ARMOR HEART IN A 30 YEAR OLD MALE: A CASE REPORT

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Case Presentation

A 30 year old Omani male presented to a district hospital with atypical chest pain. ECG showed low voltage, chest X-ray and chest CT showed pericardial calcification. He was referred to our center for further evaluation. Past medical history revealed blunt chest trauma following a fall in childhood. He did not seek any medical attention at that time. Physical examination revealed markedly elevated JVP with prominent y descent and kussmaul's sign. There was pericardial knock. There was no ascites or pedal edema.

Echocardiogram showed Doppler evidence of constrictive pericarditis with thick calcified pericardium, paradoxical ventricular septal motion with and dilated IVC/hepatic veins with loss of inspiratory collapse. Hemodynamic assessment in the cardiac catheterization laboratory showed RA 25/22/20 mmHg, RV 42/15 mmHg, LV 110/16 mmHg, PCWP 31/21/20 mmHg and PA pressure of 35/18/26 mmHg. There was near equalization of diastolic pressures in all 4 chambers with < 4 mmHg difference along with dip and plateau on LV/RV pressure tracings. LV angiography showed dense circumferential pericardial calcification (Fig.1-4), normal LV systolic function and normal coronary arteries. The patient was offered surgical intervention but he refused.

Discussion

Pericardial calcification (PC) with or without evidence of constrictive pericarditis (CP) is usually preceded by an episode of pericarditis or trauma. Infectious etiologies for pericarditis include viral agents, pyogenic, tuberculosis and histoplasmosis. Pericardial disease from radiation, previous infarction, uremia, systemic lupus erythematosis, rheumatic fever, pericardial tumors, asbestos exposure and hemopericardium (post trauma or cardiac surgery) can result in pericardial calcification. The most frequent causes of pericardial calcification are chronic idiopathic pericarditis, post cardiac surgery, mediastinal irradiation, and tuberculous pericarditis[1,2].

Blunt and penetrating chest wall trauma, although uncommon, has also been reported to cause CP and PC, presumably through an inflammatory, immunopathic and healing mechanism. Blunt trauma to the chest follows nonpenetrating thoracic impacts such as after falls, compression (crush) and blast, etc.

