

A case report of symptomatic presacral myelolipoma

Min Ho Cho, MD^{a,*}, Rohan Mandaliya, MD^b, John Liang, MD^c, Mitesh Patel, MD^b

Abstract

Rationale: Extra-adrenal myelolipoma in the presacral area often raises a concern for liposarcoma because they have similar radiologic features.

Patient concerns: A 70-year-old woman with multiple abdominal surgeries in the past presented with persistent lower abdominal pain and anemia. A presacral mass, found on a pelvic magnetic resonance image (MRI), was suspicious of liposarcoma, as it is the most common fat containing mass in the presacral area. It is often difficult to make a diagnosis of myelolipoma just based on the radiological studies, which necessitates a histopathologic examination of the resected mass for a definitive diagnosis.

Diagnoses: Myelolipoma is an encapsulated, benign tumor containing mature adipocytes and hematologic cells. Most of the patients with myelolipoma remain asymptomatic but continued growth of tumor without local invasion can lead to compression of nearby structures, causing persistent pain.

Interventions: Asymptomatic patients do not require intervention but surgical resection is indicated for persistent pain or hemorrhage inside the tumor. Our patient had 2 indications for surgery – persistent pain as well as a provisional diagnosis of liposarcoma. After the surgical resection, pelvic pain was resolved, and a diagnosis of myelolipoma was made based on histopathologic examination.

Outcomes: Patient was reassured that it was myelolipoma, a benign tumor, not requiring subsequent surveillance for recurrence.

Lessons: Despite advancement in imaging techniques, and knowledge of the radiological features of myelolipoma, it still remains as a challenge for clinicians to make the distinction between liposarcoma and myelolipoma only based on diagnostic radiology. Although myelolipoma is a benign tumor, if patient suffers from persistent pain due to local mass effect, surgical resection is required.

Abbreviations: CT = computer tomography, IV = intravenous, MRI = magnetic resonance image.

Keywords: extra-adrenal myelolipoma, pelvic mass, presacral mass

1. Introduction

Myelolipoma is a rare disease entity. It is a benign tumor with mature adipocytes and mature hematopoietic cells.^[1] It can be classified into 4 subgroups, based on their clinicopathological features: isolated adrenal myelolipoma, adrenal myelolipoma with acute hemorrhage, extra-adrenal myelolipoma, and myelolipoma associated with other adrenal disease.^[2,3]

Isolated adrenal myelolipoma is the most common type, and it is most frequently, found as an isolated adrenal incidentaloma on imaging studies. Patients are usually, asymptomatic, since most of

the tumors are non-functioning. However, when its size grows, it can have central hemorrhage as a complication. There were multiple case reports of acute massive retroperitoneal hemorrhage due to myelolipoma.^[4]

Although adrenal myelolipoma is the most common form, myelolipoma can occur in any part of the body. There are 77 English literature results in PubMed when searched for “extra-adrenal myelolipoma,” describing occurrences in various parts of the body but most commonly, presacral, retroperitoneal, and mediastinal areas.^[5] There have also been multiple cases of myelolipoma associated with underlying adrenal disease for example, in association with Cushing’s syndrome, 21-hydroxylase deficiency, and non-hyperfunctioning adrenal adenoma.^[2,3]

With advancement in imaging techniques, the incidence of myelolipoma has risen slightly, but its diagnosis, and management remain challenging. It is often misdiagnosed as a liposarcoma based on imaging because liposarcoma is the most common fat-containing presacral mass. However, unlike myelolipoma, liposarcoma is malignant, and thus, more aggressive, requiring immediate surgical intervention. Whether to obtain a core-needle biopsy, or an excisional biopsy remains controversial as core-needle biopsy can cause seeding of the tumor in the needle tract while excisional biopsy is a lot more invasive, requiring general anesthesia.^[6] Here, we present a case of symptomatic presacral myelolipoma, which was initially misdiagnosed as liposarcoma.

1.1. Presenting concerns

A 70 year-old African American woman presented with ongoing pelvic pain for a few months. She did not have any other

Editor: N/A.

Informed Consent. Ensure that the patient provided their informed consent for the publication of this case.

The authors have no funding and conflicts of interest to disclose.

^a Department of Medicine, MedStar Washington Hospital Center, ^b Division of Gastroenterology and Hepatology, MedStar Georgetown University Hospital, ^c Department of Pathology, MedStar Washington Hospital Center, Washington, DC.

* Correspondence: Min Ho Cho, Department of Medicine, MedStar Washington Hospital Center, Washington, DC 20010 (e-mail: Minho.cho86@gmail.com).

