Unusual association of diseases/symptoms

Constrictive pericarditis with a calcified pericardial band at the level of left ventricle causing mid-ventricular obstruction

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Correspondence to Dr Mani Prasad Gautam, manigautam@hotmail.com**Summary**

An adolescent presented with insidious onset and gradually progressive distension of abdomen associated with bilateral ankle swelling of few months duration. He had one episode of prolonged low-grade self-limiting febrile illness during childhood but had not consulted to doctor and never had been diagnosed as case of tuberculosis or acute pericarditis. A detail clinical evaluation showed raised central venous pressure, ascites and ankle oedema. Systemic examination was not much informative except ejection systolic murmur in third left intercostal space. Echocardiography and CT scan heart showed localised thickened pericardium with calcific band around the left ventricle at mid ventricle level. The band around the heart caused the heart to have a 'dumbbell' appearance with ballooning in apical area and a rare mid-ventricular obstruction in the left. A diagnosis of chronic constrictive pericarditis with calcific band was made and the patient was referred to another centre for cardiac surgery.

BACKGROUND

Constrictive pericarditis is not a primary condition. It is the end stage of an inflammatory process involving the pericardium. Virtually any inflammatory process, infectious or non-infectious can lead to constriction. Idiopathic, postsurgical, or radiation injury are the most common causes in developed nations,^{1–3} whereas, tuberculosis was the most common cause of constrictive pericarditis in the developed world before development of effective drug therapy and it remains important in developing countries till date. After an initial insult of acute pericarditis, constrictive pericarditis can develop as early as of few weeks to as long as of few years. The end result is dense fibrosis, often calcification, and adhesions of the parietal and visceral pericardium.⁴ Usually scarring is more or less symmetrical and impedes filling of all heart chambers. Occasionally, localised progressive scarring gives rise to pericardial band and can lead to selective impairment of filling of particular chambers. Reduced cardiac output is a consequence of impaired ventricular filling and causes fatigue, muscle wasting and weight loss. In 'pure' constriction, contractile function is preserved, although ejection fraction can be reduced as a consequence of reduced preload.⁵ The myocardium is occasionally involved in the chronic inflammation and fibrosis, leading to true contractile dysfunction that can at times be quite severe. The latter predicts a poor response to pericardiectomy. In a subset of patients, the process develops relatively rapidly and is reversible. This variant is seen most commonly following cardiac surgery.⁶ The clinical presentation is usually dominated by signs and symptoms of right heart failure. Localised constrictive pericarditis is rare, but occasionally a localised band constricts the inflow or outflow region of one or more of the cardiac chambers. The clinical picture then might simulates valve disease or venous obstruction. Constrictive pericarditis is an uncommon and progressive

but potentially curable pathology. With the exception of patients with transient constrictive pericarditis, surgical pericardiectomy is the only definitive treatment.

Here, we report a case of a patient presenting with constrictive pericarditis, with an accompanying tight constrictive band at the level of midventricle causing left mid-ventricular obstruction along with the calcification of pericardium and the atrioventricular (AV) groove, resulting in a dumbbell-shaped heart with apical ballooning.

CASE PRESENTATION

A young man in his first decade of life was admitted to our hospital with insidious onset and gradually progressive distension of abdomen first noticed 4 months

Figure 1 Chest x-ray- right lateral view showing calcification of pericardium, seen in lower right corner area (arrow).

ago. He had also noticed insidious onset breathlessness initially worsened by exertion and for last 1 year, his breathlessness became persistent. Subsequently in due course of time, he also noticed palpitation, easy fatigability and pitting pedal oedema. His history of illness was significant for a hospital admission for a prolonged febrile illness associated with cough at the age of 5 years, for which he was managed conservatively. Four years ago, he had developed sudden onset and rapidly progressive breathlessness and was managed with a course of diuretic therapy. Since then he had noticed gradually progressive distension of abdomen and exertional dyspnoea which progressed to dyspnoea on minimal exertion for last few months.

On examination, he appeared pallor and undernourished. He had pitting pedal oedema. The pulse rate was 76 beats/min, regular and of low volume. Pulsus paradoxus was absent and there was no pericardial knock. An examination of precordium and chest was unremarkable. Per abdomen examination revealed mild tender hepatomegaly, mild ascites and non-tender mild splenomegaly.

