition of fatty and myeloid tissue. Sonographically, myelolipomas typically appear as hyperechoic lesions [6]. CT often reveals a heterogeneous mass surrounded by a pseudocapsule with lipomatous component showing negative Hounsfield units and the myeloid component showing higher attenuation and contrast enhancement [7]. Calcification is rarely seen.

MRI will show the fat component of the tumor as hyperintense signal on both T1-weighted and T2-weighted sequences. The myeloid component shows low signal intensity in T1-weighted imaging. Fat suppression techniques confirms the presence of macroscopic fat. Chemical shift artifacts show the presence of Indian ink appearance along the fat and water interface of the lesion [7].

Nuclear medicine with technetium-99m sulfur colloid scan allows detection of myeloid elements in myelolipomas, which helps to differentiate from liposarcoma or angiomyolipoma due to the absence of erythroid cells in these lesions [7].

Differential diagnoses include other fat-containing lesions such as extramedullary hematopoiesis, angiomyolipoma, and liposarcoma [8]. Differentiation of these lesions with imaging alone can be difficult due to overlapping imaging appearances. Extramedullary hematopoiesis is a compensatory process in response to chronic anemia with imaging features of soft tissue masses as well as associated findings of hepatosplenomegaly and medullary expansion of bone [7]. Angiomyolipoma is a common benign neoplastic lesion consisting of varying amounts of blood vessels, smooth muscle cells, and adipose tissue [1]. Patients with tuberosus sclerosis can present with multiple bilateral angiomyolipomas. Both CT and MRI demonstrate intralésional macroscopic fat and the presence of enlarged vessels or aneurysms [6]. Well-differentiated liposarcomas can also present with varying amounts of fat tissue and enhancing soft tissues, which tend to be large with lobulated margins [1]. Occasionally, cystic components are seen within the lesion. It is important to note

the presence of calcifications or ossification within the lesion as these suggest tumor dedifferentiation [6].

Biopsy is usually required to confirm the diagnosis. Histopathologic analysis reveals mature adipocytes with varying degrees of hematopoietic cell lineages including megakaryocytes, granulocytic precursors, and erythroid precursors [8]. Immunohistochemical and molecular testing has no major role currently. Surgery is contemplated when the imaging and biopsy findings are not definitive or when the patient develops symptoms [9].

Currently, there is no standard treatment. Small tumors less than 4cm in size should be monitored while large or symptomatic tumors should be surgically intervention [8].

## Conclusion

Although most myelolipomas occur in the adrenals, it is important to consider these in extra-adrenal sites. Due to the presence of macroscopic fat on imaging with CT or MRI, extra-adrenal myelolipomas in the retroperitoneum may be mistaken for other fat-containing lesions such as liposarcoma; a more commonly encountered retroperitoneal lipomatous lesion.

## REFERENCES

- [1] Ghaouti M, Znati K, Jahid A, Zouaidia F, Bernoussi Z, Mahassini N. 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. doi:10.1186/1752-1947-7-92.
- [2] Ramirez M, Misra S. Adrenal myelolipoma: to operate or not? A case report and review of the literature. *Int J Surg Case Rep* 2014;5(8):494–6. doi:10.1016/j.ijscr.2014.04.001.
- [3] Yelikar BR, Mamatha K, Chawla A, Kathpal N. Extra-adrenal myelolipoma: tumour at an unusual site. *J Clin Diagn Res* 2017;11(12):ED01–2. doi:10.7860/JCDR/2017/30916.10935.
- [4] Stengel B, Dunker H. Extraadrenal intralymphonodular myelolipoma. *Pathol Res Pract* 1989;184(6):639–41. doi:10.1016/S0344-0338(89)80171-2.
- [5] Cox A, Offman SL, Merrimen JLO, Kew A, Norman RW. Bilateral renal sinus myelolipomas. *Can Urol Assoc J* 2010;4(6):164–8. doi:10.5489/auaj.971.
- [6] Craig WD, Fanburg-Smith JC, Henry LR, Guerrero R, Barton JH. Fat-containing lesions of the retroperitoneum: radiologic-pathologic correlation. *Radiographics* 2009;29(1):261–90. doi:10.1148/rg.291085203.
- [7] Hakim A, Rozeik C. Adrenal and extra-adrenal myelolipomas – a comparative case report. *J Radiol Case Rep* 2014;8(1):1–12. doi:10.3941/jrcr.v8i1.1551.
- [8] Hajiran A, Morley C, Jansen R, Kandzari S, Baca P, Zaslau S. Perirenal extra-adrenal myelolipoma. *World J Clin Cases* 2014;2(7):279–83. doi:10.12998/wjcc.v2.i7.279.
- [9] Temizoz O, Gençellac H, Demir MK, Unlu E, Ozdemir H. Bilateral extra-adrenal perirenal myelolipomas: CT features. *Br J Radiol* 2010;83(994):e198–9. doi:10.1259/bjr/28801968.