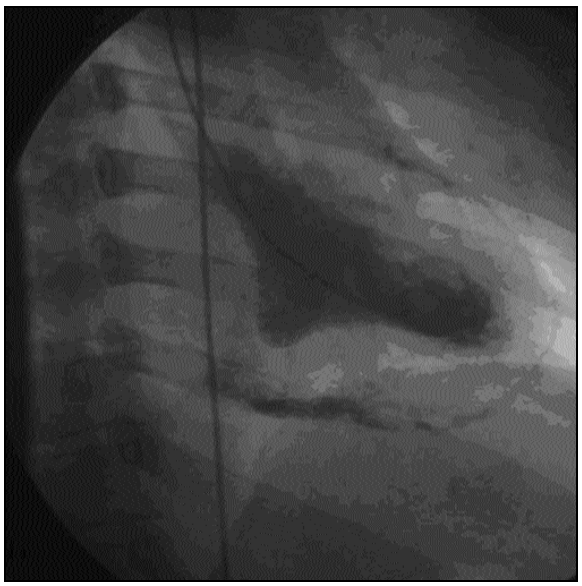


Fig. 1: *LV gram in systole.*

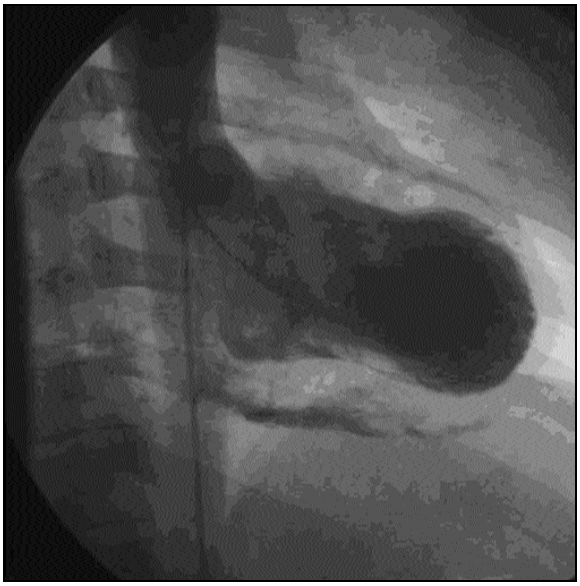


Fig. 2: *LV gram in diastole.*

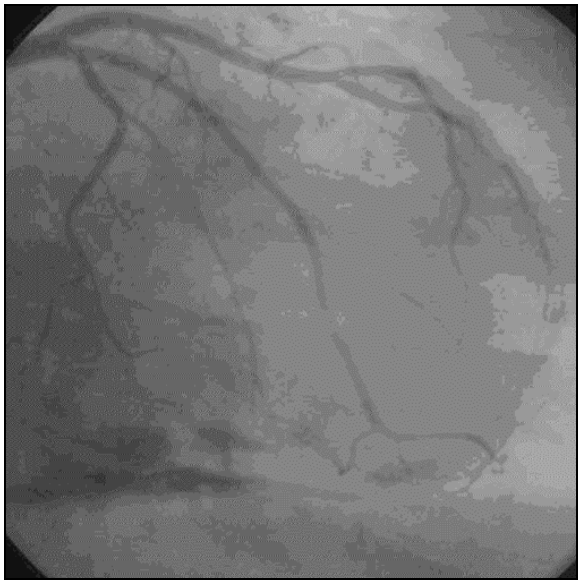


Fig. 3: *LCA injection.*

Fig. 4: *RCA injection.*

Pericardial laceration causing parietal pericardial tears are frequent, especially after falls. Other deceleration injuries and lacerations are rarely isolated or clinically silent. Symptoms occurring after blunt trauma can be nonspecific or multiple and may be overshadowed by other injuries. It is rare for patients to be asymptomatic for months or years as in our patient.

In early reports from the United States, calcification was observed in approximately 50% of cases of CP. In a recent study[3] of 135 patients with CP, pericardial calcification was seen in 27% of patients and the cause of CP was indeterminate in 67% of patients with pericardial calcification on chest radiography and 21% of patients without calcification. It was also noted that presence of calcification correlated with disease chronicity, atrial enlargement, atrial arrythmias and there was increased perioperative mortality. Long-term outcome in these patients, however, did not differ from those with non-calcific disease. They also noted that calcification was uncommon in patients who developed constriction after cardiac surgery.

In another study[2] of 163 patients with pericardiectomy, 31% had radiographically demonstrated PC which was more commonly associated with idiopathic CP and PC and was not a predictor of overall survival. In another study[4], it was noted that CP can occur with normal pericardial thickness; 18% of surgically proven CP patients had normal pericardial thickness. Constrictive pericarditis develops in 50-70% of patients with pericardial calcification. Extensive calcification may be present without signs or symptoms of CP.

Constrictive Pericarditis is a disease characterized by the encasement of the heart by a sack-like covering with or without calcification (armor) due to a rigid non-pliable pericardium secondary to dense fibrosis and adhesions. This causes impaired diastolic cardiac function leading to heart failure manifested by systemic congestion including ascites and pedal edema. The pathophysiological hall mark of CP is equalization of the end-diastolic pressures in all 4 cardiac chambers.

Dystrophic calcification signifies pericardial injury more destructive than conditions healing without calcification[5]. Yet even extensive calcifications may be well tolerated and asymptomatic. The maximal pericardial calcification occurs predominantly over the right atrium and anterior right ventricle, diaphragmatic surface, atrioventricular grooves and rarely over the LV apex. Fluid preferentially gravitates towards right side of the heart, where calcium and even bone are slowly deposited in the inspissated fluid[2]. Extensive circumferential calcification, which was seen in our patient, is rare.

On chest radiographs, pericardial calcification appears as curvilinear calcification usually affecting the right side of the heart. This is often visualized better on lateral chest radiographs than on frontal views. CT is the best technique to detect pericardial thickening (>4mm) and calcification, however, in cases of over-penetrated films, fluoroscopy, 2D/3D echocardiography with Doppler and MRI may be helpful. TEE is superior to TTE for detecting

pericardial thickening. Catheterization study may help in differentiating constrictive pericarditis from restrictive cardiomyopathy.