INVESTIGATIONS

A chest x-ray lateral view showed calcification of the pericardium (figure 1). A transthoracic echocardiogram showed a thickened and calcific pericardium (10 mm) causing constriction distal to AV groove. The left ventricular (LV) posterior wall showed sudden diastolic cessation with septal bounce. The inferior vena cava was dilated, with absent inspiratory collapse. There was mid-ventricular obstruction in the left ventricle with ballooning in the

apical area causing dumbbell shaped appearance of left ventricle (figure 2).

A cardiac contrast CT study showed changes consistent with constrictive pericarditis. A ring of calcification, encircling the right and left ventricles distal to AV groove was seen at the junction of the basal and mid cavity regions. An altered shape of left ventricle cavity was seen, the heart had a dumbbell shape with a tight constricting band seen around and immediately distal to the AV groove (posteriorly and inferiorly) (figures 3–5). The calcification was also noticed at the level of AV groove better seen in figure 2. The computer generated 3D CT image also confirms the dumbbell appearance of LV apex (figure 5).

DISCUSSION

First described as ‘concretio cordis’ 300 years ago, chronic calcific pericarditis mostly presents with symptoms and signs of right heart failure. Rarely, it can present with pericardial band causing localised constriction, and if such present, the constriction usually occurs at the AV level.⁷ Similar to constrictive pericarditis, constrictive pericarditis with pericardial band is a secondary condition preceded by an episode of acute pericarditis and time interval between these conditions may vary considerably. Acute pericarditis subsides with subsequent organisation and localised fibrous scarring of the pericardium which results in obliteration of the pericardial space and constriction of cardiac chambers. Depending on the location of pericardial constriction, clinical presentation of localised constriction may be variable including obstruction of right ventricular outflow tract and pulmonary stenosis. Usually there is paucity

Figure 2 2D echocardiography apical 4-chamber view showing calcifications (arrows) and left ventricular mid-ventricular obstruction (arrow).

Figure 3 Plain CT scans of chest with soft tissue window showing calcified pericardial band and pericardium.

Figure 4 Contrast CT scans of chest with soft tissue window showing calcified pericardial band causing left ventricular apical ballooning.

of clinical symptoms in such cases and diagnosis is made during echocardiographic study.

Among pericarditis, generalised constrictive pericarditis is a usual rule but the localised form is rare.^{8–12} Localised pericardial band has been reported after previous pericardiectomy^{8 11} and along with congenital heart disease.⁹ The most common manifestation reported with localised constrictive band involves the AV groove, along with obstruction to the right ventricular outflow tract.^{9 12} Shetty *et al* had reported a case of idiopathic isolated visceral pericardial calcification at the level of right ventricle

(RV) and its outflow tract.¹⁰ In contrast to these reported patterns, our patient had two unique features. First it had calcified constrictive band at the mid-ventricular level distal to AV groove and also extensive calcification in pericardium and AV groove. LV mid-ventricular obstruction was another rare feature recorded in our case. Although, the reason has not been discussed, localised constrictions are most common around the AV groove posteriorly and over the infundibulum anteriorly. It is believed that the AV groove and RV body–infundibulum junction is the transition gutter-like area, relatively fixed and less

Figure 5 Computer generated 3D image of the heart showing ballooning of left ventricular apex (posterior view).

moving structures between the atria and infundibulum and the dynamic ventricles. This would logically result in stasis of an organising collection in this particular region and this collection, over years, is replaced with cicatrix fibrocalcific tissue. The extensive calcification is the most notable characteristic of chronic constrictive pericarditis.¹¹ The most commonly described RV mid-ventricular obstruction is caused by constriction due to fibrocalcific tissue band to relatively less muscular RV. However, similar constriction to LV is less common in view of more muscular LV chamber and moreover, the clinical manifestation of such constriction is rare. Usually the calcification does not necessarily cause clinical constriction. In our case, the LV mid-ventricular obstruction can be explained by the localised extensive fibrosis and calcification of the pericardium which was sufficient to cause echocardiographic features of mid-ventricular obstruction. However, the specific features of constriction such as exaggerated respiratory variation in mitral and tricuspid inflow, exaggerated septal bounce during respiration, and premature pulmonic valve opening were absent in our case.

