Right atrial spleen

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SUMMARY A large cardiac tumour occupying most of the right atrium and the right ventricle and causing inflow obstruction to the right heart was confirmed by cross sectional echocardiography in a 41 year old man. After surgical resection histological examination showed that the atrial tumour had the characteristics of splenic tissue. Possible mechanisms for the development of such a tumour include an origin analogous to that of an accessory spleen or the implantation and subsequent growth of lymphoid tissue in a pre-existing superior vena caval or high right atrial angioma.

Primary cardiac tumours are rare. In necropsy series the incidence is extremely low (0.0017–0.28%). In most reported series about 75% of all cardiac tumours are considered to be benign histologically. Angiomas account for less than 5% of the total number. They are more common in the right heart chambers and generally as sessile. Histologically, the tumours consist of spaces lined with endothelium, which may contain blood, lymph, or thrombi.¹

We report a patient who presented with signs of inflow obstruction to the right heart due to an atrial tumour with an unusual histological appearance.

Case report

A 41 year old man was admitted because of slowly progressive dyspnoea and ascites. A cardiac murmur had been heard when he was 5 years old. Until 1980 he had worked as a waiter and for 20 years had consumed large amounts of alcohol. He was referred to our hospital with a presumptive diagnosis of alcoholic liver cirrhosis.

On physical examination the jugular veins were severely congested with a very prominent "a" wave. The heart was not enlarged. A grade 2/6 high frequency pansystolic murmur and a grade 2 high frequency diastolic murmur with presystolic accentuation were loudest at the left sternal border. The abdomen was distended by ascites, and the liver and spleen were not palpable. There was, however, no peripheral oedema.

A 12 lead electrocardiogram showed sinus rhythm with an undetermined mean frontal plane QRS axis.

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Fig. 1 Cross sectional echocardiogram in the apical view showing an echo dense mass at the level of the right atrioventricular valve annulus partially occupying the right atrium as well as the right ventricle. The arrow probably indicates an adherent part of the septal tricuspid leaflet.

In all 12 leads the QRS complexes showed low voltage, and the P waves had a normal voltage in the limb leads. There was a disproportionate relation between the amplitude of the P waves and that of the QRS complexes. Chest radiographs showed clear lung fields and a moderately enlarged heart in the posteroanterior projection.

The phonocardiogram showed a high frequency diastolic murmur with presystolic accentuation starting 0.10–0.12 s after the second heart sound. The raised central venous pressure, the pronounced "a" wave in the venous pulse tracing, signs of tricuspid disease, and the absence of signs of left heart failure, pulmonary hypertension, or pericardial disease suggested an inflow obstruction to the right heart. This

Fig. 2 Microscopical appearance of the right atrial tumour showing a thick walled artery surrounded by a mantle of lymphocytes forming a sheath around these arteries. (Giemsa stain $\times 150$.)

was confirmed by cross sectional echocardiography. An apical approach showed a non-mobile, homogeneous echo dense mass occupying most of the right atrium and extending across the tricuspid valve into the right ventricular cavity (Fig. 1). The echocardiographic diameter of the mass was $7 \times 4 \times 4$ cm. The nature of the mass could not be specifically identified. The pattern of motion of the echo on M mode echocardiography was not, however, characteristic of a myxoma.

SURGICAL FINDINGS

The chest was entered through a midline sternal incision. From the outside the right ventricle looked severely hypoplastic, with the left anterior descending coronary artery displaced rightwards. The right atrial wall was severely hypertrophied. The right atrium contained a dark and smooth solid tumour attached by a single short cord like pedicle to the lowest segment of the superior vena cava. The tumour extended across the tricuspid annulus into the right ventricle. The tricuspid valve leaflets were deformed and fenestrated. Half of the septal and anterior leaflets were adherent to the right ventricle and only the posterior leaflet retained some mobility. The papillary muscles were hypoplastic and the chordae tendineae were very short. The tumour and tricuspid valve were removed and a Ionescu-Shiley prosthetic valve (size 31) implanted in the tricuspid annulus. The patient died 15 days after the operation as a result of septic shock.

HISTOLOGICAL FINDINGS

The surgical specimen was fixed in 4% neutral phosphate buffered formaldehyde. Tissue samples were embedded in paraffin and epon for light microscopy

and electron microscopy respectively. Sections cut at $5 \mu\text{m}$ were stained with haematoxylin and eosin, Giemsa, Gordon and Sweet's reticulin, periodic acid Schiff (PAS), Alcian blue, van Gieson's elastic tissue stain, Perl's stain for iron, and methyl green pyronine.

The tumour, which measured $7 \times 5 \times 4$ cm, was enclosed within a thin greyish white capsule, which was continuous with the endothelial lining of the atrium. The cut surface varied in colour from deep red to greyish red.

