

A Large Intra-Abdominal, Extra Adrenal Myelolipoma- Case Report

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Abstract

Introduction: Myelolipomas are benign tumors composed of mature adipose tissue and normal hematopoietic cells. Myelolipomas develop in older age group and usual site of occurrence is adrenal gland. Their development in young individuals and at extra-adrenal site is quite rare. Very few cases of extra-adrenal myelolipomas have been reported in the literature. An extra-adrenal myelolipoma is solitary, well defined mass in abdomen, commonly occurring in retroperitoneal area. The small sized lesions are usually asymptomatic, but if large in size they can produce symptoms related to mass effect. The patient has no hematological abnormalities.

Case Presentation: We present a case of a large intra abdominal extra-adrenal myelolipoma measuring 28 x 22 x 6cm in a 29 years old male patient who presented with abdominal pain and distention. His abdominal CT scan showed a large well defined retroperitoneal lesion measuring 13.1 x 10.3 x 22cm in Anteroposterior x Transverse x Craniocaudal extent, in the region of left paraventricular gutter having fat density may represent liposarcoma. His both adrenals appeared normal. He was operated for this lesion and the specimen was sent to us for histopathological evaluation. Grossly it was solitary, encapsulated mass. Cut section showed fatty tissue with hemorrhagic areas. Microscopically, mature adipose tissue with areas of mature hematopoietic cell of all lineages was seen.

Conclusion: Since extra-adrenal myelolipoma is a rare entity diagnosis of this lesion based on radiographic studies alone is at times difficult. It is important to distinguish extra-adrenal myelolipoma from other fat-containing tumors in particular retroperitoneal liposarcoma, extra-medullary hematopoietic neoplasms and angiomyolipoma. Histopathologic evaluation is the gold standard for confirmation of the diagnosis. We have discussed the background, pathology, complications, differential diagnosis and management of this rare entity.

Keywords: Myelolipoma, Extraadrenal myelolipoma, Hematopoietic neoplasm.

Introduction

Myelolipomas are benign tumors involving the adrenal glands, composed of mature adipose tissue and normal hematopoietic cells.¹ Myelolipomas have an incidence of 0.08–0.2% and account for 3–5% of all primary tumors of adrenals.² Their occurrence in an extra-adrenal site is quite atypical and rare.³ Very few cases of extra-adrenal myelolipomas have been reported in the literature. These tumors develop in older individuals in their 5th to 7th decade of life and affect

both the genders equally.⁴ The size of these tumors varies, from less than 4cm in diameter up to 31 x 24.5 x 11.5cm reported as the largest myelolipoma⁵ in the literature. Our patient had a larger myelolipoma than the one reported in the literature. Small sized tumors are asymptomatic and picked up as incidental findings on radiographic imaging. As their size increases, these tend to produce symptoms related to their mass effect.¹ Patient has no hematological abnormalities. The natural history of these lesions is not well understood⁶, but these do not undergo malignant transformation. Myelolipoma should be surgically excised because they may result in extra-peritoneal or intra-tumoral haemorrhage.⁷

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Case Report

A 29 years old man presented with fever, abdominal pain and distention. He was admitted and his investigations were done.

His plain abdomen-pelvis ultrasound radiography showed a large well defined hyper-echoic lesion with few hypo to anechoic areas in the left hypochondrium displacing the left kidney inferomedially towards the midline, abutting the spleen superiorly with its inferior extension upto the left iliac fossa.

His abdominal CT scan showed a large well defined heterogeneously enhancing retroperitoneal lesion measuring 13.1 x 10.3 x 22cm in Anteroposterior x Transverse x Craniocaudal extent, in the region of left paraventricular gutter having fat density may represent liposarcoma. However the possibility of other sarcomatous lesions cannot be completely ruled out. Pancreas, spleen, both adrenals appeared normal.

His surgical resection was done for this lesion and the specimen was sent for histopathological evaluation.

