ANEURYSMAL DILATATION OF RIGHT SIDE OF THE HEART WITH CHYLOUS ASCITIS

A. THUKRAL, V. SRIVASTAVA, M. MISHRA, R. CHANDRA SHUKLA[a], K. TRIPATHI[1]*[*]
Department of Medicine, Institute of Medical Sciences, Banaras Hindu University Varanasi- 221005
[a]Department of Radiology, Institute of Medical Sciences, Banaras Hindu University Varanasi- 221005

A 30 year old man presented to the emergency with history of breathlessness and abdominal distension for last 8 years. He was on treatment by a general physician for last 8 years. His symptoms had exaggerated in last 3 months. General examination revealed an irregularly irregular pulse with pallor, icterus, pedal edema, JVP was raised and gynaecomastia was present. Massive ascitis was present. Cardiac examination revealed that apex beat was localized in fourth intercostal space, in mid axillary line. Auscultation revealed a pansystolic murmur best heard in third intercostals space in parasternal area. Chest X ray revealed complete opaque left hemi thorax with obliteration of left cardiophrenic angle. As a theureuptic and diagnostic measure a pleural tap was planned but due to good audibility of heart sounds even at the infrascapular area on the left side; a confusion arose regarding the existence of pleural effusion so a HRCT thorax was done which revealed that the whole left hemi thorax was occupied by the aneurysmally dilated right atrium and ventricle, left atria and ventricle appeared normal. A great disaster would have occurred had someone tried a pleural tap. Echocardiography demonstrated dialted right chambers with very thin wall with tricuspid regurgitation and a thrombus in the right atrium. Pulmonary artery and pulmonary valve were normal.Chylous ascitis was also drained. USG abdomen revealed enlarged liver with coarse echo texture. A final diagnosis of lone right sided cardiomyopathy was reached either the arrythmogenic right ventricular dysplasia or the parchment heart syndrome (UHl's disease).

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Case Summary

In March, 2009 a 30 year old male presented to us in the emergency with complaints of abdominal distension, pedal edema and breathlessness for the last 8 years. As stated by the patient he was apparently asymptomatic 8 years back when he started having abdominal distension along with pedal edema. After one month of this he started having symptoms of breathlessness on exertion which gradually progressed and over next 15 days patient started having orthopnea; but this was accompanied by massive abdominal distension and patient ascribed his symptoms of breathlessness and orthopnea to abdominal distension and stated that his symptoms got relieved on drainage of ascitic fluid 8 years back . He was at that time diagnosed as a case of cardiac disease and put on decongestive treatment and digoxin. The exact nature of his cardiac disorder was not known to the patient and his previous records were misplaced. However patient stated that he was taking diuretics and digoxin for the last 8 years uninterrupted; he was apparently asymptomatic 3 months back when he started with the same complaints of progressive abdominal distension with

*Corresponding Author: kamlakar_tripathi@yahoo.co.in

breathlessness and orthopnea. At no point of time patient stated any complaints of joint pains, syncope, cyanosis, chest pain, hemoptysis, hematmesis or malaena. However he did complain of palpitations this time which were not present 8 years back. On basis of history it was evident that the patient's symptoms were consistent with right heart failure with recent onset exaggeration of symptoms.

There was no significant past history of tuberculosis, hypertension or diabetes and no other significant illnesses in his childhood.

General examination revealed mild pallor, icterus was present, JVP was raised, pedal edema was present, grade 1 clubbing was present in bilateral hands and pedal edema was present. Gynaecomastia was also present. There was no cyanosis or lymphadenopathy.

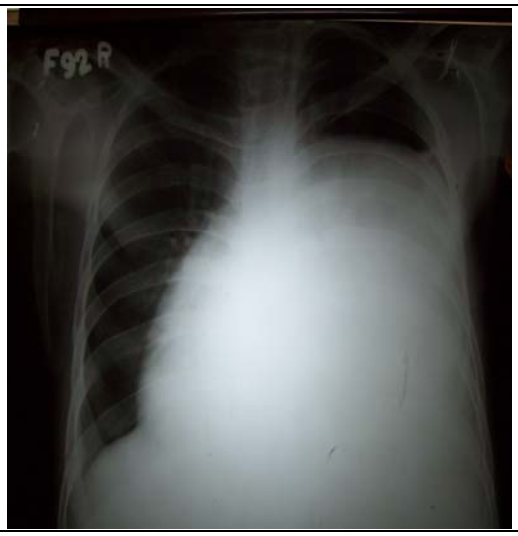
Pulse was irregularly irregular with a heart rate of 40/min. Blood pressure was 110/70 mmHg in right arm supine position.