Light microscopy showed that the tumour was surrounded by a collagenous capsule. The pedicle contained only collagenous connective tissue and was traversed by both arteries and veins. The parenchyma showed a reticular framework and endothelial lined sinusoids, some of which had cystic dilatations. The sinusoids were filled with red cells and had foci of haematopoiesis. Thick walled arteries were present and small lymphocytes formed a sheath around these arteries. The microscopical appearance was identical to that of normal splenic tissue except for a paucity of trabeculae (Fig. 2).

Electron microscopy showed an intermingling of small capillaries, red blood cells, and some lymphocytes. The capillaries consisted of a single layer of irregular endothelial like cells with cell junctions and a periendothelial basal lamina. Pericytes and histiocytes were not found. The appearance was similar to that of splenic tissue.

When the tumour was compared with splenic tissue obtained at necropsy by immunoperoxidase techniques for identifying immunoglobulins in plasma cells as well as histiocytic markers no differences could be found.

NECROPSY FINDINGS

At necropsy an enlarged spleen weighing 320 g was present at the normal site in the left upper abdomen and showed no abnormalities other than signs of septicæmia and small foci of haematopoiesis. The liver was moderately enlarged, weighing 1940 g. The parenchyma showed severe cardiac fibrosis and steatosis.

Discussion

The most striking feature of this case was the hitherto unreported finding of a cardiac tumour consisting of splenic tissue. Several possible mechanisms for the pathological development of a right atrial spleen exist. Embryologically, the primordium of the spleen appears as a thickening of coelomic epithelium on the left side of the dorsal mesogastrium. Several separate splenic masses originate here simultaneously and fuse to form a lobulated spleen.² Failure of the adjoining

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splenic areas at the dorsal mesogastrium to fuse could give rise to the formation of an accessory spleen. Invagination of a splenic area during cardiac development could explain the finding of accessory intracardiac splenic tissue, but this seems to be rather unlikely. Another possibility is a primary cardiac tumour of an angiomatous nature. Such tumours probably arise from angioblasts normally dormant in the myocardium. The angioblasts originate from primitive mesenchymal tissue and produce blood vessels, primitive plasma, and red blood cells.^{3,4} One theory focuses on the vasoformative reserve cell. Islands of vasoformative reserve cell hyperplasia may occur in the endocardium near the fossa ovalis. The spleen is essentially a part of the vascular system, and the vasoformative reserve cells could presumably also give rise to the development of an angiomatous tumour differentiating as splenic tissue. This angiomatous tissue might have become seeded by lymphocytes. The lymphoid tissue in the tumour is thought to be formed by a proliferation of lymphocytes in the area. Foci of haematopoiesis were present in both the tumour and the spleen. This myeloid metaplasia in the spleen is due to the undertaking of haemopoietic functions by the reticuloendothelial cells.⁵ Both the tumour and the patient's spleen cells showed this myeloid metaplasia. Furthermore, even the neoplastic nature of the tumour in our patient was unknown, since it is almost impossible to differentiate between malformations—such as hamartomas—and benign neoplasms of blood vessels on purely anatomical or clinical grounds.

From a clinical point of view the following findings are of special interest. The average delay between the onset of symptoms and the diagnosis of right atrial tumours in most reported cases was three years.¹ In our patient the history of a large consumption of alcohol and the pronounced ascites many years previously had led to an initial false diagnosis of alcoholic liver cirrhosis. A combination of features—namely, the high central venous pressure, the pronounced “a” wave in the venous pulse tracing, and the diastolic murmur with presystolic accentuation—suggested the possibility of a right sided inflow obstruction at the level of the tricuspid valve. The high frequency of the diastolic murmur is, however, unusual in tricuspid

inflow obstruction. Nevertheless, because the large solid tumour was wedged in the tricuspid orifice this characteristic of the murmur may be explained by the very high central venous pressure (25 mm Hg) and the subsequent contraction of the hypertrophied right atrium forcing the blood around the tumour via the slit like opening in the right ventricle. A definitive diagnosis was made by cross sectional echocardiography, which allowed the size of the tumour to be accurately estimated. Cross sectional echocardiography seems to be ideally suited for the non-invasive diagnosis of a right sided cardiac mass.⁶ The degenerative changes of the tricuspid valve were unusual. Fenestration of the atrioventricular valve leaflets, short chordae tendineae, and hypoplasia of the papillary muscles have not been reported in cases of atrial myxoma.

In conclusion we were unable to differentiate this cardiac atrial tumour from splenic tissue. The clinical findings suggest that the tumour had been present for a long time, which may indicate a congenital origin. Nevertheless, the true origin of this unusual tumour remains unknown.

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