Copyright © 2018 the Author(s). Published by Wolters Kluwer Health, Inc. This is an open access article distributed under the terms of the Creative Commons Attribution-Non Commercial License 4.0 (CCBY-NC), where it is permissible to download, share, remix, transform, and buildup the work provided it is properly cited. The work cannot be used commercially without permission from the journal.

Medicine (2018) 97:15(e0337)

Received: 21 July 2017 / Received in final form: 13 February 2018 / Accepted: 14 March 2018

<http://dx.doi.org/10.1097/MD.00000000000010337>

associated symptoms such as fever, dysuria, vaginal bleeding, diarrhea or rectal bleeding.

1.2. Clinical findings

The patient’s comorbidities included hypertension, chronic obstructive lung disease, chronic kidney disease, and anemia. She also had multiple abdominal surgeries, including partial colectomy, and total abdominal hysterectomy with bilateral salpingo oophorectomy. Physical examination was unremarkable other than abdominal tenderness in the lower quadrants.

1.3. Timeline

Date	Relevant past medical history and interventions		
July 22, 2016	A 70 year-old African American woman presented with ongoing pelvic pain for a few months without any other associated symptoms such as fever, dysuria, vaginal bleeding, diarrhea, or rectal bleeding. She also reported that she was recently found to have a “pelvic mass” at another hospital.		
Date	Summaries from initial and follow-up visits	Diagnostic testing (including dates)	Interventions
July 22, 2016	No pelvic mass identified. Pelvic ascites present	Ultrasound of pelvis and transvaginal	Clinical monitoring and further investigation
July 23, 2016	Dilated common bile duct and small ascites.	Ultrasound of abdomen	Clinical monitoring and further investigation
July 25, 2016	Magnetic resonance image (MRI) showing the pelvic mass. Patient left the hospital prior to being evaluated by surgical team.	MRI pelvis with and without contrast	GI consult to rule out GI malignancy.
November 18, 2016	Delay in colonoscopy due to loss to follow up. Diverticulosis in colon. Non-bleeding hemorrhoids. No further intervention	Colonoscopy	Loss to follow up
January 01, 2017	Patient came back with persistent abdominal and pelvic pain. Staging computer tomography (CT) did not show any evidence of metastasis.	CT chest, abdomen, pelvis with intravenous (IV) contrast	Surgical planning
January 18, 2017	Came for elective surgery	Not applicable	Resection of the pelvic mass

CT = computer tomography, GI = gastrointestinal, IV = intravenous MRI = magnetic resonance image

1.4. Diagnostic focus and assessment

The patient did have chronic normocytic anemia (mean corpuscular volume was 92 to 96; hemoglobin was 8.6 gm/dL; and hematocrit was 25.3%). Her other routine laboratory values

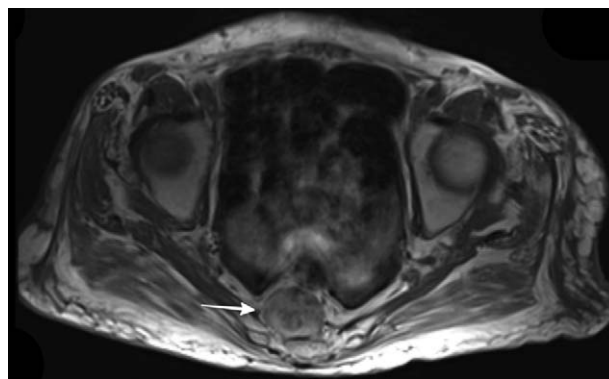


Figure 1. T1 sequence of MRI abdomen, and pelvis with, and without MultiHance. A focal mass which measures 3.5cm transverse × 3cm anteroposterior × 3.6cm craniocaudally within a presacral soft tissue. MRI = magnetic resonance image.

including white blood cell counts and platelet counts were normal as was basic metabolic profile. A transvaginal ultrasound showed pelvic ascites without a visible mass. MRI of abdomen, and pelvis with, and without gadobenate (MultiHance) showed a focal mass which measures 3.5cm transverse × 3cm anteroposterior × 3.6cm craniocaudally within a presacral soft tissue (Fig. 1). The mass had heterogeneous high T2 signal; internal high T1 signal; and heterogeneous T1 signal. Heterogeneous T2 area is suggestive of hematopoietic cells in the tumor while high T1 signifies lipid portion of myelolipoma. There was no invasion to the surrounding structures although there was mild remodeling. Based on the MRI findings, differential diagnoses included liposarcoma, germ cell tumor, neurogenic tumor, and extramedullary hematopoiesis. Colonoscopy was normal and ruled out colonic malignancy associated with the mass. CT chest, abdomen, and pelvis with oral, and IV contrast for staging purpose showed a stable fat-containing well-circumscribed presacral mass measuring 4.2 × 3.9 × 3.0cm (Fig. 2). There was no evidence of local invasion or metastatic lesion.