Gross examination showed an encapsulated large mass measuring 28 x 22 x 6cm. External surface showed congestion. Cut surface had a variegated appearance showing tan yellow areas of fatty tissue and hemorrhagic areas. No foci of necrosis or calcification were identified on serial sectioning. (Figure 1 and Figure 2)



Figure 1. Gross appearance of the mass



Figure 2. Cut surface of this large mass showing the fatty and hemorrhagic areas.

Microscopic examination revealed a benign tumor composed of intricate mixture of fat cells and normal hematopoietic cells of all lineages. No evidence of cellular atypia was seen in sections examined. Diagnosis of a large extra adrenal myelolipoma was rendered. (Figure. 3 & 4) (H & E X 100)

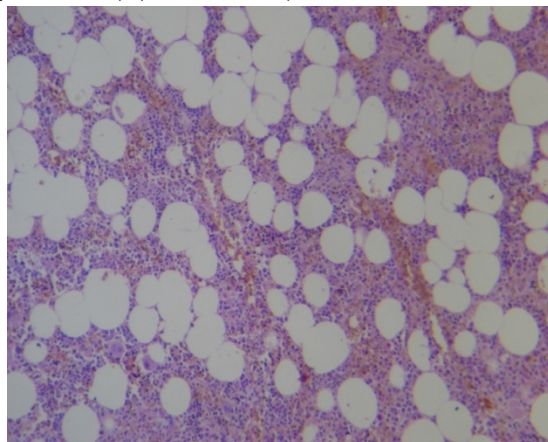


Figure 3. This micrograph shows a mixture of fat cells and normal hematopoietic cells of all lineages.

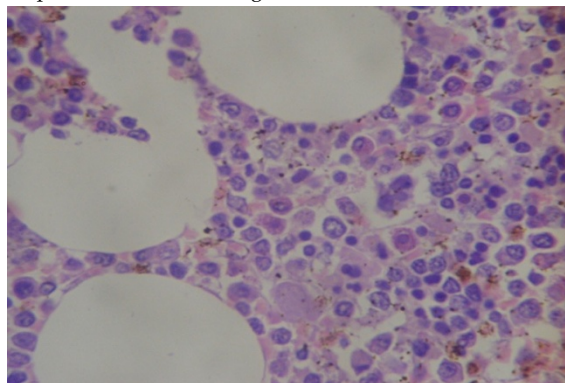


Figure 4: A high power view (H&EX 400) of the same area, showing hematopoietic cells of all lineages.

Discussion

Myelolipoma was initially described by Gierke in 1905 and subsequently termed as "formations myelolipomatoses" by Oberling in 1929.⁴ Over the years, several hypotheses have been proposed about the causative process of these rare lesions but it is still not validated. Recent experiments suggest that both the lipomatous and myeloid elements are monoclonal in origin, suggesting that myelolipomas are neoplastic lesions.⁸

These usually develop in adrenal glands. Extra-adrenal myelolipomas may occur in retroperitoneum, pelvis, presacral area, thorax, mediastinum, stomach, liver, kidney and even in the thyroid glands.³ Their prevalence appears to be increasing upto 10% due to increased use of radiographic imaging.⁹

These are usually asymptomatic¹ but depending upon the location and size these can cause pain in that region. Mechanical compression by large tumors, tumor necrosis or spontaneous retroperitoneal hemorrhage can be the reasons for painful sensations.¹⁰ Kenney et al. showed that the risk of hemorrhage was over four times greater for tumors >10 cm in diameter.¹¹

Retroperitoneal extra-adrenal myelolipomas must be distinguished from other adipose tissue containing tumors. These include both benign and malignant lesions such as angiomyolipoma, retroperitoneal liposarcoma, extramedullary hematopoiesis, and pelvic lipomatosis.¹²

Fortunately, the various tumors in the differential diagnosis can be distinguished microscopically. But the distinction between extra-adrenal myelolipoma and extramedullary hematopoiesis can be very challenging.¹²

