Front. Med. 2016, 10(3): 356–359
DOI 10.1007/s11684-016-0467-6

CASE REPORT

A rare chronic constrictive pericarditis with localized adherent visceral pericardium and normal parietal pericardium: a case report

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Abstract **Classic constrictive pericarditis (CP) is characterized by fibrous scarring and adhesion of both the visceral pericardium and the parietal pericardium, which leads to restricted cardiac filling. However, diagnosing CP with normal thickness pericardium and without calcification is still a challenge. The predominant cause in the developed world is idiopathic or viral pericarditis followed by post-cardiac surgery and post-radiation. Tuberculosis still remains a common cause of CP in developing countries. In this report, we describe a rare case of idiopathic localized constrictive visceral pericardium with normal thickness of the parietal pericardium in a middle-aged man. The patient presented with unexplained right heart failure and echocardiography showed moderate bi-atrial enlargement which should be identified with the restrictive cardiomyopathy. After 10 months of conservative treatment, the progression of right heart failure was remaining. A pericardiectomy was performed and the patient recovered. This case serves as a reminder to consider CP in patients with unexplained right heart failure, so that timely investigation and treatment can be initiated.**

Keywords constrictive pericarditis; heart failure; pericardiectomy

Introduction

Constrictive pericarditis (CP), caused by an inelastic pericardium, is a disorder of cardiac filling. The etiology of CP is tuberculosis, trauma, autoimmune, tumor, idiopathic causes, etc. The pericardium is a fibroserous membrane composed of the visceral and parietal pericardia. CP is often diagnosed when a fibrotic, thickened, frequently calcified, shell-like, and adherent pericardium restricts diastolic filling of the heart [1]. However, pericardial thickening is not required for the diagnosis. Typical clinical manifestations are absent in the early stages of CP, thus, early diagnosis is extremely difficult. CP with thickened adhesions in both the visceral pericardium and the parietal pericardium is common. In past case reports (Table 1), only one patient with no calcification and no patients with normal parietal pericardium thickness have been described [2–13]. However, CP with localized visceral pericarditis and normal thickness of the parietal pericardium is possible. The inelastic can limit cardiac volume and make cardiac output decreased. Early diagnosis and treatment is important. We herein describe a rare case of idiopathic and localized constrictive visceral pericardium with normal thickness of the parietal pericardium in a middle-aged man.

Received February 24, 2016; accepted June 12, 2016

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Case report

A 50-year-old man presented with shortness of breath, fatigue, and swelling of the legs, which were particularly evident after exertion, and had lasted for one month. The patient was treated for common cold in a private clinic for a week with no improvement, and there was no history of any diseases. Physical examination revealed a mildly elevated jugular venous pressure and pitting edema in the lower limbs. His heart rate was normal, but the rhythm was irregular. An electrocardiogram revealed atrial fibrillation (AF).

Table 1 Previous case reports of idiopathic constrictive pericarditis (published in English)

References	Year	Patient		Pericardium		Characteristic	Therapy	Prognosis
		Age (year)	Sex	Calcified	Thickened			
Lambelin *et al.* [2]	2014	55	M	Yes	NM	Pheochromocytoma	Subtotal pericardectomy	Recovery
Kushida *et al.* [3]	2014	61	F	NM	NM	Neoplastic constrictive pericarditis	Chemotherapy	Death
Samara *et al.* [4]	2010	66	F	NM	Yes	Transient, pericardial constriction	Steroid treatment	Recovery
Nachimuthu *et al.* [5]	2009	54	M	Yes	NM	Gitelman's syndrome	Pericardectomy	Improved
Tugcu *et al.* [6]	2008	77	M	Yes	Yes	Narrowing the ascending aorta	Pericardectomy	Improved
Godoy *et al.* [7]	2007	55	M	NM	Yes	Voluminous ascitis	Pericardectomy	Improved
Yamauchi *et al.* [8]	2007	58	F	Yes	Yes	Large mediastinal tumor	Pericardectomy	Improved
Al-Sarraf *et al.* [9]	2007	73	M	Yes	NM	Pericardial hematoma	Pericardectomy	Death
Akhter *et al.* [10]	2006	40	F	No	Yes	Thorough physical examination	Pericardectomy	Improved
Barbieri *et al.* [11]	2004	28	M	NM	Yes	Transient, acute, healthy young man	Nonsteroidal anti-inflammatory drugs	Recovery
Chien *et al.* [12]	2003	16	M	Yes	Yes	Pulmonary stenosis	Pericardectomy	Recovery
Lim *et al.* [13]	2002	46	F	Yes	Yes	Recurrent cardiac failure	Pericardectomy	Improved

M, male; F, female; NM, not mentioned.