Constrictive pericarditis is a potentially curable chronic pericardial disease. Whether antituberculous treatment prevents constrictive pericarditis is debatable.¹³ Total pericardiectomy is the treatment of choice for constrictive pericarditis and has good long-term results.¹⁴ In high risk subjects, subtotal pericardiectomy can also be done with satisfactory result. Our patient was referred for pericardiectomy in another centre where total pericardiectomy was done. The detail investigations in our subjects did not reveal any evidence of active or healed tuberculosis except that from history of febrile illness which we believe could be that of primary tuberculosis and had caused cicatrization that led to chronic constrictive pericarditis.

Learning points

- Chronic constrictive pericarditis can have subtle manifestation, high degree of suspicion should be kept in cases where ascites is disproportionate that is, ascites is more prominent than ankle oedema and is associated with distended jugular veins.
- Calcific band around ventricles causing mid-ventricular obstruction is a rare condition and usually occurs on right ventricle, left mid-ventricular obstruction as in our subjects, is a rarer phenomenon.
- Management of constrictive pericarditis with pericardial band is surgical and it is a curable condition.

Acknowledgements We would like to acknowledge Professor Rabindra Bhattacharya, Dr Lekhjunga Thapa and Dr Usha Ghimire for valuable suggestions, necessary corrections and revision.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Troughton RW**, Asher CR, Klein AL. Pericarditis. *Lancet* 2004;**363**:717–27.
2. **Hoit BD**. Management of effusive and constrictive pericardial heart disease. *Circulation* 2002;**105**:2939–42.
3. **Little WC**, Freeman GL. Pericardial disease. *Circulation* 2006;**113**:1622–32.
4. **Hoit BD**. Pericardial disease. In: Fuster V, O'Rourke RA, Walsh RA, eds. *Hurst's the Heart*. Twelfth edition. New York: Mc Graw Hill Medical 2008:1951–74.
5. **Lewinter MM**. Pericardial diseases. In: Libby P, Bonow RO, Mann DL, eds. *Braunwald's Heart Disease, a Textbook of Cardiovascular Medicine*. Eighth edition, Philadelphia, PA: Saunders 2008:1842–6.
6. **Haley JH**, Tajik AJ, Danielson GK, et al. Transient constrictive pericarditis: causes and natural history. *J Am Coll Cardiol* 2004;**43**:271–5.
7. **Hamdulay ZA**, Kumar P, Ali M, et al. Localized pericardial constriction resulting in a "dumbbell" heart. *Ann Thorac Surg* 2007;**83**:2222–4.
8. **Vallance PJT**, Gray HH, Oldershaw PJ. Diagnostic features of localised pericardial constriction. *Int j cardiol* 1988;**20**:416–9.

9. **Chein SJ**, Liang CD, Ko SF. Idiopathic calcific constrictive pericarditis causing pulmonary stenosis associated with a ventricular septal defect mimicking tetralogy of fallot. *J clin ultrasound* 2003;**31**:222–5.
10. **Shetty RK**, Vivek G, Dias LS. Calcific visceral pericardial constriction. *BMJ case reports* 2011.
11. **Bhagia ST**, Patel AR, Reul GJ. Coronary obstruction by a calcific pericardial ring. *Ann Thorac Surg* 2002;**74**:595–7.
12. **Nigri A**, Mangieri E, Martuscelli E, *et al*. Pulmonary trunk stenosis due to constriction by a pericardial band. *Am Heart J* 1987;**114**:448–50.
13. **Dwivedi SK**, Rastogi P, Saran RK, *et al*. Antitubercular treatment does not prevent constriction in chronic pericardial effusion of undermined etiology: a randomized trial. *Ind heart J* 1997;**49**:411–4.
14. **Nataf P**, Cacoub P, Dorent R, *et al*. Results of subtotal pericardiectomy for constrictive pericarditis. *Eur J Cardiothorac Surg* 1993;**7**:252–5; discussion 255–6.

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Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Gautam MP, Gautam S, Sogunuru G, Subramanyam G. Constrictive pericarditis with a calcified pericardial band at the level of left ventricle causing mid-ventricular obstruction. *BMJ Case Reports* 2012;10.1136/bcr.09.2011.4743, Published XXX

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