

MULTIFOCAL HAEMANGIOMATOSIS INCLUDING A GIANT HEPATIC HAEMANGIOMA WITH CAPSULAR RETRACTION AND KASABACHMERRITT'S SYNDROME

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PJR July - September 2013; 23(3): 114-117

ABSTRACT

Haemangiomas are the most common benign lesions of liver with female gender preponderance. These are usually silent and become symptomatic when larger in size (> 4 cm – giant haemangioma). Multifocality and capsular retraction on computerized tomography are rare features. We are presenting a case of multifocal haemangiomas with a giant hepatic lesion showing capsular retraction on CT images.

Key words: Giant haemangioma; multifocal haemangiomas; capsular retraction; Kasabach Merritt's syndrome

Haemangiomas are the most common benign neoplasm of liver. These are congenital lesions with mesenchymal origin although some consider them hamartomas. They are frequently diagnosed as an incidental finding on imaging and most patients are asymptomatic and less than 10% become symptomatic primarily due to their size.¹ Incidence of haemangiomas is 20% with a female preponderance and appreciable enlargement during pregnancy or during hormone replacement therapy suggesting a correlation with estrogen.² There is no reported age specific distribution but most commonly diagnosed in adults.³ Multiple haemangiomas of the liver can occur in about 10% of cases.⁴ There is poor agreement in the literature as to the exact definition of what constitutes giant hepatic haemangiomas.⁵ A giant haemangioma is considered when its size is more than 4 cm. Most of them remain stable with no symptoms over time. But some of them may become symptomatic and the most common symptom is abdominal pain or discomfort.¹ Goodman noted that symptoms are

experienced by 40% of patient with 4 cm haemangiomas and by 90% of patient with 10 cm haemangiomas.⁶

Most of the haemangiomas are diagnosed on ultrasound and computerized tomography (CT) but capsular retraction in haemangiomas may pose a diagnostic challenge as it is commonly associated with malignant neoplasms. We present a case of giant hepatic cavernous haemangioma with capsular retraction on CT and associated smaller lesions in spleen and thoracic vertebrae as well.

Case History

This is a 40 year old hypertensive male presented with short history of upper abdominal pain and vomiting. On examination he was hemodynamically stable but tender over right hypochondrium and epigastric region. His previous history was positive for an episode of abdominal pain with thrombocytopenia (platelet <80,000/ mm³). His recent

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Submitted 7 November 2014, Accepted 18 December 2014

laboratory tests revealed a platelet count of 280,000/mm³ while rest of hematological parameters, serum potassium, liver function tests, alpha fetoprotein and serum creatinine were normal. An upper abdomen ultrasound revealed a large echogenic lesion in the right hepatic lobe suggestive of a mass lesion. Subsequently a contrast enhanced CT examination was done which did show a vascular lesion 13/6 x 10.5 cm involving VI and VII hepatic segments with mild capsular retraction (Fig. 1). In addition, high density lesions were also found in spleen (1.1 cm) and T8 and T9 bodies. Findings were highly suggestive of multifocal haemangiomas with a giant one in liver. But the mild capsular retraction raised a possible suspicion of a malignant lesion. A planar