Laboratory examinations investigating blood sedimentation, immune indices, autoantibodies, blood biochemical indices, brain natriuretic peptide, and C-reactive protein were performed, and the results were all normal.

Echocardiography showed moderate bi-atrial enlargement (the left atrium was 51 mm × 53 mm, and the right atrium was 58 mm × 48 mm). The ventricles were deformed; the intermediate part of the left and right ventricles was narrower than the cardiac apex (Fig. 1A). Mitral inflow with early diastolic velocity by Doppler did not show any respiratory change based on the AF. The left ventricular ejection fraction (LVEF) was 61%. The quantitative assessment of longitudinal mitral annular motion by tissue Doppler imaging was normal.

CP was suspected based on the above information. The patient then underwent magnetic resonance imaging which showed a narrow, tubular deformation of the lower biventricular that coincided with the echocardiography findings. The pericardium thickness was normal, and calcification was not observed (Fig. 2A). The patient was referred for pericardiectomy, and was provided with aspirin, bisoprolol, hydrochlorothiazide, and spironolactone, which abated his stuffiness and edema symptoms. He refused surgery and was discharged from the hospital.

After 10 months of treatment with diuretics, the patient developed dyspnea after 20 to 30 feet of walking, which indicated the progression of right-sided heart failure, and he returned to hospital for further diagnosis and treatment. Echocardiography showed diastolic interventricular septal bounce. The left atrium was 48 mm × 49 mm, the right atrium was 64 mm × 46 mm, and the LVEF was 51%. Computed tomography (CT) revealed that the ventricles

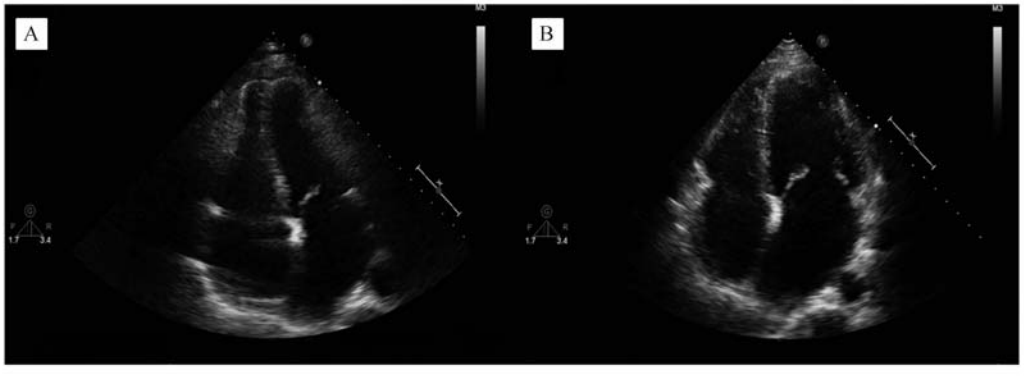

Fig. 1 (A) Echocardiography showed biatrial enlargement. The ventricles were deformed; the intermediate part of left and right ventricles was narrower than the cardiac apex. (B) The shape of the ventricles was almost normal after pericardiectomy.

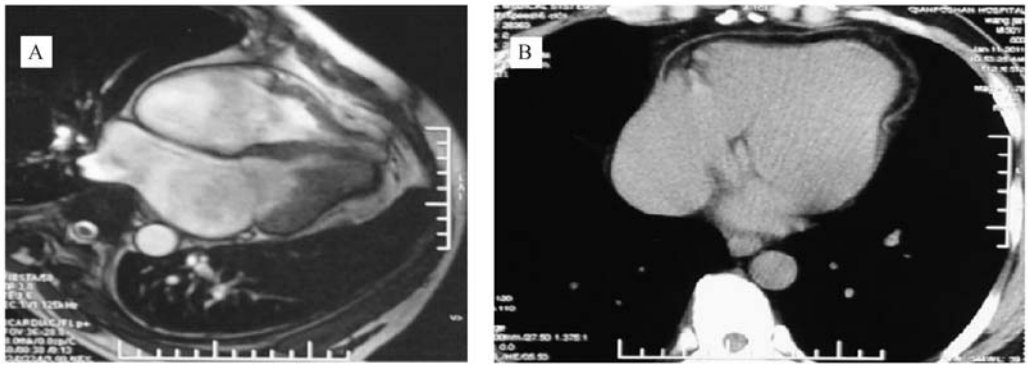

Fig. 2 (A) MRI showed a narrow, tubular deformation of the lower bi-ventricular that coincided with the echocardiography findings. The thickness of the pericardium was normal and calcification was not observed. (B) CT revealed gourd-shaped ventricles. The pericardium thickness was normal. CT, computed tomography; MRI, magnetic resonance imaging.

had a gourd-like shape (Fig. 2B). Pericardiectomy was performed via a median sternotomy over the anterior surface of the left and right ventricles, and as far as the phrenic bundle. The parietal pericardium, with 2.0-mm thickness, was easily removed from the visceral pericardium without calcification. The visceral pericardium was thin, and was closely combined with the epicardium in the lower third of both the left and right ventricles, resulting in a constrictive band. After stripping the pericardium, the central venous pressure was immediately reduced by half.

