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CASE REPORT
Cardiac tumors

Right atrial myxoma in a patient with Budd-Chiari syndrome (RCD code: VI-1A.1)

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Abstract

Budd-Chiari syndrome (BCS) is a rare clinical disorder which is caused by obstruction of the major hepatic veins or suprahepatic portion of the inferior vena cava. Either thrombosis or a mechanical venous obstruction may be present. BCS has been shown to be associated with a number of diseases including polycythaemia vera, myeloproliferative disorders, malignancy or congenital web of the gastro-intestinal tract. Right-sided cardiac tumours, although rarely reported as a cause for BCS, are generally considered as a surgically curable cause for this otherwise potentially lethal clinical condition. JRCD 2018; 3 (6): 218–220

Key words: rare disease, cardiac tumour, cardiac magnetic resonance imaging, computed tomography, echocardiography, cardiac surgery

Case presentation

A 29-year-old female was referred for cardiac magnetic resonance (CMR) for evaluation of a right atrial mass, found on a computed tomography (CT) scan and an echocardiogram. She presented with right hypochondrial pain and abdominal distension and was diagnosed with Budd-Chiari syndrome (BCS). CT scan showed evidence of thrombosis in the hepatic veins and hepatic and suprahepatic portion of the inferior vena cava (IVC). The CT scan revealed a filling defect in the right atrium and evidence of subsegmental pulmonary embolism. Echocardiogram showed a lesion in the right atrium, with thrombus or tumour being the differential diagnoses. No evidence of haematological malignancy or polycythaemia was found on routine blood tests.

CMR with gadolinium enhancement was performed, which showed a large, mobile, irregular shaped, pedunculated mass in the right atrium. It measured 32 mm × 21 mm and was attached to the interatrial septum at the site of the fossa ovalis, obstructing the tricuspid valve. The mass was isointense on T1 weighted images and iso- to hyperintense on T2 weighted images. The mass showed some perfusion on first pass perfusion imaging with gadolinium injection, ruling out the possibility of thrombus. No hyperenhancement was seen in late gadolinium images (Figures 1–3).

A diagnosis of right atrial myxoma was made on the basis of the above findings.

Patient management and follow-up

The patient underwent surgical removal of the mass and histopathology confirmed it to be a myxoma (Figures 4–5). The patient's clinical condition improved after surgical removal of the mass.

Discussion & review of literature

BCS is a rare clinical condition caused by hepatic venous outflow obstruction. The obstruction may be at any level between the small hepatic veins and the IVC junction with the right atrium. It results in centrilobular hepatic congestion with the subsequent development of fibrosis, portal hypertension, and cirrhosis.

The BCS is called primary when the obstruction is due to thrombosis or congenital web and is classified as secondary when the obstruction is caused by an extrinsic compression or invasion by an abscess, cyst, or tumour [1–4]. The risk factors for thrombosis and hypercoagulable state are polycythaemia vera, myeloproliferative disorders, paroxysmal nocturnal haemoglobinuria, and condi-

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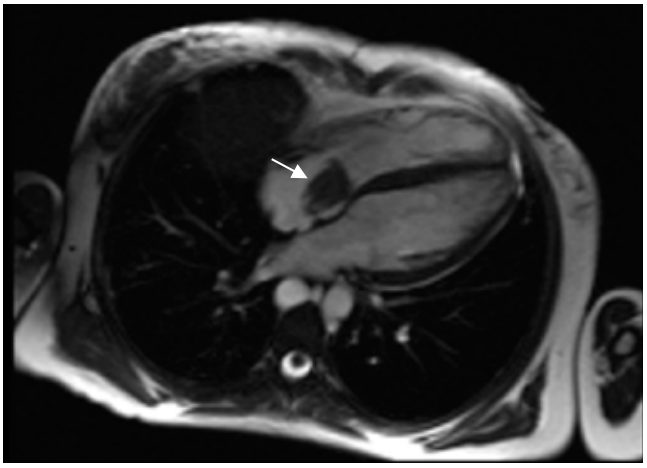

Figure 1. Steady-state, free precession cardiac magnetic resonance image showing large, mobile, irregular shape, pedunculated mass in the right atrium (arrow)

