

A Disease that Constricts my Heart, Drown me in my Own Fluid, and Affected my Kidneys can be Cured by Surgery

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Case History

A 62 year-old Caucasian male, with past medical history of coronary heart disease, type-2 diabetes mellitus, obstructive sleep apnea, and hyperlipidemia, admitted to the hospital with an episode of hypoglycemia and unresponsiveness. He was diagnosed in the past by his gastro-enterologist as having “liver disease”, the nature of which was not clear. His physical examination showed that he was well nourished man with regular heart rate, no edema or jugular venous distension. His serum creatinine was 1.9 mg/dl and had unquantifiable proteinuria in his urine analysis triggered a nephrology consultation. He returned to the nephrology clinic after discharge from the hospital on 12/15/16 with increasing serum creatinine from 1.9 to 2.5 mg/dl and an estimated GFR of 30 ml/min. During this encounter he denies any nausea, vomiting, diarrhea, or use of non-steroidal anti-inflammatory medications (NSAID). In the interim, he stated that he had paracentesis of 17 liters on 12/07/16 for his ascites.

On his follow up visit on 1/31/17 his examination was unremarkable and his serum creatinine had gone down to 1.9 mg/dl. During this time he was diagnosed with liver cirrhosis but had no liver biopsy or more detailed information about the diagnosis. His hepatitis serology was negative and liver

function test were within normal range. He returned to the nephrology clinic on 3/8/17 with increasing serum creatinine to 3.82 mg/dl and blood urea nitrogen (BUN) increasing from 37 to 73 mg/dl. He had a transthoracic echocardiogram which showed left ventricular dysfunction.

The patient returned to the nephrology clinic once again on the 3/17/17, with stable serum creatinine of 3.8 and BUN of 79 mg/dl. But this time he had fluid overloaded with increased edema of the lower extremities and tense ascites. He stated that he had been scheduled for paracentesis and cardiac catheterization on 3/29/17.

On the next visit on 4/24/17, his eGFR had gone down and was classified to CKD stage-4. During this visit he was diagnosed with chronic constrictive pericarditis (CP) on cardiac catheterization. His edema had been controlled with medications, and paracentesis. He was scheduled to have pericardiectomy in the following few days. His serum creatinine had gone up to 4.4 mg/dl. He continued to have paracentesis and hemodialysis to control his edema and renal function. His last paracentesis was on 5/8/17 with 10.7 L removed.

On 5/17 he had surgery on the pericardium with chest tube insertion × 3. During this hospital admission and following surgical procedure his heart function, renal function, and urine output had improved. He was taken off hemodialysis and his final serum creatinine was 1.53 mg/dl before discharge. He had a total of 4 cardiac catheterizations and the diagnosis of CP was clinched on the 3rd catheterization. He had multiple paracentesis with total fluid loss of 111 Lb. and total fluid removal by paracentesis of 21.6 L.

Discussion of the Case

This case illustrated the importance of vanishing arts of clinical history, examination, and analytical data interpretation. The patient presented with edema, ascites, fluid overload, normal hepatitis serology and liver function, and progressive increase in blood urea nitrogen (BUN) and creatinine. He underwent a total of 4 cardiac catheterization before the final clinical diagnosis is attained. The sensitivity of transthoracic echocardiography (TTE) is not high (40%) compared with the

trans-esophageal (TEE) for the diagnosis of valvular diseases, diseases of the myocardium, and pericardium linings [1].

Serology for hepatitis B and C were negative as other liver function tests. A diagnosis of cirrhosis without liver biopsy was given because of ascites. Considerations of pre-hepatic, hepatic and post-hepatic causes of ascites were not analyzed in the context of patient's history and clinical examinations. As a matter of fact very often CP is misdiagnosed as cirrhosis because of hepatomegaly and ascites. On the other hand ultrasound examination of the abdomen in this case revealed no splenomegaly, evidence of fibrosis, or obstruction to the portal or hepatic veins. I am not sure if portal vein and hepatic venous pressures and their variation with respiration were measured during this encounter. The patient was treated symptomatically with diuretics, paracentesis, and eventually hemodialysis to control ascites, peripheral edema, and deteriorated kidney function.

The case is also unique as it presented with increasing BUN and creatinine in the absence of significant proteinuria, making intrinsic renal disease unlikely. The ultra-sound of the kidneys excluded obstruction of the genitourinary tract making pre-renal diseases with congestive nephropathy a more likely scenario. One wonders why repeated cardiac catheterization was performed to achieve the final diagnosis. I do not know whether the right and left cardiac pressures were measured in the first 2 catheterizations or not.

On 5/12/16 both right and left cardiac catheterization was done which showed no coronary artery disease or evidence of CP. His transthoracic echocardiogram was also normal with ejection fraction of 60% which is very commonly encountered in CP. The third cardiac catheterization showed equalization of the pressure in the cardiac chambers which triggered the correct diagnosis. The fourth cardiac catheterization was done for follow up after the surgery. The etiology of the CP remained idiopathic as no cause where found to account for the disease process.

If we trace his serum creatinine as far back as 1/27/14 and 6/26/15 (1.0 and 0.84 mg/dl, respectively), one would find that his serum creatinine were normal to start with and progressively getting worse up until his recent admission and the time of surgery after which relief of congestion and improvement of kidney function began to appear, (Table 1). In the follow up visit 4 months after surgery his kidney function reverted to normal with resolution of ascites and congestion.

Constrictive pericarditis is a scarring process which causes loss of normal elasticity of the pericardial sac. It is typically chronic disease which impedes cardiac filling by the external forces. In the most severe form of CP the pericardium becomes almost inelastic which compromises volume changes during respiration.