Biopsy studies of the pericardial tissue gave no evidence of its etiology. The pathological diagnosis was "fibrotic pericardium with some hyaline degeneration." At the 3, 6, 12-month follow-up appointments, symptoms of heart failure had disappeared, though AF still remained. On the other hand, the quality of life of the patient clearly improved. The echocardiogram revealed an almost normally shaped heart (Fig. 1B).

Discussion

The unique aspects of this case were that only the visceral pericardium was locally constricted, obvious calcification was not observed, and the parietal pericardium was of normal thickness, which prevented early diagnosis of CP. Zacharaki *et al.* [14] reported a case of localized CP mimicking the cardiac apex diverticulum that was similar to our case, however, his patient had severe pericardial calcification and thickened pericardium. In addition, echocardiography revealed that the heart was almost normal in shape after pericardiectomy. Therefore, heart deformation was due to constriction, and not caused by a congenital defect.

Classic CP is characterized by fibrous scarring and adhesion of both the visceral and parietal pericardia, leading to loss of pericardial cavity. The thickened pericardium severely restricts cardiac filling. Pericardial thickness is generally increased in CP; however, normal pericardial thickness does not exclude CP during diagnosis, because inelastic fibrous visceral pericardium can physiologically constrict the heart without a demonstrable increase in thickness [15]. Talreja *et al.* [15] reported that a total of 143 patients with proven constriction underwent pericardiectomy at the Mayo Clinic in Minnesota between 1993 and 1999. The pericardium was of normal thickness ($\leqslant$ 2 mm) in 26 patients (18%), and was thickened ($>$ 2 mm) in 117 patients (82%). The symptoms observed between the two patient groups differed very little. In addition, pericardiectomy was equally effective in both patient groups in terms of relieving symptoms. Pericardiectomy is a relatively safe procedure with an average perioperative mortality rate of 6%, and patients with idiopathic CP showed the best results, with a reported survival rate of 88% in 7 years [16]. Compared with routing coronary artery bypass graft and aortic valve replacement surgery, CP resection may carry a moderate surgical risk. However, previous reports have also shown that pericardiectomy is the best choice for idiopathic CP [2,5–10,12,13]. The patient's symptoms generally improve within 6–12 months after pericardiectomy. Szabó *et al.* [17] reported that idiopathic CP was associated with increased survival rates after pericardiectomy. Pericardiectomy for idiopathic CP is safer than CP with other causes, such as prior cardiac surgery, tuberculosis, radiation treatment, etc. However, if the preoperative New York Heart Association (NYHA) classification is IV, the prognosis is unfavorable. Khandaker *et al.* [18] also discovered that if medical management fails patients with chronic pericarditis, surgical pericardiectomy can safely relieve symptoms.

Echocardiography can provide important information for the diagnosis and differentiation of CP from restrictive cardiomyopathy. The three basic signs of CP are as follows [19]. The septal notch shows a sudden shift in the ventricular septal position caused by the asymmetry

between the right and left ventricular filling; ventricular septal shift occurs with respiration; and moderate biatrial enlargement is present (whereas severe enlargement is more likely due to restrictive cardiomyopathy). In our case, the patient met two of the three conditions, but did not have ventricular septal shift with respiration cause by AF. The patient also had an irregularly shaped heart, which was revealed by magnetic resonance imaging and CT imaging. Tissue Doppler imaging (TDI) showed a prominent Ea-wave (peak early velocity of longitudinal axis expansion), which would have been significantly lower in restrictive cardiomyopathy [20]. Therefore, Doppler echocardiography should be the initial non-invasive imaging modality of choice for the differential diagnosis of CP. This case serves as a reminder to consider CP in patients with deformed hearts and unexplained right heart failure, and thus, the timely investigation and treatment can be initiated.

Conclusions

Our case demonstrates that while CP cannot be cured through medical treatment, pericardiectomy is effective. In this case, the patient received 10 months of conservative treatment, including diuresis and anticoagulation, which reduced the lower limb swelling but did not stop the progression of right-sided heart failure. When medical management has failed to improve the right ventricular failure, surgery should be considered in idiopathic CP. With 6% mortality, the clinician should consider the risks versus the benefit to a particular patient.

Compliance with ethics guidelines

Qingqiang Ni, Lin Yun, Rui Xu, Guohua Li, Yucai Yao, and Jiamin Li confirm that there are no conflicts of interest. The local Ethics Committee approved the study, and the patient gave written informed consent prior to participation.

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