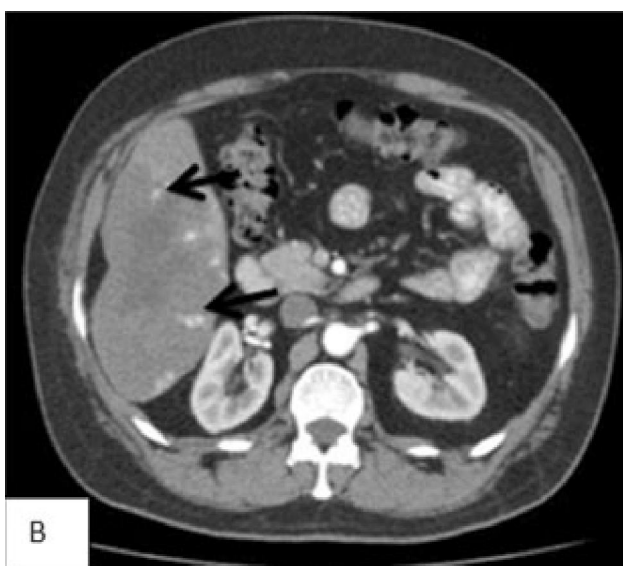
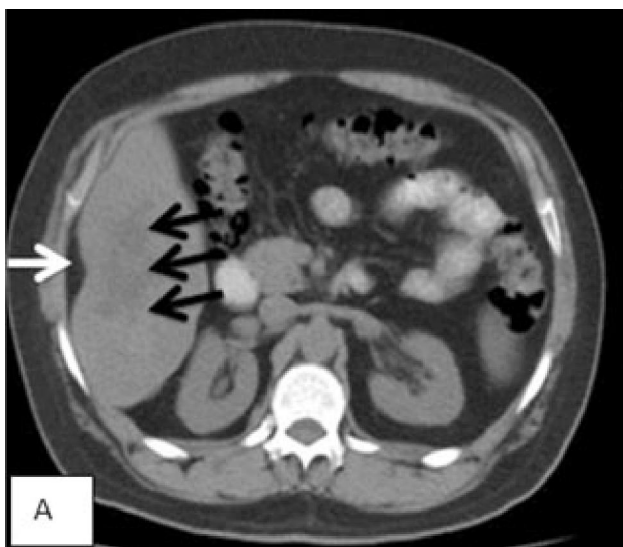
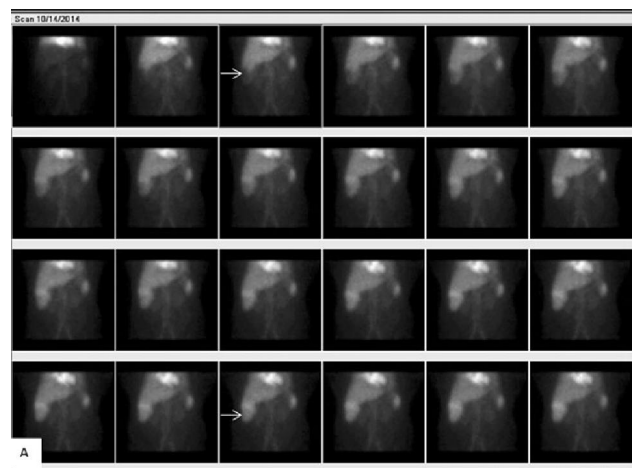


Figure 1: (A) Non-enhanced axial CT abdomen showing relatively low attenuation area in the liver (black arrows) with a focal area of capsular retraction of overlying liver (white arrows). (B) contrast enhanced CT abdomen axial image showing a low area demonstrated on plain scan (Figure 1A) shows peripheral puddling (black arrows) characteristic of a hepatic haemangioma; (C) contrast enhanced image showing a small hyperintense haemangioma of spleen (white arrow).

Technetium-99m tagged RBC scan (20 mCi) was performed and dynamic images showed relatively reduced perfusion over segment VI of liver in early dynamic frames. Blood pool images revealed a large area of progressive enhancement of blood pool activity involving segment VI of liver (Fig. 2). Splenic and vertebral lesions seen on CT were not outlined most likely due to limitation of camera to resolve small sized lesions. All these findings were



