Localized constrictive pericarditis compressing and obstructing the right ventricular inflow tract due to a giant anterior calcified cardiac mass. A case report

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1. Introduction

Constrictive pericarditis is a rare disorder, characterized by thick pericardial fibrosis and calcification that causes progressively impaired diastolic filling of the heart with associated symptoms of heart failure [1]. The most common cause of this disease in underdeveloped areas of the world is either viral or bacterial, and when bacterial, it is likely related to be tuberculosis. Localized pericardial constriction was reported as a rare form of constrictive pericarditis [2,3]. Depending on the location of pericardial constriction, clinical presentation may be variable, including compression and obstruction of the right ventricular outflow tract, coronary obstruction, or pulmonary stenosis [4]. Obstruction of the right ventricular inflow is rarely reported. Small fluid collections are commonly observed between adhesions in patients with constrictive pericarditis; however large amount of hematoma are extremely rare. We report in line with the SCARE criteria [5] a case with localized anterior calcified constrictive pericarditis with a large hematoma formation, compressing and obstructing the right ventricular inflow tract.

1.1. Case presentation

A 72-year-old man presented a 2-year history of dyspnea and atrial fibrillation. During the last three months, the patient exhibited occasional paroxysmal nocturnal dyspnea and two-pillow orthopnea. The patient denied any chest pain, history of myocardial infarction or thoracic trauma in the past. Physical examination revealed moderate elevation of jugular venous pressure and mild hepatomegaly without other gross features of heart failure, and no pericardial knock or rub could be heard. A contrast enhanced angio computerized tomography clearly demonstrated a large spherical mass about 11 × 9 × 4 cm in the anterior pericardium, presenting as
a mediastinal tumor and compressing right ventricle inflow tract (Fig. 1A and B). At this time, the origin of the mass was unclear. However, the differential diagnosis included obstructive pericarditis and teratoma. A transthoracic echocardiogram identified the anterior cardiac mass and revealed again the presence of partial compression of the right ventricular free wall (Fig. 1C). The echocardiographic data are given in Table 1. Cardiac catheterization was performed to better elucidate the hemodynamic effect of suspected obstructive pericarditis on right ventricle filling, through observation of dynamic respiratory changes that occur in the heart during cardiac catheterization of the right cardiac side. During the same procedure the coronary angiography was performed and non coronary stenosis was identified.

With evidence of constriction confirmed, the patient underwent pericardectomy and resection of the mass. Before sternotomy, the central venous pressure was 20 mmHg. Upon gross examination before removal, the mass was noted to extend from the diaphragm to the top of the right atrium, and from the right phrenic nerve to the anterior interventricular groove and right ventricular infundibulum (Fig. 2A). The pericardium was thick. The dissection was started from the anterior surface of the right ventricle toward the mass on the right atroventricular groove (Fig. 2B). The outer calcified layer of the pericardial mass was a thick layer of calcification surrounded an inner amorphous low density material. This layer was easily opened and the contents of the mass appeared like old coagulated blood (Fig. 2C) which was evacuated with a sterile spoon (Figs. 2 D and 3 A). Then the inferior calcified layer of the pericardial mass which was extremely adherent with the epicardium, was carefully excised, without the use of cardiopulmonary bypass, from the origin of the great vessels to the diaphragm and all areas between the right and left phrenic nerves (Fig. 3B). The removed spec-