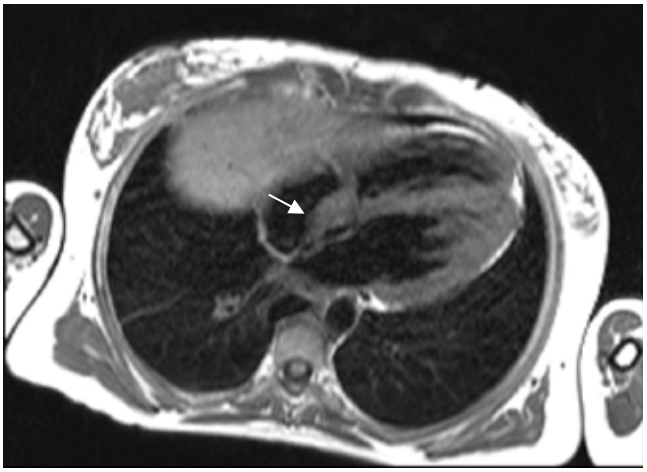

Figure 2. T-1 weighted cardiam magnetic resonance image showing an isointense pedunculated mass attached to the interatrial septum at the site of fossa ovalis, obstructing the tricuspid valve (arrow)

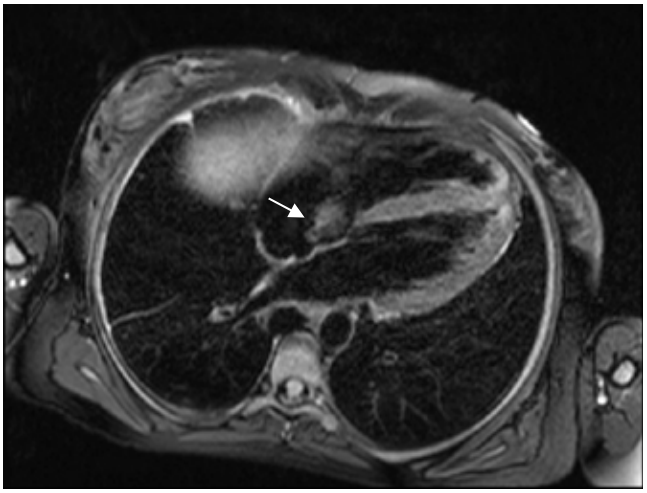

Figure 3. T-2 weighted cardiac magnetic resonance image showing and iso to hyperintense mass attached to the interatrial septum at the site of fossa ovalis, obstructing the tricuspid valve (arrow)

tions such as protein C and protein S deficiency or factor V Leiden mutations. Cardiac tumours are rarely associated with the development of BCS [2, 3].

Clinical presentation of BCS may be acute, subacute, or chronic and depends upon the rapidity of venous occlusion and development of collateral circulation. The acute form of BCS may present with fulminant hepatic failure, encephalopathy, jaundice, ascites, and tender hepatomegaly [1]. The chronic form of BCS is usually characterized by vague symptoms for about 6 months, presenting with portal hypertension and onset of ascites. Blood tests are generally consistent with chronic liver disease and show low albumin, prolonged prothrombin time and near normal aminotransferase. In our case, the patient presented with the chronic form.

Non-invasive imaging modalities are often used for the diagnosis of BCS. Ultrasound is useful and can reveal the location of the obstruction. Abnormalities which can be detected by ultrasound are the non-visualization of the hepatic veins, areas of stenosis, the prominence of collateral veins, and proximal dilatation. Doppler can improve diagnostic accuracy by detecting altered or absent hepatic vein flow [5]. CT scan is another non-invasive imaging modality, commonly used for the diagnosis of BCS. Post-contrast CT scan can show hypodense filling defects in the IVC and hepatic veins, which are highly specific for the diagnosis of thrombosis. Other diagnostic findings on CT scan include non-visualization of the hepatic veins or IVC, and changes in the attenuation of hepatic parenchyma [5]. An initial diagnosis of BCS was made using ultrasound in our case, however CT scan was done to confirm the diagnosis.

