HEPATIC PSEUDOTUMOURS IN A PATIENT WITH IVEMARK SYNDROME - A CASE REPORT

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ABSTRACT

Cardiac failure secondary to hepatic venous congestion is the predominant cause of mortality as cases of asplenia associated with cyanotic heart disease may present with hepatomegaly. C.T. imaging; may give, the appearance of 'Pseudotumours' secondary to hepatic venous congestion. We presents case of Ivemark syndrome with multiple hypodense areas in the liver as a recognised case of pseudotumour secondary to hepatic venous congestion. This entity should be recognised to avoid pitfall in the diagnosis of this condition and to avoid unnecessary cost and morbidity.

INTRODUCTION

Computed tomography (CT) has a remarkable capacity for evaluating the liver. It is clinically a valuable tool and is useful imaging modality in determining presence and the extent of mass lesions in the liver. CT has a high degree of specificity in the diagnosis of hepatic masses,[2] however there remains a variety of conditions which may mimic neoplastic changes and present as 'pseudotumour' appearance on contrasted CT scan imaging of the liver and if these condition is not recognised, it may caused a pitfall in the diagnosis of liver masses. We would like to illustrate a case of Ivemark Syndrome as one of the recognised cause of hepatomegaly resulting in hepatic pseudotumour, and emphasised the importance of recognising this condition to avoid an unnecessary mismanagement in the diagnosis of liver masses.

CASE REPORT

A 2 year child with known complex cyanotic heart disease with dextrocardia, primary atrio-septal defect, double outlet right ventricle, single atrio-ventricular valve and pulmonary stenosis presented with facial puffiness following an upper respiratory tract infection. On examination the child was cyanosed with facial puffiness, and also pitting oedema of the lower limbs. There was dextrocardia, cardiomegaly, hepatomegaly and ascites. A clinical diagnosis of nephrotic syndrome in child with cyanotic heart disease was made. However in view of the hepatomegaly a CT of the abdomen performed. This confirmed hepatomegaly and dextrocardia. The hepatomegaly occupied the entire right and left hypochondria and was of homogenous density pre-contrast. Post-intravenous iodinated contrast medium showed multiple low attenuation areas within both lobes of the liver (Figure 1). The spleen was absent with the stomach fundus on the right side. A diagnosis of multiple liver lesions possibly tumour deposits was made. The differential diagnosis of perfusion defects secondary to venous congestion was also considered. An ultrasound done subsequently showed a homogenous echo-pattern with no focal

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lesions within the liver. the hepatic veins and the intrahepatic IVC were dilated (Fig. 2) suggesting right heart failure. In the peripheral blood film Howell Jolly bodies were present. The a-foeto-protein was normal. The patient was treated with antifailure medication, steroids for the nephritis and penicillin for the asplenia. The patient responded and was discharged. On further follow up, there is no increase in size of hepatomegaly.

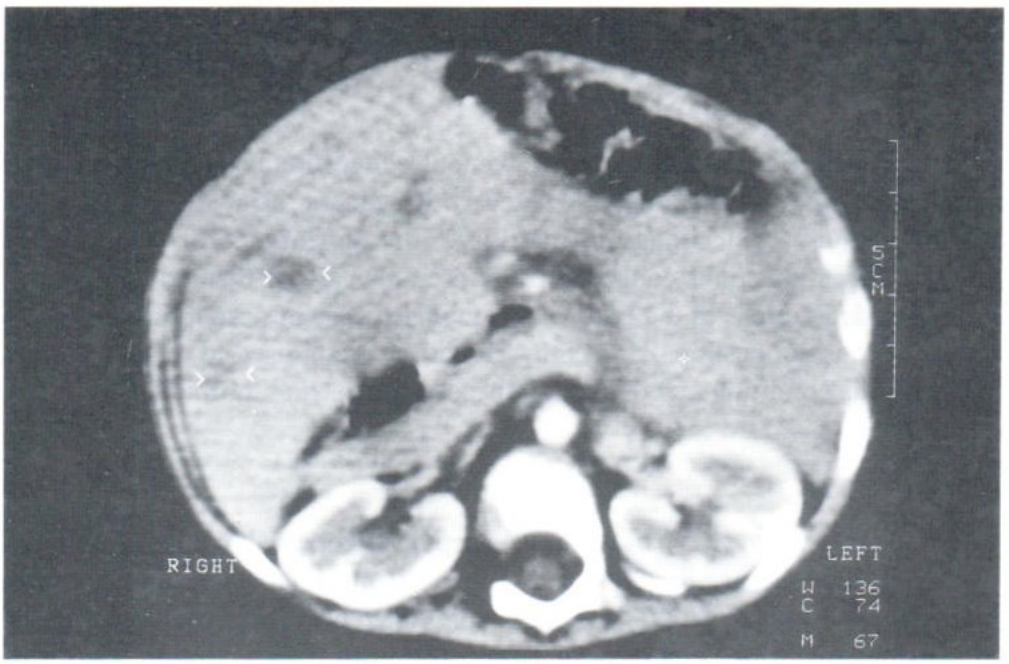

Fig 1. Axial CT scan of the abdomen after IV contrast enhancement shows hepatomegaly with multiple hypodense areas within the liver (arrowheads).

