

IMAGES in PAEDIATRIC CARDIOLOGY

Calleja Stafrace D, Vella C. Tuberous sclerosis associated with multiple hepatic lipomatous tumours. *Images Paediatr Cardiol* 2016;18(4):1-4.

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MeSH: Tuberous Sclerosis, hepatic multiple angiomyolipomas.

Introduction

Tuberous sclerosis is a genetic multisystem disorder characterised by widespread hamartomas in several organs, including brain, heart, skin, eyes, kidney, lung and liver. Hepatic multiple angiomyolipomas (AML's) are a rare and usually asymptomatic complication in patients with tuberous sclerosis.^{1,2} We present such a case.

Case presentation

We report a 15 year old girl with tuberous sclerosis who was noted to have multiple AML's of the liver and kidneys found on routine ultrasound screening. She was asymptomatic at this stage and in view of the small size of the AML's a repeat ultrasound of the liver and kidneys was performed twelve months later (Fig-1).

Figure 1- Routine ultrasound screening shows multiple AML's of the liver and kidneys



This showed a dramatic increase in the size of the AML's. The patient was asymptomatic and had normal liver biochemistry and clotting. MRI of the liver (Fig-2,3) showed signal dropout on opposed phase imaging and also on spectral fat suppression, in keeping with fat content of the AML's.

Fig-2 - MRI of the liver, in keeping with AML's

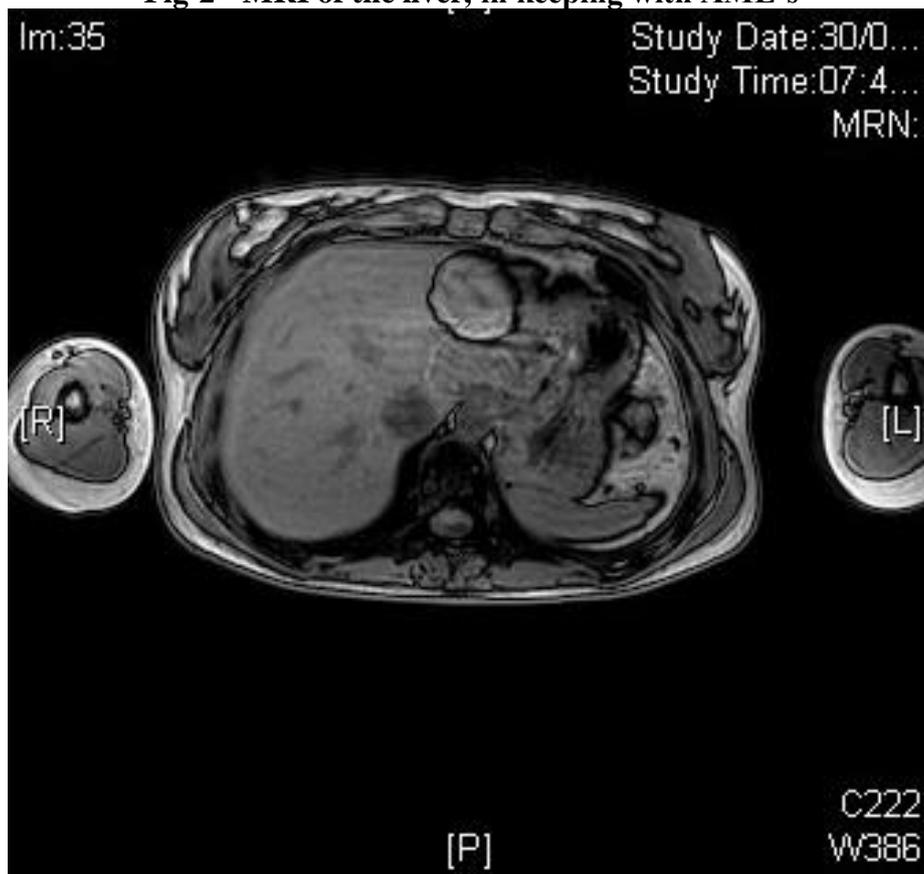


Fig-3 - MRI of the liver, in keeping with AML's



The lesions also enhanced avidly in the arterial phase with washout in the venous phases. These findings on MRI of the liver were typical of AML's.³ In view of the rapid progression in the growth of the lesions an ultrasound guided biopsy of the suspected AML's was performed despite the typical MR findings in order to exclude malignancy. Histological examination showed the blood vessel (angiod), smooth muscle (myoid) and mature fat (lipoid) components. No malignant components were identified.

Discussion

Angiomyolipoma (AML) is a benign, unencapsulated mesenchymal tumor that is composed of varying proportions of three elements: smooth muscle cells, thick-walled blood vessels, and mature adipose tissue.

A significant amount of literature with regard to hepatic AML comes from Asia where the condition is more common. Previously considered to be hamartomas, there is evidence for monoclonality and cytogenetic abnormalities suggesting a possible neoplastic aetiology. AML is more common in women and rare before puberty.

Liver AML are much rarer than renal AMLs and both are more likely to occur in patients with TS and in these patients the lesions can be multiple. From a clinical view point, hepatic AML's in tuberose sclerosis are commonly asymptomatic and most cases are detected incidentally on routine ultrasound screening.² This case was noted on routine U/S screening. Basic evaluation consist of U/S and CT /MRI which yield enough information in most cases to enable a reasonably certain diagnosis. Sometimes the picture less characteristic or the lesion appears to be growing rapidly raising the spectre of malignancy and justifying a biopsy procedure.

Radiologically, AML's can range from entirely lipomatous to completely solid lesions. A characteristic finding of hepatic involvement in this disease appears to be the multiplicity of the lesions. In the setting of tuberous sclerosis it is reasonable to assume that intrahepatic lesions represent AML's especially in the context of the typical MR findings, however in cases where the lesions increase rapidly in size a biopsy is needed to exclude malignancy.

Patients with tuberous sclerosis should be screened routinely for liver and kidney lesions and screening should be more frequent once AML's have been detected.

Management of Hepatic AML's may be conservative or interventional. Management is conservative follow up for most lesions. This is suggested in patients with asymptomatic AML's.⁵ Further criteria for conservative management include tumour size smaller than 5 cm, AML's confirmed on fine needle aspiration biopsy and patients with good compliance to follow-up.⁶ Surgical intervention is recommended when symptomatic, when diagnostic imaging cannot exclude malignancy, and when lesions increase rapidly in size.⁷ Malignant change has been reported rarely as has spontaneous rupture of the lesions. Hepatic AML's may be treated effectively with surgery and the prognosis is good.⁸ Embolisation of the lesions prior to surgery has been reported in the literature. Potential rupture of an angiomyolipoma is a life-threatening complication.

Although usually asymptomatic, occasionally AML's of the liver may result in hepatic insufficiency and very rarely require liver transplantation.⁹

Patient/Parental consent: Obtained

Conflicting interests: None

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