Adrenal myelolipoma: To operate or not? A case report and review of the literature

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A B S T R A C T

INTRODUCTION: Myelolipoma is a rare, benign neoplasm that predominantly occurs in the adrenal gland and is composed of mature adipose tissue and scattered islands of hematopoietic elements. Although usually small and asymptomatic, there are some cases of adrenal myelolipoma that cause symptoms such as chronic pain. Elective surgery can prevent a more severe symptom presentation and life threatening progression while also allowing accurate diagnosis in patients with tumors larger than 6 cm.

PRESENTATION OF CASE: This report presents an unusual case of a 28-year-old female who suffered with chronic pain from a growing left-sided adrenal myelolipoma. Without the financial means for additional testing along with many symptoms warranting a high suspicion for malignancy, this patient decided to have the mass surgically excised, which was both diagnostic and curative.

DISCUSSION: For this specific patient, surgery was the most cost effective option, as well as both diagnostic and curative. Surgery can also prevent complications such as spontaneous rupture and hemorrhage of the mass, which can lead to subsequent cardiovascular shock.

CONCLUSION: Management of adrenal myelolipoma should be considered on an individual basis. Although it is a benign tumor, surgery plays an important role for symptomatic cases and those lesions that cannot be distinguished reliably from malignancy.

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1. Introduction

Myelolipoma is a rare, non-functional, benign neoplasm that predominantly occurs in the adrenal gland and is composed of macroscopic fat and mature hematopoietic tissue, resembling bone marrow. 1 Less than 300 cases were reported in the literature before 2000.2 In the past, these tumors were discovered at autopsy, with an incidence ranging from .08% to .4%. Today, with the widespread use of radiological studies such as ultrasonography, computed tomography (CT), and magnetic resonance imaging (MRI), the incidental detection of myelolipoma has become more common, constituting up to 10–15% of incidental adrenal masses.3

The tumor affects men and women equally and is most commonly found between the fifth and seventh decades of life, with a mean age of 62 years.2,4 They are generally non-secreting in nature and are often smaller than 4 cm in diameter, with the largest reported adrenal myelolipoma measuring 31 cm × 24.5 cm × 11.5 cm and weighing 6 kg.4

While myelolipomas are mostly incidental findings, they may become symptomatic causing flank pain and abdominal discomfort by causing pressure of surrounding structures and may even present with necrosis, rupture, hemorrhage, or hemorrhagic shock.4 Management of adrenal myelolipoma is usually conservative because these tumors are typically asymptomatic and rarely show signs of spontaneous hemorrhage if the tumor is small. Some studies suggest surgical intervention if the tumor is symptomatic, growing, or larger than 6 cm.2 Ultimately, the optimal treatment for myelolipoma depends on the size and symptoms of the mass, and the needs of the patient.

2. Case report

A 28-year-old Hispanic female presented to the surgery oncology clinic with chronic left sided flank pain that had been worsening over the past 2–3 days. 2 years prior she had presented to the Emergency Department with flank pain due to a non-obstructing kidney stone and the CT had shown a well-circumscribed incidental adenoma of the left adrenal gland measuring 4.70 cm × 3.77 cm. After 6 months of chronic back and flank pain, re-imaging showed the adenoma had increased in size, measuring 5.3 cm × 6.6 cm. On CT there was a loss of fat plane between the adenoma, spleen, and pancreas (Picture 1). Initial labs showed potassium of 3.4 mEq/L.
Her past medical history is significant for anxiety, carpal tunnel syndrome, ulnar nerve palsy of the right hand, esophageal reflux, and muscle spasm. Her past surgical history involved a laparoscopic cholecystectomy.

3. Physical exam

On presentation the vital signs were blood pressure 139/80, heart rate 83, and temperature 99.2 Fahrenheit. The patient appeared ill, nauseated, tearful and anxious. She is morbidly obese with a BMI of 46.6. She has a cushingoid appearance, with a moon faces, truncal obesity, and abdominal striae present. Posterior cervical, anterior cervical, supraclavicular and axillary nodes were not palpable. The abdomen was tender to deep palpation in the epigastric region and the left upper quadrant, with no rebound tenderness or guarding. Costovertebral angle tenderness was mild.

4. Treatment and plan

The patient had labs done to test for a pheochromocytoma displaying the following:

- Normetanephrine: 0.20 nmol/L (reference range 0.00–0.89 nmol/L).
- Metanephrine: 0.11 nmol/L (reference range 0.00–0.49 nmol/L).
- VMA 24 h urine: normal.
- VMA/creatinine ratio: 2 mg/g (reference range 0–6).

High resolution CT and MRI as well as labs to rule out hypercortisolism and functional adrenocortical carcinoma were refused by the patient and her mother due to lack of finances.

Given the fact that she had worsening pain along with a rapidly growing adenoma, the patient opted for surgical resection of the mass. Due to the patient’s morbid obesity (BMI 46.6), suspected local invasion of the spleen and pancreas (Picture 1), and the high clinical suspicion of malignancy, an exploratory laparotomy was performed. It was discovered that the tumor was adherent to the perinephric fat, spleen, and distal tail of the pancreas. The adrenal mass had to be separated from the perinephric fat and then the tumor, adrenal gland, spleen and a small portion of the distal tail of the pancreas were removed en bloc. The patient’s blood pressure remained stable throughout the surgery, which was along expected lines given the fact that pheochromocytoma had been ruled out. A drain was placed in the left upper quadrant and the skin was closed with staples.