Calcifications increase the technical difficulties of pericardial resection and where calcification invades the myocardium, the procedure is particularly difficult and increases failure rates. Perioperative mortality in CP is around 6% and seven- year survival is more than 85%2. Complete relief of symptoms occurs in 50% of survivors; around 10% have persistent symptomatic heart failure. Extensive calcification is a marker for poor postoperative out come.?

Heart Views 2007;8(3)106–108.

References:

1. Pericardial Disease, WC Little, GL Freeman, *Circulation* 2006; 113:1622-1632.
2. Constrictive pericarditis: etiology and cause-specific survival after pericardiectomy, SC Bertog, SK Thambidorai, K Parakh et al, *J Am Coll Cardiology*, 2004; 43:1445-1452.
3. Calcific Constrictive Pericarditis: Is It Still with Us? LH Ling, Jk Oh, JF Breen et al: *Annals of Internal Medicine*, 2000; 132:6:444-450.
4. Constrictive Pericarditis in 26 Patients With Histologically Normal Pericardial Thickness, DR Talreja, WD Edwards, GK Danielson et al, *Circulation*, 2003; 108:1852.
5. Pericardial diseases. In: Braunwald E, ed.Heart Disease.7th ed.

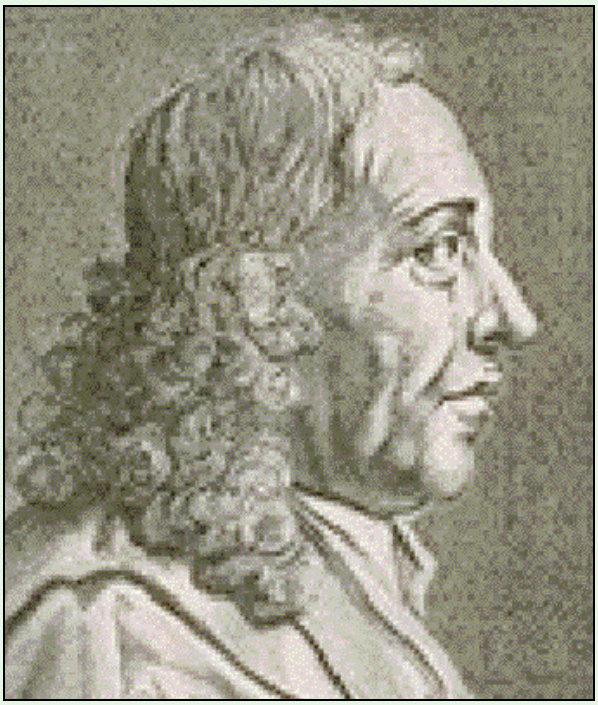
THE FIRST HISTOLOGIST

Malpighi discovered capillaries, a network of tiny thin-walled microtubular anastomosis between the arteries and veins. His discovery supplied the missing link in the circulation of the blood.

He described the vesicular nature of the lung, showing how the divisions of the trachea terminated in the dilated air vesicles, thus supplying the anatomic basis for the true conception of the respiratory process.

He was the first to demonstrate the existence of red corpuscles but failed to interpret them properly. Under the microscope, he observed flat red blood cells in the mesenteric blood vessels of the hedgehog. He mistook the red blood corpuscles as fat globules passing from fatty tissues into the current of the blood. This was later clarified by van Leeuwenhoek in 1674.

Malpighi did much to advance the understanding of the physiology of the liver, spleen and kidneys through his monumental work on the structure of the viscera, "De Viscerum Structura, Exercitato Anatomica" which was published in Bonn in 1666.



Marcello Malpighi (1628-1694)

Malpighi was born in Crevalcuore, near Bologna, Italy. He obtained his doctorate in medicine and philosophy from the University of Bologna at the age of 25. For most of his career, Malpighi combined an intense interest in scientific research with a fond love of teaching. He used the microscope toward biological investigations and became one of the greatest microscopists of all time. Many historians regard Malpighi as the father of microscopic anatomy in both animals and plants.

Other works and discoveries:

- Insects, particularly the silkworm, do not use lungs to breathe, but small holes in their skin called tracheae
- "Rete mucosum", or Malpighian layer of the skin
- Papillae of the tongue are organs of taste
- His treatise, "De polypo cordis" (1666 was important towards understanding how blood clots and its composition.
- He was the first to notice that there were patterns in fingerprints but suggested no practical application.