Figure 2. CT abdomen and pelvis with contrast. A fat-containing well-circumscribed presacral mass measuring 4.2 × 3.9 × 3.0cm. CT = computer tomography.

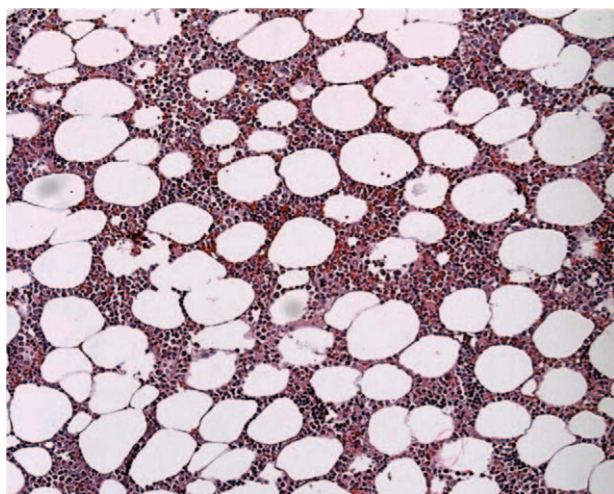


Figure 3. Hematoxylin and eosin stain of the resected tissue (medium magnification, $\times 100$). A medium magnification image showing hematopoietic cells interspersed between the adipocytes.

1.5. Therapeutic focus and assessment

The patient underwent surgical resection of the mass without complication. Although not included in this article, the resection was complete, and margins were clear without evidence of local invasion. The histopathological examination of the resected mass showed numerous adipocytes with hematopoietic cells interspersed in between the adipocytes on hematoxylin and eosin stain under $\times 100$ magnification (Fig. 3). Under higher magnification ($\times 400$), individual hematopoietic element, including erythrocytes, megakaryocytes, and granulocytes, is well visualized (Fig. 4). There is no dominance in 1 particular lineage. Hematopoietic components appeared mature although immunohistochemistry was not performed. Overall, the histology of the sample resembled a bone marrow biopsy without bone trabeculae. Lipoblasts, and stromal cells, typically seen in liposarcoma, were also absent.

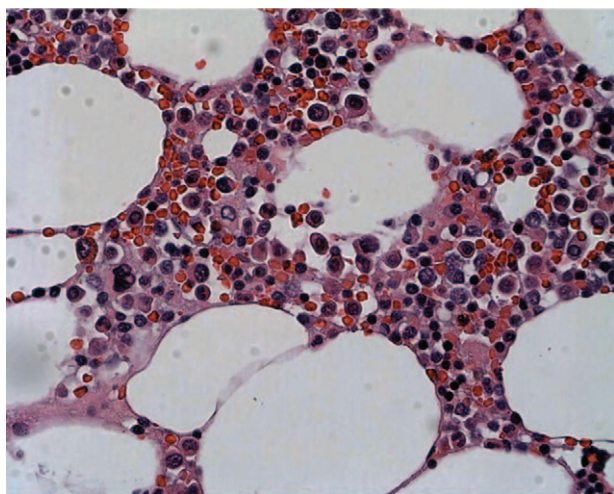


Figure 4. Hematoxylin and eosin stain of the resected tissue (high magnification, $\times 400$). A high magnification image showing adipocytes with surrounding numerous mature trilineage hematopoietic cells, including erythrocytes, megakaryocytes, and granulocytes.

1.6. Follow-up and outcomes

There was no need for follow-up as the mass was a benign tumor on histopathology and patient's abdominal pain resolved after the surgery.

2. Discussion

Myelolipoma is most often found as an adrenal incidentaloma on imaging. There have been about 50 case reports, and literature reviews. It has a woman predominance with 2:1 ratio, and occurs among middle-aged to elderly population with a mean age of 60. Various parts of the body have been involved: presacral, retroperitoneum, mediastinum, liver, stomach, lungs, pelvis, and spleen.^[7]

Most patients with myelolipoma are asymptomatic, as the tumor is non-functional. However, with continued growth, it can lead to local mass effect, compressing surrounding structures. Rarely, it can also adhere to a nearby structure, making surgical resection difficult. When the mass enlarges, it can also result in central hemorrhage.