Notable clinical, anatomical and pathological differences exist between these two entities as indicated by Fowler et al in his report. Clinically, the patients with extramedullary hematopoietic tumor present with hepatosplenomegaly, anemia and bone marrow hyperplasia, while patients with extra-adrenal myelolipoma lack these signs. The extra-adrenal myelolipomas are usually solitary and lie in the abdominal cavity, whereas the extramedullary hematopoietic tumors are multiple and located retropleurally lateral to the

vertebral column. Extramedullary hematopoietic tumor shows bony spicules and connection to medullary bone cavity while extra-adrenal myelolipoma lacks these features. Grossly the extra-adrenal myelolipoma shows yellow fatty areas because of predominance of adipose tissue on cut section, while extramedullary hematopoietic tumor lacks this appearance. Microscopically, if we note the myeloid to erythroid ratio, extra-adrenal myelolipoma shows mainly lymphocyte aggregates while erythroid hyperplasia is seen in extramedullary hematopoietic tumor.¹³

The myelolipomas can be managed as follows:

1. Close follow up with regular radiographic imaging for small asymptomatic tumors.
2. Surgical excision for large sized, symptomatic or asymptomatic tumors, to prevent the risk of spontaneous hemorrhaging¹⁰.

Conclusion

The purpose of this case report is to make the clinicians aware of the atypical presentation and location of a myelolipoma. Pre-operative diagnosis of extra-adrenal myelolipoma is at times difficult on radiographic imaging alone. Histopathology is the gold standard for definitive diagnosis so CT-guided fine needle aspiration cytology or tissue biopsy can be done preoperatively to confirm the diagnosis. In this atypical case of extra-adrenal myelolipoma, surgical resection of the mass was successfully done to relieve the patient's symptoms and to confirm the diagnosis.

References

1. Conley A, Klein E, E dhayan E, Berri R. Extra-adrenal myelolipoma presenting as efferent limb obstruction. *Case Rep Surg.* 2012;2012:718383.
2. Wani NA, Kosar T, Rawa IA, Qayum A. Giant adrenal myelolipoma: Incidentaloma with a rare incidental association. *Urol Ann.* 2010;2(3):130-33.
3. 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 Apr 4;7(1):92.
4. 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.

5. Brogna A, Scalisi G, Ferrara R, and Bucceri A. M. Giant secreting adrenal myelolipoma in a man: a case report. *Journal of Medical Case Reports*, vol. 5, article 298, 2011.
6. Temizoz O, Genchellac H, Demir MK, Unlu E, Ozdemir H. Bilateral extra-adrenal perirenal myelolipomas: CT features. *Br J Radiol*. 2010 Oct;83(994):e198-9.
7. Orlicz-Szcześna G, Prystupa A, Miturski A, Trojnar M. Large adrenal myelolipoma - case report. *Journal of Pre-Clinical and Clinical Research*, 2011, Vol 5, No 1, 41-42.
8. McNicol AM. A diagnostic approach to adrenal cortical lesions. *Endocr Pathol*. 2008 Winter;19(4):241-51.
9. Porcaro A. B, Novella G, Ficarra V, Cavalleri S, Antoniolli S. Z, and Curti P. Incidentally discovered adrenal myelolipoma. Report on 3 operated patients and update of the literature. *Archivio Italiano di Urologia, Andrologia*, vol. 74, no.3, pp. 146-151, 2002.
10. Shih H-J, Chang H-K. A Huge Adrenal Myelolipoma: A Case Report. *JTUA* 18:102-104, 2007.
11. Kenney PJ, Wagner BJ, Rao P, Heffess CS. Myelolipoma: CT and pathologic features. *Radiology* 1998;208:87-95.
12. Clark P.E, Farver C.F, Ulchaker J.C, Angermeier K. A rare case of an extra-adrenal myelolipoma arising in the renal sinus: a case report and review of the literature. *ScientificWorldJournal*. 2005 Jan 28;5:109-17.
13. Fowler, M.R., Williams, R.B., Alba, J.M., and Byrd, C.R. (1982) Extra-adrenal myelolipomas compared with extramedullary hematopoietic tumors: a case of presacral myelolipoma. *Am. J. Surg. Pathol*. 6, 363-74.