Table 1

| Echocardiographic data. | Preoperative | Postoperative |
|------------------------|--------------|--------------|
| Left ventricular ejection fraction (%) | 43           | 60           |
| Mitral E velocity in inspiration (cm/s) | 82           | 112          |
| Mitral E velocity in expiration (cm/s) | 98           | 125          |
| Percent change in mitral E velocity | 32           | 14           |
| Lateral e‘ velocity (cm/s) | 12           | 9            |
| Medial e‘ velocity (cm/s) | 13           | 7            |
| E/A ratio in inspiration | 1.5          | 2            |
| E/A ratio in expiration | 1.9          | 2            |
| Deceleration time | 1.3          | 1            |
| expiration/inspiration |              |              |
| Hepatic veins systolic reversal velocity in inspiration(cm/s) | 2.2          | 2            |
| Hepatic veins systolic reversal velocity in expiration(cm/s) | 16           | 23           |
| Hepatic veins diastolic velocity in inspiration(cm/s) | 17           | 34           |
| Hepatic veins diastolic velocity in expiration(cm/s) | 24           | 33           |
| Hepatic veins diastolic reversal velocity in inspiration(cm/s) | 50           | 70           |
| Hepatic veins diastolic reversal velocity in expiration(cm/s) | 17           | 49           |
| Percent change in superior caval vein velocity | 1.4          | 0.6          |
| Tricuspid regurgitation velocity max (m/s) | 2.2          | 1            |
mens were examined pathologically and were noted to have no evidence of granulomatous inflammation, acute inflammation, or tumor. The removed mass consisted solely of thickened and fibrotic pericardium with nodular calcifications (Fig. 3C) and coagulated blood. Polymerase chain reaction culture of the specimen of pericardium and pericardial fluid revealed no tuberculosis bacilli. The final diagnosis was idiopathic constrictive pericarditis. The patient was discharged 6 days later. Three months later, the clinical status of the patient was significantly improved with normal left ventricular ejection fraction.
2. Discussion

This case represents a very rare form of localized constrictive pericarditis consisting in a giant anterior calcified pericardial mass. Other cases with anterior localized constrictive pericarditis have been rarely reported [2–4,6]. Our patient experienced right ventricular free wall compression and obstruction of the right ventricular inflow as the cause for his clinical symptoms. Constrictive pericarditis should be taken into consideration when generalized symptoms of right-sided heart failure and decreased cardiac output are present [7]. Elevation and equalization of ventricular filling pressures is also often present in patients with constrictive pericarditis.

The differential diagnosis with other calcified masses in the anterior mediastinum such as teratoma, hemopericardium after blunt trauma and idiopathic or tuberculosis constrictive pericarditis should be considered [8]. The most common etiologies of this disorder are viral infection, renal failure, tuberculosis, radiation therapy, collagen vascular disease, prior pericardiopathy, and idiopathic constrictive pericarditis. In this patient no specific cause was identified for the calcified constrictive pericarditis with the associated mass on the anterior pericardium. Often there is an occult event that triggers inflammation of the pericardium, leading to fibrosis with calcification and sometimes adherences on the pericardium. Sometimes small collections of fluid entrapped between adhesions in patients with constrictive pericarditis are reported [9]. The giant mass observed in the case presented here consisted of a thick layer of calcified tissue within the pericardial cavity. The patient’s previous disease history, clinical course, pathological examinations of the resected pericardium, and the content of the mass showed no specific etiology for its formation or calcification of the pericardium. The mechanism by which such a large amount of coagulated blood was entrapped within the mass was unclear. The inflammatory changes of the pericardium might lead to neovascular process, which is often fragile, easily ruptured, and results in a large amount of blood in the cavity. Indeed, blood was confined in the cavity mass. Pericarditis is a common cause of compression of cardiac structures, and when compression is severe, may cause obstruction. Several cases of right ventricular outflow tract obstruction are reported, however there are only two reports in which the right ventricular inflow tract was obstructed because of chronic constrictive pericarditis [10,11]. It needs to be remembered that constrictive pericarditis can form a large mass, which is difficult preoperatively to distinguish from a mediastinal tumor, and it might obstruct the cardiac chamber, including the right ventricular inflow tract as in our case. Pericardectomy is the only established treatment for chronic constrictive pericarditis. Careful dissection is mandatory for a successful procedure.

Conflict of interest

The authors declare that there is no conflict of interest regarding the publication of this paper.

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Ethical approval

The study received ethic committed approval, by the institutional ethic committee who fully approved.

Consent

The patient and family gave their total consent.

Author contribution

Edvin Prifti and Krakulli performed the first and the second procedure. Hortensa Gjerjo and Nedri Hasimi performed the diagnosis and paper writing.

Guarantor

Edvin Prifti.

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