Constrictive Pericarditis Presenting as a Large Mediastinal Mass Causing Functional Tricuspid and Pulmonary Stenosis

Niklas F. Ehl, MD; Simon Sundermann, MD; Reinhard Geyer, MD; Svenja Thies, MD; Carola Epp, MD; Daniel Weilenmann, MD; Volkmar Falk, MD, PhD; Lucas Joerg, MD; Hans Rickli, MD; Micha T. Maeder, MD

A 71 year-old man was admitted with progressive shortness of breath and signs of right heart failure. Chest x-ray showed extensive pericardial calcifications (Figure 1). Echocardiography and computed tomography revealed a large mass (77×56 mm) in the anterior mediastinum compressing the right heart cavities, particularly at the level of the atrioventricular groove, resulting in hemodynamically significant functional tricuspid stenosis (Figure 2 and online-only Data Supplement Movie I). In addition, there was mild functional pulmonary stenosis caused by a circular calcified plaque (Figure 3 and online-only Data Supplement Movie II). Contrast uptake by the mass in the computed tomography images was minimal. Computed tomography–guided needle biopsy revealed only necrotic tissue. Cardiac magnetic resonance imaging demonstrated that the mass was heterogeneous, did not exhibit significant contrast uptake, and did not infiltrate the wall of the right-sided cardiac chambers, and also highlighted flow acceleration across the tricuspid valve by external compression (Figures 4 and 5 and online-only Data Supplement Movie III). A detailed history of a scar obviously from a median sternotomy revealed that the patient had undergone partial pericardectomy for calcific constrictive pericarditis of unknown origin 55 years ago. He underwent redo surgery, and the mass was excised; parts of the calcified right atrium also had to be resected (Figure 6) and were replaced by a pericardial patch. Histology of the macroscopically inhomogeneous and partially calcified fragments of the mass revealed a mix of extensive fibrosis with evidence of a chronic inflammatory process with reactive calcification and ossification, as well as necrotic areas (Figures 7 and 8). There were no signs of malignancy and no sign of specific inflammation such as granuloma. Immunochemistry for pancytokeratin-positive epithelial cells was negative. Findings were consistent with a chronic inflammatory process resulting in the formation of a pseudotumor.

We present a rare case of a mass in the anterior mediastinum, which became clinically manifest by compression of the right heart, causing functional right ventricular inflow and outflow tract obstruction. The patient’s history, the presence of pericardial calcifications, the absence of significant contrast uptake in computed tomography and magnetic resonance imaging, and the histological findings suggest that the mass corresponds to massive constrictive pericarditis, which rarely can present as a pseudotumor causing right heart compression including tricuspid stenosis. It may be hypothesized that the formation of the mass occurred in part as a consequence of the previous operation, which likely had been performed at the anterior aspect of the heart. Given that there were no granuloma within the mass and there was no evidence of a specific chronic inflammatory disease, the cause underlying pericardial constriction remains unknown. Large cardiac fibroma would have been a potential different...
tial diagnosis. However, the patient’s age (older adult versus infant/child/younger adult), the localization of the mass (extracardiac versus intramyocardial), and the histological findings (mix of extensive fibrosis and chronic inflammation versus a homogeneous mix of fibroblasts/fibrocytes and collagen only) argue against the presence of cardiac fibroma.

In this unusual case of a mass in the anterior mediastinum, the findings of different imaging techniques allowed for an excellent morphological and functional understanding of the cause of the patient’s symptoms and signs of heart failure, which was the basis for a successful surgical intervention. The patient recovered from the operation, and his symptoms markedly improved.

Disclosures

None.

References

Computed tomography of the chest as well as transesophageal echocardiography showing pulmonary stenosis (peak systolic pressure gradient 20 mm Hg) due to a circular calcified plaque (arrows). PT indicates pulmonary trunk; LPA, left pulmonary artery; Ao, ascending aorta; AV, aortic valve; and RVOT, right ventricular outflow tract.

Figure 4. Cardiac magnetic resonance imaging (still frame image from a SSFP 4-chamber cine series) showing an inhomogeneous mediastinal mass (MM) compressing the right atrium and right ventricle and causing flow acceleration across the tricuspid valve (arrow). The border of the mass is well defined, and there is no evidence of infiltration.
Figure 5. Cardiac magnetic resonance imaging. A, T2-weighted half-Fourier acquisition single-shot turbo-spin-echo showing a round mass compressing the right heart. B, Delayed enhancement images (10 minutes after 0.1 mmol/kg Gd-DOTA) showing mild patchy hyperenhancement of the tumor mass.

Figure 6. Intraoperative situs with opened calcified right atrium (arrow).

Figure 7. Hematoxylin and eosin–stained section demonstrating evidence of chronic inflammation and granulation tissue with fibroblasts, collagen fibers, and inflammatory cells (magnification ×50).
Figure 8. Hematoxylin & eosin–stained section demonstrating inhomogeneous tissue with extensive reactive calcification and ossification as well as necrotic areas with neutrophiles and fibrinous exudate (magnification ×50).
Constrictive Pericarditis Presenting as a Large Mediastinal Mass Causing Functional Tricuspid and Pulmonary Stenosis

Circulation. 2011;124:e487-e491
doi: 10.1161/CIRCULATIONAHA.110.017830

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/124/19/e487

Data Supplement (unedited) at:
http://circ.ahajournals.org/content/suppl/2011/11/04/124.19.e487.DC1

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org/subscriptions/