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 TYPE OF ARTICLE: Case Report

TITLE: Hepatic angiomyolipoma presenting with chronic epigastric pain: A case report and review of literature

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ABSTRACT

Introduction
Angiomyolipomas primarily arising from the liver are rare. Hepatic angiomyolipoma (HAML) is a rare tumor characterized by the presence of blood vessels, muscle tissue and adipose tissue with only about 300 reported cases to date. This the first case reported in the literature on a Filipino patient.

Case report
A 49-year old Filipino female presented with a 2-year history of recurrent epigastric pain. Abdominal magnetic resonance imaging revealed a fat-containing lesion in the left liver lobe. Serial follow up revealed progressive enlargement on imaging studies. Serologic tests were negative for malignancy or infection. The patient underwent left hepatic lateral segmentectomy which revealed a 7.5 x 7 cm friable mass involving hepatic segments II and III. Histopathology showed mature fat cells with a few thick-walled blood vessels and spindled smooth muscle cells with no atypia. Homatropine mehtylbromide-45 test showed strong and diffused staining confirming angiomyolipoma.

Conclusion
Clinical evaluation and a comprehensive analysis of US, CT, MRI imaging characteristics are essential for correct preoperative diagnosis. Symptoms, malignant potential and inability to distinguish from liver malignancies may indicate surgical resection as the best treatment option.

Keywords: Angiomyolipoma, Hepatic tumors
INTRODUCTION

Hepatic angiomyolipoma (HAML) is a rare tumor characterized by the presence of blood vessels, muscle tissue and adipose tissue. It was first described by Ishak in 1976 and was classified under the group of perivascular epitheloid tumors in the World Health Organization classification of tumors in 2002 [1,2]. Approximately 300 reported cases have been reported to date and to the best of our knowledge, this is the first reported case of HAML in a Filipino patient.

HAML is difficult to diagnose because of its rarity and varying composition of adipose and muscle tissue. Tsui et al categorized the disease according to its composition: mixed, lipomatous (>70% fat), myomatous (<10% fat) and angiomatous types [3]. These varying classifications resembles other hepatic tumors, and makes preoperative diagnosis only 11% to 50% accurate, where majority are misdiagnosed as hepatocellular carcinoma (HCC) [3-5].

While HAML is considered a benign tumor, several malignancies and recurrences have been reported. Predisposing characteristics of its invasive nature have been unclear. It is important to review the characteristics that improve accuracy in the diagnosis of HAML as well as the features that might predispose to invasiveness.

CASE REPORT

A 49-year old Filipino female presented with a 2-year history of liver mass with recurrent epigastric pain. She was being managed as a case of gastritis with a benign liver mass for 2 years until she consulted at our institution for increasing severity and recurrence of epigastric pain. There was no abdominal tenderness no palpable mass noted on physical examination. Abdominal magnetic resonance imaging (MRI) with intravenous gadolinium contrast revealed a fat-containing lobulated lesion in the left liver lobe with increasing size measuring 7.12 x 6.2 cm compared to 6.15 x 5.17 cm from the previous imaging study done 2 years prior (Figure 1). Serologic tests showed no indication of viral Hepatitis infection and normal Alpha fetoprotein and Carcino embryonic antigen.

The patient was presented with treatment options and eventually requested surgical resection. Routine preoperative testing revealed no significant co morbidities. Intraoperative fings confirmed the location of the mass and a lipomatous
appearance of the liver lesion (Figure 2). She underwent left hepatic lateral
segmentectomy with unremarkable perioperative course. The gross specimen
showed a 7.5 x 7 cm spongy, friable and fatty yellowish mass involving hepatic
segments II and III (Figure 3). Histopathology revealed a non-encapsulated mass
composed mainly of mature fat cells with a few scattered islands of thick-walled
blood vessels and spindled smooth muscle cells with no atypia (Figure 4).
Homatropine methylbromide-45 (HMB45) test showed strong and diffused staining of
the specimen which confirmed the diagnosis of angiomylipoma (Figure 5). After 1
year of follow-up, the patient did not show any recurrence or post-operative
complications.

DISCUSSION
Hepatic angiomylipoma is a benign mesenchymal tumor, often mistaken for
hepatocellular carcinoma (HCC) pre-operatively because of its similarity in its
diagnostic imaging characteristics. Improving knowledge of clinical features and
imaging characteristics will help decrease misdiagnosis and overtreatment.
A large case series of 33 HAML patients revealed that majority occur in women with
a peak incidence age range of 30-50 years, and with 66% of the cases coming from
Asian countries while 23% come from European countries [6]. Most patients are
asymptomatic and diagnosed incidentally. Symptomatic patients present with right
upper abdominal pain, abdominal distention, fever, discomfort and weight loss [4,7].
Although symptoms are not always correlated with size, gastrointestinal symptoms
as a result of intralesional bleeding are generally observed with tumors >5.0 cm [6,8].
Differences in imaging features of HAML and HCC are important to avoid
misdiagnosis and overtreatment. Zhu et al compared imaging findings of 14 cases
with existing literature and noted that HAML usually have an early draining vein in
the arterial phase and tortuous vessels in the tumor. These features are absent in
patients with HCC [9]. Furthermore, tumor capsule, liver cirrhosis, bile duct dilatation
and lymphadenopathy, which are usually present in HCC, are absent in HAML [9].
These distinguishing features significantly contributed in considering
angiomylipoma in our patient preoperatively.
Although HAML is considered a benign tumor, several cases have reported recurrence or malignancy. Dalle et al. reported the first case of HAML with malignant vessel invasion that recurred 5 months after surgical resection [10]. Definite factors predisposing to malignancy is still unknown. However, a tumor size of 10 cm or greater seems to have greater risk of recurrence [11]. Although the occurrence of malignancy is very rare among patients with HAML, it is still important to advise close follow-up particularly among patients with large tumors.

CONCLUSION
Hepatic angiomyolipoma is a rare and benign entity. It has a wide variation and poses a diagnostic challenge clinically, radiologically, and pathologically. Its prognosis is good regardless of management, including hepatic resection or conservative treatment after tissue biopsy by echo-guided needle. Reports of aggressive growth patterns, malignant degeneration, recurrences and difficulty in definitive diagnosis mandates individualized treatment planning. As in this case, surgery may be the most appropriate treatment for symptomatic and progressively enlarging HAML. This case report adds to the pool of information on hepatic angiomyolipoma and hopefully raises awareness of the disease among Filipino surgeons. Improving knowledge by continuously updating and comparing characteristics of angiomyolipoma cases with existing literature can provide better understanding, diagnosis and effective care for patients.

CONFLICT OF INTEREST
No conflict of interest or financial disclosures

AUTHOR’S CONTRIBUTIONS
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REFERENCES


FIGURE LEGENDS

Figure 1: Magnetic Resonance imaging with Gadolinium Contrast.

Figure 2: Intraoperative Findings

Figure 3: Resected Specimen

Figure 4: Histopathology showing adipose cells, muscle cells and blood vessel (HE x10).

Figure 5: HMB45 test showing strong and diffused staining (x40).

FIGURES

Figure 1: Magnetic Resonance imaging with Gadolinium Contrast.
Figure 2: Intraoperative Findings

Figure 3: Resected Specimen
Figure 4: Histopathology showing adipose cells, muscle cells and blood vessel (HE x10).
Figure 5: HMB45 test showing strong and diffused staining (x40).