Eight days following the surgery the patient was ambulating without assistance, tolerating a normal diet, and she was

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**Picture 1.** (A) CT scan showing loss of a fat plane between the mass and the pancreas (yellow arrow). (B) CT scan showing local invasion of the spleen (red arrow). **Picture 2.** (A) Histopathologic picture of myelolipoma composed of mature fat cells mixed with hematopoietic elements. (B) Magnified view of (A).
discharged home. Two weeks following the surgery the patient had no significant complaints and reported that her left upper quadrant and flank pain had since subsided.

5. Pathology report

The adrenal mass consisted of a 93.84 g, 7.5 cm × 6.0 cm × 4.0 cm myelolipoma (Picture 2). The capsule was red-brown and smooth with areas of tan-yellow discoloration. The capsule weighed 230 g and measured 15.4 cm × 9.3 cm × 5.4 cm, displaying passive congestion and mild sinusoidal dilatation. The distal pancreas weighed 60 g and measured 6.0 cm × 3.4 cm × 1.9 cm, consisting of normal pancreatic tissue. No evidence of malignancy was found in either the spleen or the pancreas.

6. Discussion and conclusion

This case has an unusual presentation due to the fact that the patient was in the third decade of life (28 years old), was symptomatic, and the dimensions of the adrenal myelolipoma (AML) were larger than 4 cm. Also, the patient had the clinical signs of a hormone-secreting tumor (truncal obesity, abdominal striae, hypertension, and anxiety), although labs ruled out pheochromocytoma. This patient’s clinical signs and symptoms along with the CT imaging warranted concern for malignancy such as liposarcoma, a fat containing adenocortical carcinoma (ACC), or adrenal teratoma. Adrenal liposarcomas are typically large at presentation, with the average fat content ranging from 25% to a little over 50%. ACC’s are more common in women, can secrete cortisol, aldosterone, and sex hormones, are typically large and ill-defined, and preferentially affect the left adrenal gland. Her adrenal mass was located on the left side and had ill-defined borders near the pancreas and spleen, warranting suspicion for local invasion.

Although specific MRI techniques such as frequency-selective fat suppression could have been done to diagnose AML, it was not done because regardless of the findings, surgery was needed to alleviate her symptoms. Also, the patient did not have the financial means for additional testing and this would not have been cost effective for her treatment. While the laparoscopic approach is gaining ground for management of AML, this procedure is not indicated when there are adhesions or if the lesion shows infiltration of the surrounding structures, both of which were seen in this case (Picture 1).  

Although traditionally treated conservatively, some studies suggest surgical intervention for symptomatic tumors, growing tumors, or tumors larger than 6 cm in order to reduce the risk of developing abdominal pain or life threatening rupture and hemorrhage. After excision, AML generally does not recur, with recurrence-free survival rates of up to 12 years being reported. While the tumor is benign, surgery has an important role for symptomatic cases and those lesions that cannot be distinguished reliably, and treatment and management should be done on a case-by-case basis.

Conflict of interest statement

There are no potential sources of conflict declared by any of the authors.

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Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contributions

Mary Ramirez: first author, data collection and Subhasis Misra: corresponding author.

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References

1. Feng C, Jiang H, Ding Q, W en H. Adrenal myelolipoma: a mingle of progenitor cells? Med Hypotheses 2013;80:819–22.
2. Nabi J, Rafiq G, Authoy F, Sofi GN. Incidental detection of adrenal myelolipoma: a case report and review of literature. Case Rep Urol 2013;2013:1–3.
3. Wani N, Kosar T, Raw a I, Qayum A. Giant adrenal myelolipoma: incidentaloma with a rare incidental association. Urol Ann 2010;2:3:130 [print].
4. D aneshmand S, Quek ML. Adrenal myelolipoma: diagnosis and management. J Urol 2006;171:71–4.
5. Vajtai Z, Korngold E, Hooper JE, Sheppard BC, Foster BR, Coakley VF. Suprarenal retroperitoneal liposarcoma with intracaval tumor thrombus: an imaging mimic of adenocortical carcinoma. Clin Imag 2014;38:1:75–7 [print].
6. Low G, Dhilwayo H, Lomas DJ. Adrenal neoplasms. Clin Radiol 2012;67:988–1000 [print].
7. Pereira JM, Sirin CB, Pinto PS, Casola G, CT and MR imaging of extrahepatic fatty masses of the abdomen and pelvis: techniques, diagnosis, differential diagnosis, and pitfalls. Radiographics 2005 [print].
8. Takahashi K, Honda M, Okubo RS, Hyodo H, Takakuski H, Yokoyama H, et al. CT pixel mapping in the diagnosis of small angiomylipoma of the kidneys. J Comput Assist Tomogr 1993;17:98–101.
9. Ioannidis O, Papaemmanouil S, Chatzopoulos S, Paraskevas G, Konstantara A, Kotronis A, et al. Giant bilateral symptomatic adrenal myelolipomas associated with congenital adrenal hyperplasia. Pathol Oncol Res 2011 [Epub ahead of print].
10. Hsu S-W, Shu K, Lee W-C, Cheng Y-T, Chiang P-H. Adrenal myelolipoma: a 10 year single-center experience and literature review. Kaohsiung J Med Sci 2012;37:77–82 [print].

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