Constrictive Pericarditis Presented by Generalized Edema (Anasarca)

Shahar Zimand MD[1], Patricia Benjamin[1], Mira Frand MD[1], David Mishaly MD[2] and Julius Hegesh MD[1]

[1]Cardiac Institute, Department of Pediatric Cardiology and [2]Department of Pediatric Cardiovascular Surgery, Sheba Medical Center, Tel-Hashomer and Sackler Faculty of Medicine, Tel Aviv University, Israel

Key words: constrictive pericarditis, restrictive cardiomyopathy, atrial natriuretic factor

IMAJ 2000;2:316–317

Constrictive pericarditis should be suspected in any patient with jugular venous distention, general edema (anasarca) and pleural effusion. Echocardiography, computerized tomography or magnetic resonance imaging, and angiography should be performed to confirm the diagnosis. Due to the vague and insidious nature of the symptoms, constrictive pericarditis is often mistaken for primary hepatic disease, interabdominal malignancy, or nephrotic syndrome. In the past tuberculosis was the leading cause of constrictive pericarditis, but today the most common source of the sub-acute or chronic form of the disease is idiopathic or viral. This unique complication of simple viral disease should be familiar to any pediatrician investigating anasarca of unknown origin.

Case Description

A previously healthy 2-year-old boy was admitted for tachypnea, dyspnea and anasarca. Four weeks prior to his admission he had a right bronchopneumonia for which he was treated with erythromycin. This was accompanied by progressive tachypnea, weakness and general deterioration. Physical examination revealed a pale, sick-looking child with tachypnea, dyspnea, cutis marmurata, pedal edema, and ascitis with hepatomegaly (7 cm) but no splenomegaly. The heart sounds were normal, ventilation was limited with dullness in both lung bases, and the jugular veins were distended. Chest X-ray revealed bilateral pleural effusion, and the electrocardiogram showed low voltage complexes. Blood examinations for renal and liver functions were normal, and serological tests for common bacteria and viruses including tuberculosis were negative. Echocardiography showed the superior and inferior vena cavae to be dilated without pericardial fluid, thickening or calcification. Cardiac catheterization demonstrated the classical findings of constrictive pericarditis: namely elevation of the vena cavae pressure, a characteristic M (or W) configuration of the right atrium pressure curve, and equilibrium of left and right ventricular end-diastolic pressure with a dip and plateau component of the ventricular wave forms.

A mid-sternotomy performed several days later revealed a thickened and edematous pericardium with involvement of the epicardium in a symmetric fashion. The pericardium was painstakingly removed from the right atrium and both ventricles, with immediate improvement in the ventricular expansion. The postoperative course was uneventful; the child returned to normal life and is free of symptoms.

Comment

Tuberculosis was previously the leading cause of constrictive pericarditis. Today, however, the etiology of the sub-acute or chronic form of the condition is idiopathic or viral, although post-radiation therapy and post-thoracotomy for heart surgery have also been considered the cause of the disease [1]. Less common sources include post-infectious pericarditis [2], chronic renal failure, neoplastic involvement of the pericard, connective tissue disorders [1], and mulibrey nanism – a rare autosomal recessive condition.

Clinically, constrictive pericarditis may mimic hepatic disorders, nephrotic syndrome and other cardiac diastolic dysfunctions [3]. Advanced tuberculous constrictive pericarditis is associated commonly with dense pericardial calcifications and has been termed "Concretia cordis" in Latin and “Panzerherz” in the German literature [1]. Nonetheless, the main diagnostic challenge is to discriminate between constrictive pericarditis and restrictive cardiomyopathy [1,3]. The main etiology for restrictive cardiomyopathy includes idiopathic factors, metabolic storage disease, amyloidosis and hemochromatosis.

The difficulty in distinguishing between constrictive pericarditis and restrictive cardiomyopathy has been the subject of numerous reviews. Vaitkus et al. [3] developed a clinical algorithm to apply when either disease is suspected. Their algorithm considered three well-known hemodynamic criteria: a) equalization (difference <5 mmHg) of right and left end-diastolic pressure; b) restrictive cardiomyopa-