Echocardiography is useful for diagnosing right-sided cardiac tumours as a rare cause of BCS. Differential diagnosis of a mass in the right atrium using echocardiogram includes thrombus, myxoma, malignant tumour, or metastasis [6]. Thrombi in the right atrium are very rare and risk factors for thrombi include central venous lines, enlarged cardiac chambers, and arrhythmias. Co-existence of a thrombus in the right atrium and IVC is rare and usually occurs in malignancy such as renal cell carcinoma. Primary tumours of the heart account for only 0.3% of cardiac tumours [7]. Atrial myxomas are the most common primary tumours of the heart and account for 35–50% of primary cardiac tumours [8]. They are usually located in the left atrium and only 20% of cardiac myxomas originate from the right atrium [9]. There are a few cases of BCS reported in the literature which are associated with right atrial myxoma [2, 3].

CMR imaging and CT scans are also useful in diagnosing right atrial myxomas. Myxomas usually have a gelatinous nature and may manifest as a low attenuation mass on CT. In some cases, calcification may be present [10]. Interestingly, in our case, the right atrial mass was first seen on CT and then on echocardiogram and finally confirmed as myxoma using CMR.

CMR is particularly sensitive for detecting thrombi and differentiating cardiac tumours due to better tissue characterization. Thrombi appear as dark structures surrounded by contrast enhanced blood. Atrial myxomas are heterogeneous structures on contrast CMR imaging and usually appear isointense on T1 weighted images and hyperintense on T2 weighted images.

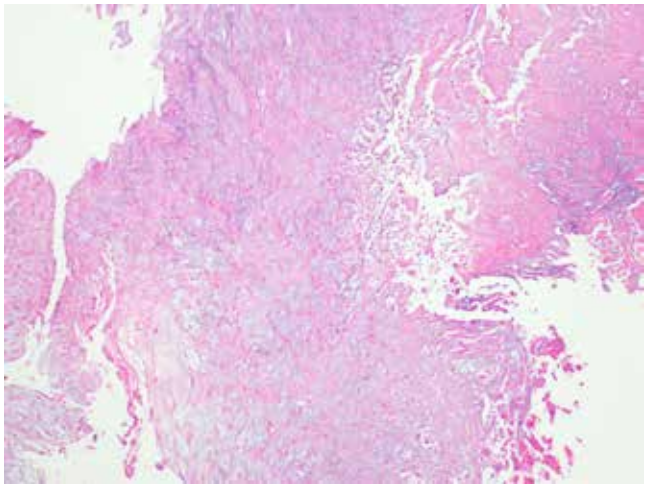

Figure 4. Hematoxylin and eosin stain photomicrograph at 100X magnification of atrial myxoma

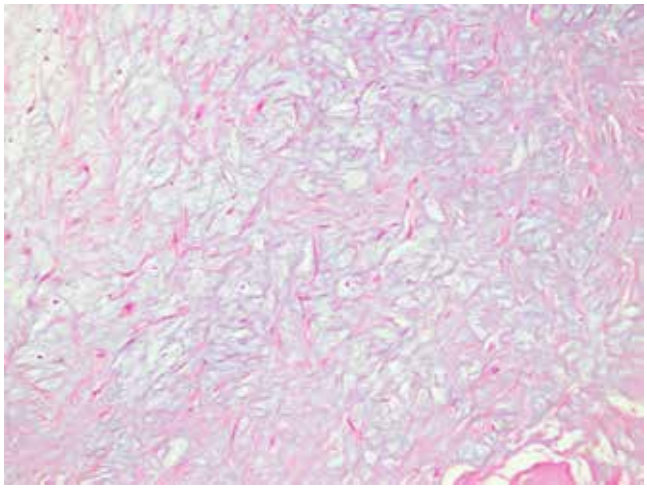

Figure 5. Hematoxylin and eosin stain photomicrograph at 200X magnification showing stellate fibroblasts present in a myxoid matrix

In our patient, successful removal of the myxoma resulted in rapid resolution of the BCS. Thus, right atrial myxoma is a curable cause of BCS. It should be considered in the differential diagnosis, particularly when other more common causes of BCS are absent.

Conclusion

Although rare, the possibility of right atrial myxoma should be considered in the differential diagnosis among the causes of BCS. This is critical, as surgical resection of the myxoma is curative for BCS, which is otherwise a potentially lethal condition.

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