Case report - Cardiac general

A diagnostic challenge – an unusual right atrial mass, 12 years following atrial septal defect surgery

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Abstract

A young girl presented with a right atrial mass, 12 years following ostium secundum atrial septal defect surgery. The unusual pathology and the various diagnostic considerations for an atrial mass are discussed.

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1. Clinical presentation

A 21-year-old woman presented with a five-year history of atypical chest pain. She had undergone surgical closure of an asymptomatic, 8 mm secundum atrial septal defect by primary suture at the age of nine years. Clinical examination was unremarkable. She was in sinus rhythm. A transesophageal echocardiogram (Fig. 1a) revealed a 1.8 cm × 1.0 cm lobulated, highly mobile, echogenic mass in the right atrium. A prominent eustachian valve and a Chiari network were present. MRI confirmed a mass attached to the posterolateral right atrial wall distal to the inflow of the superior vena cava and anterior to the sinus venosus portion of the atrial septum. The heterogenous 'salt and pepper' MRI appearance suggested an atrial myxoma.

On cardiopulmonary bypass, the mass was excised completely (Fig. 1b). It did not look like a myxoma. It was attached to the right atrial wall with a short narrow stalk. It seemed to arise from the site of an old suture piercing into the atrial wall at that site. The suture was most likely that placed for holding the temporary atrial pacemaker wires at the original operation. The postoperative course was essentially uneventful. Histopathology is described in Fig. 2.

2. Discussion

Most atrial septal defects are now electively closed in early childhood to prevent late onset complications. In a 25 years’ reoperation-rate review, Monroe had a 0.013% incidence for ASD [1]. Here, the preoperative features suggestive of a myxoma were the MRI picture, the female sex and the long duration elapsed since initial surgery. However, the absence of any myxoma cell, unexpectedly, excluded this diagnosis. A more detailed review reveals the atypical features in our case against this knee-jerk diagnosis.

A mass in the atrium commonly results from thrombus, infections with vegetations, and tumors. Primary cardiac tumors occur in 0.2 to 0.3% of all individuals; nearly 50% of which are myxomas. The usual sporadic variety occurs in the third and sixth decades of life and preferentially in women. Seventy-five percent of myxomas occur in the left atrium and only about 20% in the right atrium. Seventy-five percent originate from the interatrial septum at the inferior border of the fossa ovalis. Right atrial myxomas tend to have a broader base of attachment and are also more calcified.

The origin of the myxoma cell is debated [2]. This predilection for the fossa ovalis is important and was explained by the clustering of the ‘Prichard structures’ here. These are multipotent subendocardial mesenchymal cells; embryonic remnants capable of differentiating into different cell lines, explaining the occurrence of bone, cartilage, etc. in myxomas. Heavy calcification is reported in about 10% of myxomas. Rarely, the myxoma cells may almost be completely replaced or mummified, termed a cardiac ‘lithomyxoma’ [3]. A wide spectrum of other cardiac tumors, as well as thrombus are known to calcify.

In our case, the location away from the fossa ovalis, its small size, and the much younger age group were atypical. We found only one report of a myxoma developing after
Fig. 1. (a) Transesophageal echocardiogram showing mass, separate from the interatrial septum. (b) The mass within the opened right atrium.

Fig. 2. The mass had diffuse calcification (pink) enclosed by a rim of collagen on the surface. It showed focal ossification with a few inflammatory cells and a microfocus of cartilaginous metaplasia. The core of the mass had the occasional small vascular space. There were no identifiable myxoma cells (Hematoxylin and Eosin stain, ×16 magnification).

ASD repair [4]. It developed after 4 years on the atrial septum between the orifices of the coronary sinus and inferior vena cava. The suture line was uninvolved. We found three reports of the co-association of a myxoma with an ASD resulting in symptomatic right to left shunting in adults [5]. An anecdotal report described an atrial myxoma at the site of a trans-septal puncture for percutaneous balloon dilatation of the mitral valve. They concluded that some atrial myxomas might be reactive to trauma rather than neoplastic in origin [6].

There are few reports of atrial thrombus following atrial septal defect surgery. This has been described with primary suture and the patch closure techniques. This was usually within six months of surgery and usually arose from the repair site. However, a thrombus has been described occurring as a thin, stalked mobile mass on the free wall of the right atrium near the inferior vena cava, and was attributed to endocardial damage [7]. The main pre-operative feature against a thrombus in our case was its remote timing, its site away from the surgical secundum site and the presence of sinus rhythm.

Prominent right atrial anatomy can sometimes be misinterpreted. Such a 'pseudomass' include Prominent crista terminalis, lipomatous hypertrophy of the septum [8], eustachian valve mimicking a cystic mass, and the persistent embryologic right sinus venous valve known as the Chiari network. Werner found the Chiari network in 2–3% of normal hearts and is more often associated with the orifice of the IVC [9]. The prominent anatomical structures did not confuse the issue in our case.

3. Conclusion

In this case, after review of all the available pre and postoperative information, we conclude that the suture material served as an inflammatory focus, subsequent thrombus formation with eventual dystrophic calcification and ossification. One should be guided by a systematic approach in such an atypical case.

References