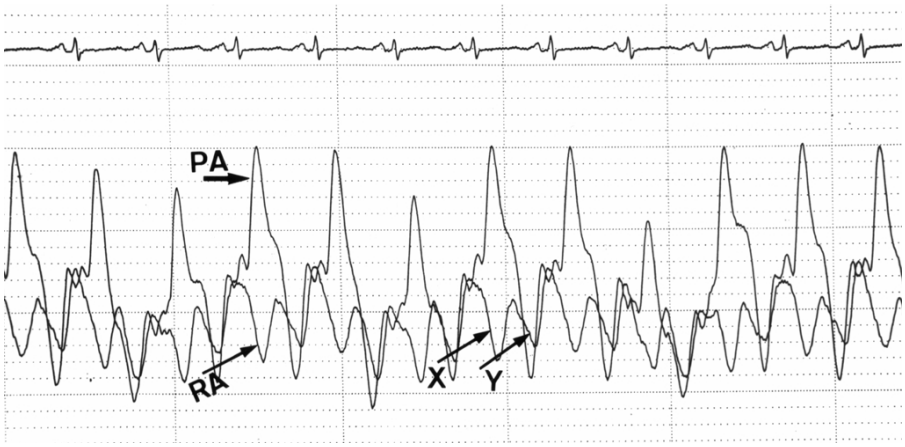

Figure 1. Recording of simultaneous right atrial (RA) and pulmonary artery (PA) pressures in a 2-year-old patient with constrictive pericarditis, illustrating that both are elevated throughout diastole. The prominent X and Y descents give the right atrial wave form the characteristic M (or W) shaped appearance typical of constrictive pericarditis. Note the prominent Y descent, which indicates that right atrial emptying is rapid and unimpeded in early diastole. Maximal PA systolic pressure is 40 mmHg and maximal RA pressure (a wave) 25 mmHg.

thy is more often associated with elevation of right ventricular systolic pressure above 50 mmHg compared to the modest elevation in constrictive pericarditis; and c) constriction is associated with elevation of the ratio RV end-diastolic pressure to RV systolic pressure above 1:3, whereas this ratio is characteristically less than 1:3 in restrictive cardiomyopathy.

Pericardial imaging by MRI and CT are very useful for demonstrating pericardial thickening and small calcifications that may be missed by echocardiography. The recognition of a thickened pericard with or without calcifications, together with appropriate hemodynamic criteria, are very sensitive and specific for constrictive pericarditis [3] and no other evaluation is required to perform a surgical pericardiotomy.

In selected cases, endomyocardial biopsy with the finding of amyloid heart disease is useful to avoid unnecessary thoracotomy [4]. Myocarditis in this instance must be considered a non-specific finding since the inflammatory process may involve the pericardium as well.

The pathogenesis of sodium and water retention in constrictive pericarditis is not well understood; it may differ from the pathogenesis of fluid accumulation in a patient with congestive heart failure due to myocardial disease. The clinical features of constrictive pericarditis reflect the gradual and often insidious development of systemic and pulmonary venous hypertension. In moderate elevation of right atrial pressure (10–18 mmHg), symptoms and signs of systemic venous congestion predominate. These include leg edema, hepatic congestion and ascites [1]. As right and left heart filling pressures become elevated (18–30 mmHg), dyspnea, orthopnea and pleural effusion develop [1]. Some authors state that restricted distensibility of the atria in constrictive pericarditis limits the secretion of atrial natriuretic factor and thus reduces its natriuretic and diuretic effect. Other investigators [5] contend that atrial enlargement is part of the new hemodynamic balance, secondary to preload elevation and diastolic dysfunction. There is evidence that down-regulation of atrial natriuretic factor receptors may be the cause of massive fluid retention.

Although the exact mechanism of fluid retention is still unclear, our case described here demonstrates that constrictive pericarditis is a life-threatening condition that may rapidly complicate a simple viral (or bacterial) infection at any age. In view of the fact that the condition is not always detected by echocardiography alone, we believe that all pediatricians should be familiar with this unique clinical presentation as well as its update of diagnostic criteria. In selected cases, CT, MRI and cardiac catheterization with or without endomyocardial biopsy are necessary to establish the diagnosis.

References

1. Lorel BH, Grossman W. Profiles in constrictive pericarditis, restrictive cardiomyopathy and cardiac tamponade. In: Cardiac Catheterization, Angiography and Intervention. 4th edition Philadelphia: Lea & Febiger, 1991: 633–53.
2. Dupuis C, Gronnier P, Kachaner J, Farru O, Hernandez I, Ducoulombier H, Vliers A. Bacterial pericarditis in infancy and childhood. *Am J Cardiol* 1994;74 8):807–9.
3. Vaitkus PT, Kussmaul WG. Constrictive pericarditis versus restrictive cardiomypathy: a reappraisal and update of diagnostic criteria. *Am Heart J* 1991;22(5):1431–41.
4. Schoenfeld MH, Supple EW, Dec GW Jr, Fallon JT, Palacios IF. Restrictive cardiomyopathy versus constrictive pericarditis: role of endomyocardial biopsy in avoiding unnecessary thoracotomy. *Circulation* 1987;75(5):1012–7.
5. Mantri RR, Bajaj R, Radhakrishnan S, Sinha N, Bidwai PS. Pathogenesis of edema in constrictive pericarditis. *Circulation* 1992;85(4):1634–5.

Correspondence: Dr. S. Zimand, Cardiac Institute, Dept. of Pediatric Cardiology, Sheba Medical Center, Tel-Hashomer 52621, Israel. Tel: (972-3) 530 3123; Fax: (972-3) 530 3127; email: zimandsh@netvision.net.il.

RV = right ventricular