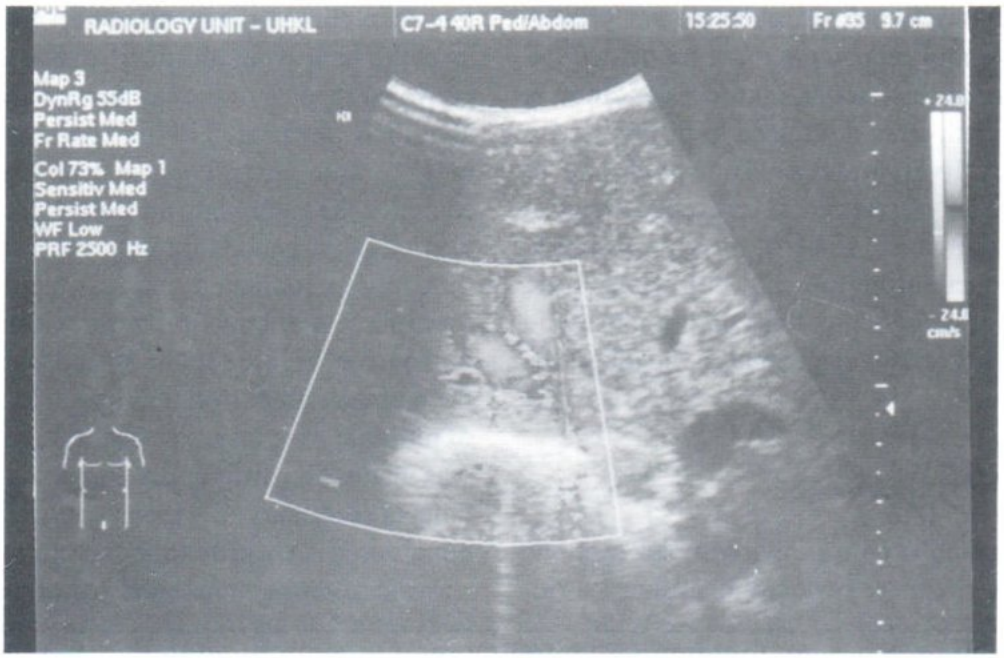

Fig 2. Doppler Ultrasound of the liver shows flow within the hepatic veins which are dilated but patent. The liver is of homogenous echogenicity.

DISCUSSION

CT is excellent at the demonstration of lesions in the liver.[2] In addition this modality is cheaper and more readily available than MRI and allows better access to the patient although the use of ionising radiation is a drawback. The diagnosis of liver masses with CT depend on the differential attenuation and enhancement pattern. The sensitivity of CT in defining the tumour is well established. There however remain several conditions that may mimic tumour giving rise to 'Pseudotumour' appearance. If these pitfalls are not recognised it may lead to an increase patient morbidity in term of unnecessary longer hospitalisation, extra investigation and even intervention such as biopsy or surgery.

Ivemark syndrome was first described in 1955[3] and is characterised by asplenia, malformation of conotruncus of the heart and abnormal lobulation of the lung. Abnormalities of the conotruncus of the heart include transposition of great artery in 72% of the cases and also pulmonary venous drainage in 72%. Death occurs in 79% of the cases in the first year of life due to cardiac failure and anoxia.[3,4,5] Cardiac failure occurring in this patient may cause a hepatic venous congestion and obstruction due to egress of blood from hepatic vein to the right heart. There is no associated increase risk of hepatic tumour in these patients. Multiple low attenuation areas within an enlarged liver in the patient presented raised the possibility of pseudotumour and the ultrasound of abdomen revealed dilated IVC and hepatic veins. In our patient although no biopsy was done to confirm the diagnosis, the clinical correlation of the clinical problem, and the correct choice of imaging modality plus the recognition of hepatic 'pseudotumour' has helped in making the correct diagnosis. In addition there was no progression of the hepatomegaly.

Recent report by Kane et al[1] illustrates multiple causes of hepatic pseudotumour appearance in children. These pseudotumours may result from chronic liver diseases such as cirrhosis and also fatty infiltration secondary to various conditions. In Tyrosinemia Type I also known as hepatorenal tyrosinemia in which there is a deficiency of fumaryl acetoacetate hydrolase. Accumulation of fumaryl and malelylacetate within the liver results in liver failure and cirrhosis which on CT gives rise to the appearance of multiple nodules with high or low density.

Other recognised causes of pseudotumour include Neimann Pick disease, Gaucher disease, Alpha I Anti Trypsin and also fatty infiltration secondary to alcoholic liver disease. In Neiman Pick disease the appearance of multiple low attenuation lesion in the liver is secondary to the accumulation of complex lipid, while in Gaucher disease it is secondary to accumulation of glucocerebroside. In alcoholic liver disease fatty in filtration may be diffuse or focal areas of low attenuation within the liver.

There have been no reported cases of hepatic venous congestion secondary to cardiac failure that have produced a pseudotumour appearance. The causes of hepatomegaly with multiple low attenuation areas include necrotic metastases, abscesses and other cystic masses.[6] In hepatic abscesses, the clinical history and pattern of rim enhancement may help in differentiating it from other cause of pseudotumour while the CT value may help in the differentiating from cystic liver disease.[7] In addition the correlation with other imaging modalities like ultrasound or MRI has help differentiate this from pseudotumour as was in the case presented. MRI with its multiplanar capability, higher soft tissue contrast resolution and pattern of enhancement is another modality that may be helpful in differentiation of various causes of focal hepatic masses and may also be able to differentiate them from pseudotumour.[8]

In summary, the presence of low attenuation lesions on the post intravenous contrast

enhancement CT scan of the liver may not always be real and that the pseudotumour has to be considered in the case presenting with congestive cardiac failure, where the clinical problem, symptoms, signs and ultrasonography have been helpful in making the correct diagnosis of a pseudotumour. This pitfall must be recognised to avoid unnecessary cost in further investigation and morbidity from inappropriate treatment.

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