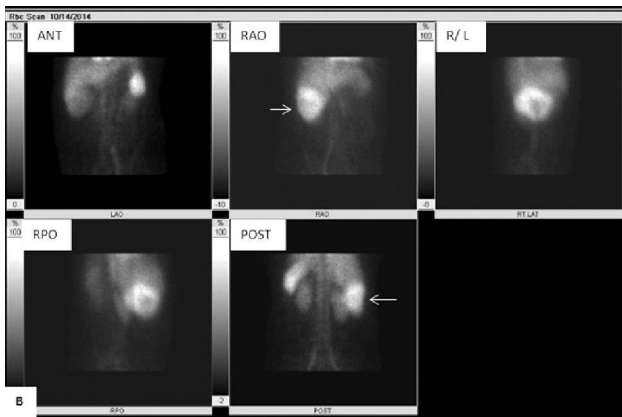


Figure 2: Tc-99m tagged RBC scan. Dynamic images (A) show an area of reduced arterial flow in early image over liver segment VI with progressive enhancement in blood pool images (arrow); multiple static (B) images a large well outlined area of enhanced tagged RBC involving liver segment VI (arrow).

consistent with multi focal haemangiomas with a giant one involving the liver. On the basis of these evidences biopsy was considered unjustified. Patient was referred to vascular surgeon for resection of the lesion.

Discussion

Haemangiomas are the most common benign slow flow venous lesions which are usually asymptomatic and diagnosed incidentally. They become symptomatic due to their size and associated mass effects resulting in pain, nausea, vomiting, rupture, hemorrhage and thrombosis. In our case, there was a past history of similar abdominal pain with thrombocytopenia and this raised the possibility of consumptive coagulopathy associated with giant haemangiomas (Kasabach Merritt's syndrome).⁷ However, our case did not have a full complete spectrum of this syndrome and it is considered as a self-limiting case as reported in literature as well.⁸ As haemangiomas are detected incidentally, one has to be familiar with its imaging features to differentiate it from primary or secondary hepatic neoplasms.

Ultrasound is the most commonly performed procedure which initiates the diagnostic algorithm in most of the cases. Most of these lesions appeared homogeneous hyperechoic lesions. However, about

10% of haemangiomas are hypoechoic particularly in a fatty liver. Giant haemangioma may appear non-homogeneous due to internal thrombosis and necrosis and acoustic enhancement is common. On color Doppler feeding vessels may not be identified.²

Computerized tomography (CT) clearly defines most of the haemangiomas and post-contrast enhancement on dynamic sequences is related to the volume of vascular spaces. Plain or noncontrast images show focal, well-circumscribed, low attenuation lesions relative to liver parenchyma. Arterial phase images show non-homogeneous nodular peripheral enhancement (although smaller lesions may show a uniform enhancement as well). Portal venous phase reveal a centripetal pattern of peripheral enhancement. Finally delayed images reveal further irregular fill-in of lesion rendering it iso- or hyper-attenuating to liver parenchyma.⁹ In this case CT images were pathognomonic for haemangioma as mentioned above but presence of capsular retraction was worrisome as it is commonly seen in malignant lesions. However, literature search have revealed few reports of capsular retraction in giant haemangioma due to fibrosis.^{10,11}

Magnetic Resonance Imaging (MRI) is the modality of choice with high diagnostic accuracy. They are hypointense on T1-weighted and markedly hyperintense on T2 weighted images giving characteristic light bulb sign.² Contrast enhanced images show peripheral nodular enhancement which progresses centripetally (inward) on delayed images. While diffusion weighted images (DWI) show hyperintense haemangioma due to T2 shine through rather than restricted diffusion.⁸

Radionuclide Nuclear Imaging using Technetium-99m labeled colloid (radiocolloid scan), haemangioma appears as cold area due to lack of reticulo-endothelial (RE) cells and considered a non-specific examination. While Tc-99m labeled RBC considered more specific as haemangiomas being slow flow venous lesion typically demonstrate decreased activity on initial dynamic images followed by increased activity on delayed blood pool images.

Single photon emission computerized tomography (SPECT) imaging is more sensitive than planar imaging and for lesions greater than 2-3 cm due to limited spatial resolution of gamma camera.¹² In this case we did only planar imaging and failure to outline splenic and vertebral lesions is due to these facts.

Conflict of Interest: None

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