Different imaging modalities have been used in the past to characterize myelolipoma. Such modalities include ultrasound, CT, and MRI. On ultrasound, myelolipoma has various appearances, depending on the proportion of fat and hematopoietic elements – fatty parts will appear as hyperechoic areas while hematopoietic cells appear as hypoechoic areas. On CT, myelolipoma presents as an encapsulated mass with a macroscopic fatty tissue admixed with a soft tissue, representing hematopoietic elements. Also, hematopoietic parts may enhance after IV contrast injection. MRI is the best modality to characterize myelolipoma. Fatty area has high signal intensity on T1-weighted images which can be confirmed on fat suppression sequence while hematopoietic parts appear as lower signal intensity on T1-weighted and intermediate signal intensity on T2-weighted images.^[8]

Despite advancement in imaging techniques, and knowledge of the radiological features of myelolipoma, it still remains as a challenge for clinicians to make the distinction between liposarcoma and myelolipoma only based on diagnostic radiology.^[9,10] However, it is very important to make a correct diagnosis as the prognosis, and management for myelolipoma are very different from liposarcoma, as the former is benign while the latter is malignant. Most patients with extra-adrenal myelolipoma are asymptomatic, and need serial assessment with imaging for monitoring without surgical intervention.

Differential diagnoses for a fat-containing presacral mass includes liposarcoma, extramedullary hematopoiesis, and germ cell tumor. These disease entities have distinctive pathophysiology as well as different features on imaging.

Liposarcoma is the most common retroperitoneal fat-containing mass.^[11] It is malignant and more aggressive in nature. It appears as more infiltrative growth with less well-defined borders.^[4] Because, liposarcoma lacks encapsulation, it often involves local invasion. Histology often shows lipoblasts and stromal cells. Management involves surgical radical resection.

Extramedullary hematopoiesis also contains fat although usually at a microscopic level. The predilection site is mediastinum but it can infrequently occur in a presacral area. It is usually multifocal, and poorly circumscribed.^[5] Also, it is more commonly, seen among young to middle-aged men in association with myeloproliferative disorders, and chronic hemolytic anemia.

Germ cell tumor is another fat-containing mass that can occur in the pelvic region. Germ cell tumors appear as well-circumscribed heterogeneous masses on image,^[5] because they contain fat, soft tissue, and calcification. Calcification is often a distinctive feature of teratoma although it can also be seen in myelolipoma following hemorrhage.

Our patient carried a misdiagnosis of liposarcoma from another facility. The first MRI of abdomen and pelvis showed a well-circumscribed fat-containing mass, with a provisional diagnosis of liposarcoma again. Surgical resection relieved her pain, and provided a definitive tissue diagnosis of myelolipoma.

Author contributions

Data curation: John Liang.

Resources: John Liang.

Supervision: John Liang, Mitesh Patel, Rohan Mandaliya.

Writing – original draft: Min Ho Cho.

Writing – review & editing: Mitesh Patel, Rohan Mandaliya, Min Ho Cho.

References

- [1] Plaut A. Myelolipoma in the adrenal cortex (myeloidipose structures). *Am J Pathol* 1958;34:487–515.
- [2] Rao P, Kenney PJ, Wagner BJ, et al. Imaging and pathologic features of myelolipoma. *Radiographics* 1997;17:1373–85.
- [3] Oliva A, Duarte B, Hammadeh R, et al. Myelolipoma and endocrine dysfunction. *Surgery* 1988;103:711–5.
- [4] Fuss IJ, Friend J, Yang Z, et al. Nodular regenerative hyperplasia in common variable immunodeficiency. *J Clin Immunol* 2013;33:748–58.
- [5] Baker KS, Lee D, Huang M, et al. Presacral myelolipoma: a case report and review of imaging findings. *J Radiol Case Rep* 2012;6:1–9.
- [6] Hassan I, Wietfeldt ED. Presacral tumors: diagnosis and management. *Clin Colon Rectal Surg* 2009;22:84–93.
- [7] Sutker B, Balthazar EJ, Fazzini E. Presacral myelolipoma: CT findings. *J Comput Assist Tomogr* 1985;9:1128–30.
- [8] Gagliardo C, Falanga G, Sutura R, et al. Presacral myelolipoma. a case report and literature review. *Neuroradiol J* 2014;27:764–9.
- [9] Itani M, Wasnik AP, Platt JF. Radiologic-pathologic correlation in extra-adrenal myelolipoma. *Abdom Imaging* 2014;39:394–7.
- [10] Tokuyama N, Takeuchi H, Kuroda I, et al. Incidental presacral myelolipoma resembling the liposarcoma: a case report and literature review. *Case Rep Urol* 2016;2016:6510930.
- [11] Craig WD, Fanburg-Smith JC, Henry LR, et al. Fat-containing lesions of the retroperitoneum: radiologic-pathologic correlation. *Radiographics* 2009;29:261–90.