In normal situation the normal pericardium does not interfere with the decrease in intra-thoracic pressure during inspiration which leads to increase in venous return to the right side of the heart. This increase in venous return to the right side of the heart does not impair left ventricular filling.

However, in CP cardiac volume is constrained which prevents normal inspiratory decrease in intra-thoracic pressure from being transmitted to the heart chambers. Pulmonary venous pressure decrease during inspiration, however, left ventricular pressure does not. This divergence will results in decreased left ventricular volume as a results of a decrease in trans-pulmonary gradient. These pathophysiological abnormalities lead to shifting of interventricular septum to the left during expansion of the right ventricle further compromising left ventricular end-diastolic volume. In severe CP the ventricular filling occurs in early diastole with little or no filling subsequently [2,3]. As CP becomes more severe the ventricular volumes and stroke volumes are reduced [4-9] (Table 2).

Orthotropic heart transplant and tuberculosis pericarditis have been associated with high incidence of CP (49%) in developing world [10,11]. IgG4 diseases and Whipple's disease have also been incriminated in cases of CP [12,13].

The clinical presentation of CP are protean, and includes volume overload manifested as peripheral edema, ascites and anasarca, low cardiac output with exertional dyspnea, fatigability and signs of heart failure or symptoms of cardiac tamponed.

On physical examination, patients with CP often exhibit raised jugular venous pressure (JVP), pulsus paradoxus [4, 14], Kussmaul's signs [8,9], a pericardial knock [8], edema, ascites, pulsatile liver, and cachexia. JVP is reported in 93% of cases of CP [8]. Unexplained raised JVP in patients with predisposing conditions like malignancy, irradiation, or previous cardiac surgery should alert the physician to the presence of CP.

Evaluation of CP should include chest x-ray which might show calcification. Echocardiography both 2-D and M-mode which can be done as transthoracic or trans-esophageal echocardiography (TTE or TEE). The TEE is more sensitive [1-3,15-20]. CT and cardiac magnetic resonance (CMR) are very sensitive modalities for detection of pericardial thickening and calcification in CP [3,15,21-24]. Hemodynamic evaluation during cardiac catheterization are sometimes needed to confirm the diagnosis of CP [2,3,15,25].

Plasma BNP are elevated in cardiomyopathy and left ventricular dysfunction but less so in CP. The differential diagnoses of CP include cardiac tamponed, restrictive cardiomyopathy, and cirrhosis. Table 3 showed the difference between CP and restrictive cardiomyopathy.

Table 1: Time-line for serum creatinine

Date	5/11/16	12/25/16	1/31/17	3/8/17	3/17/17	4/24/17	5/22/17	7/8/17
Serum creatinine in mg/dl	1.38	2.5	1.9	3.82	3.8	4.4	2.41	1.8

Table 2: The etiology of CP is identified in case series reported by previous workers [4-9]

Idiopathic or viral	42-61%
Post-radiation	20-31%
Connective tissue disorders	3-7%
Post-cardiac surgery	11-37%
Post-infectious	3-15%
Miscellaneous causes (malignancy, Trauma, drug induced, asbestosis, Sarcoidosis, uremic pericarditis)	up to 10%

Table 3: Differences between constrictive pericarditis and restrictive cardiomyopathy

Characteristics	CP	RCMP
History		
- Prior pericarditis	Yes	No
- Cardiac surgery	Yes	No
- Irradiation	Yes	No/Yes
- DM, amyloidosis, sarcoidosis	No	Yes
Physical examination		
- JVP	more common	Yes
- Pericardial knock	Yes	No
- S3	No	Yes
ECG findings		
• BBB	No	Yes
• Ventricular hypertrophy	No	Yes
• Pathological Q wave	No	Yes
• impaired atrio-ventricular conduction	No	Yes
B-type BNP	Usually in normal range	Usually raised
CXR		
- Calcification	Yes	No
- cardiomegaly	No	Yes
Imaging (Doppler, echo, CT, CMR)		
- increased thickens of pericardium or calcification	Yes	No
- Thick ventricular wall or septum	No	Yes
- MR or TR	No	Yes
Doppler		
- Respiratory variation in ventricular filling velocity	High usually in 30-40%	Minimal <10%
- Hepatic venous flow reversal	Expiratory	Inspiratory
- Diastolic MR	No	Yes
Tissue Doppler imaging		
- Early diastolic Doppler tissue velocity @M annulus (E')	Increased >12 cm/sec	Decreased to <8cm/sec
- Doppler myocardial velocity gradient @ LVW in early diastole and during ventricular ejection	Comparable to normal	Significantly lower
Cardiac catheterization		
- Right and left ventricular end-diastolic pressure (RVEDP & LVEDP)	Equal or near equal	LVEDP>RVEDP
Biopsy		
- Endomyocardium or less commonly pericardium	May be helpful	May be helpful

CP- constrictive pericarditis; RCMP- restrictive cardiomyopathy.

Acute kidney injury (AKI) is rare manifestation of CP. The pathophysiology is probably linked to congestive nephropathy with high renal vein pressure and little in the way of proteinuria as illustrated in this case. The high pressures in the renal veins are transmitted back to the glomeruli with decrease in the glomerular filtration rate. Once the pressure is relieved by pericardiectomy the renal function usually reverted to normal.

The treatment of CP is pericardiectomy [26-29]. Striping of as much as possible of the pericardium is advisable to have the gratifying effects of surgery especially if surgery is carried out early in the disease process. In many instants relief of ascites, peripheral edema and congestive hepatopathy and nephropathy would ensue more quickly.

The take home message in this case is never abandoned good clinical history; physical examination looking for signs of

CP especially raised JVP, pulsatile liver, and ascites. The art of history taking and physical examination together with analytical interpretation of the data will sharpen our clinical acumen and in most cases would lead you to the correct diagnosis.

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