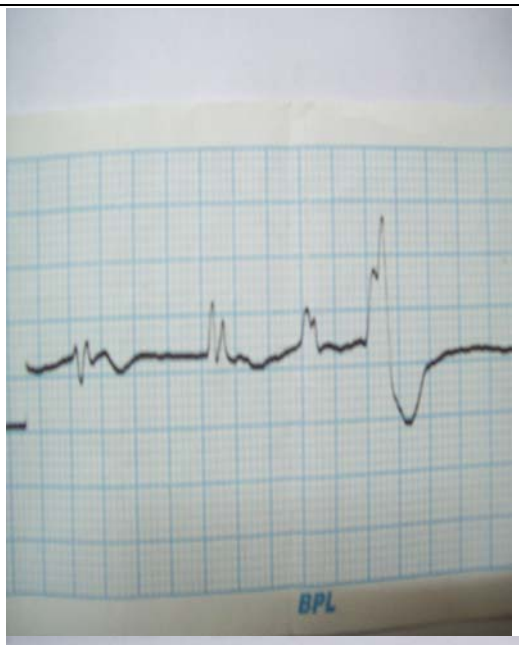
Abdominal examination revealed distended abdomen with venous prominences and everted umbilicus. Ascitis was present, as was evident by fluid thrill; the flow in the veins was away from the umbilicus. Liver was palpable 6cm below the right costal margin in the mid-clavicular line. There was no splenomegaly. On diagnostic tap of the ascitic fluid it revealed a turbid whitish fluid which on analysis was found to be transudative with ether test positive suggesting that it was chylous fluid.

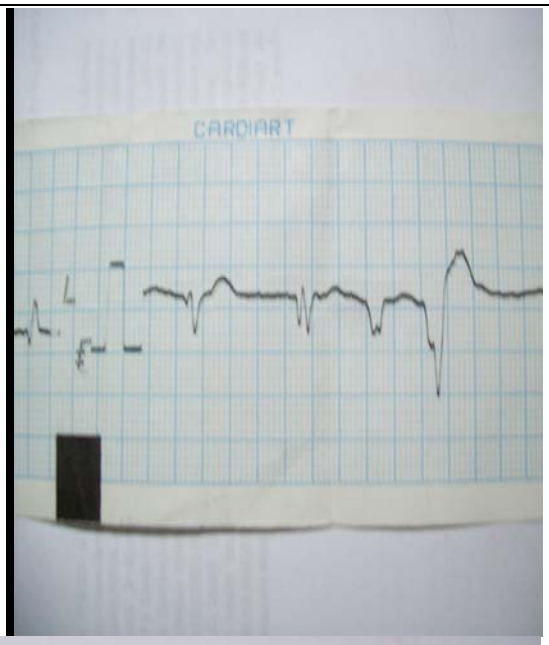
Chest examination revealed prominent veins in the lateral chest wall with direction of flow from below upwards. There was dullness on percussion in lower left hemi thorax with decreased breath sounds in the lower left hemi thorax. This raised a clinical suspicion of it being a pleural effusion; however although the breath sounds were dull but prominent cardiac sounds could be heard even in the lower left hemi thorax in the back. Other wise chest auscultation was normal with no crepts or rhonchi.

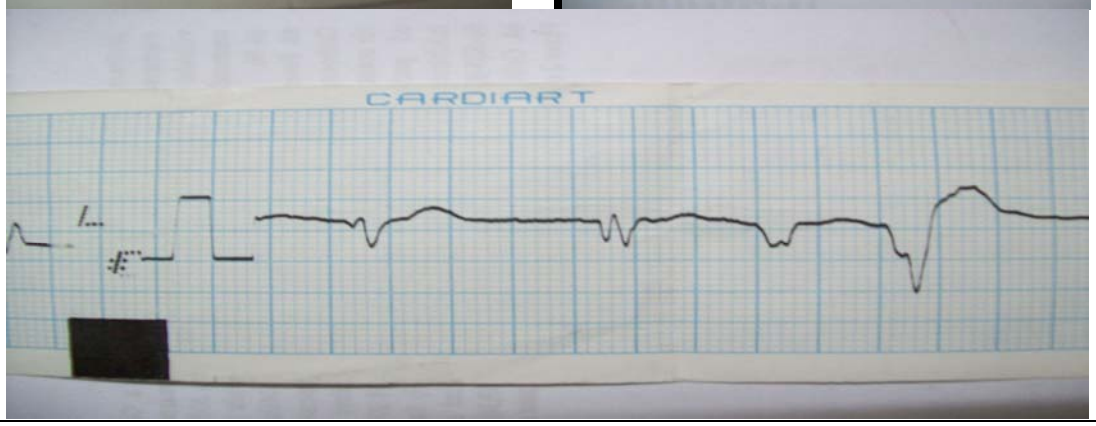
Cardiovascular examination revealed that apex beat was diffuse and could be seen in the 4th intercostals space in the mid axillary line. Palpation confirmed the apex impulse in the mid axillary line in the 4^{th} intercostals space, there was also a systolic thrill in the apex. On auscultation 1^{st} and 2^{nd} heart sounds were loud. P2 was soft. A pansystolic murmer best heard in the 3^{rd} intercostals space in parasternal area was heard radiating towards the axilla but was audible all over the precordium.

A chest X ray revealed a opaque left hemi thorax. ECG showed absent 'p' waves with a triplet of ventricular ectopics repeating at intervals with RBBB pattern. So ECG suggested a possibility of Atrial fibrillation with complete heart block with ventricular ectopic with varying RR interval suggesting a ventricular Wenkebach phenonmenon.



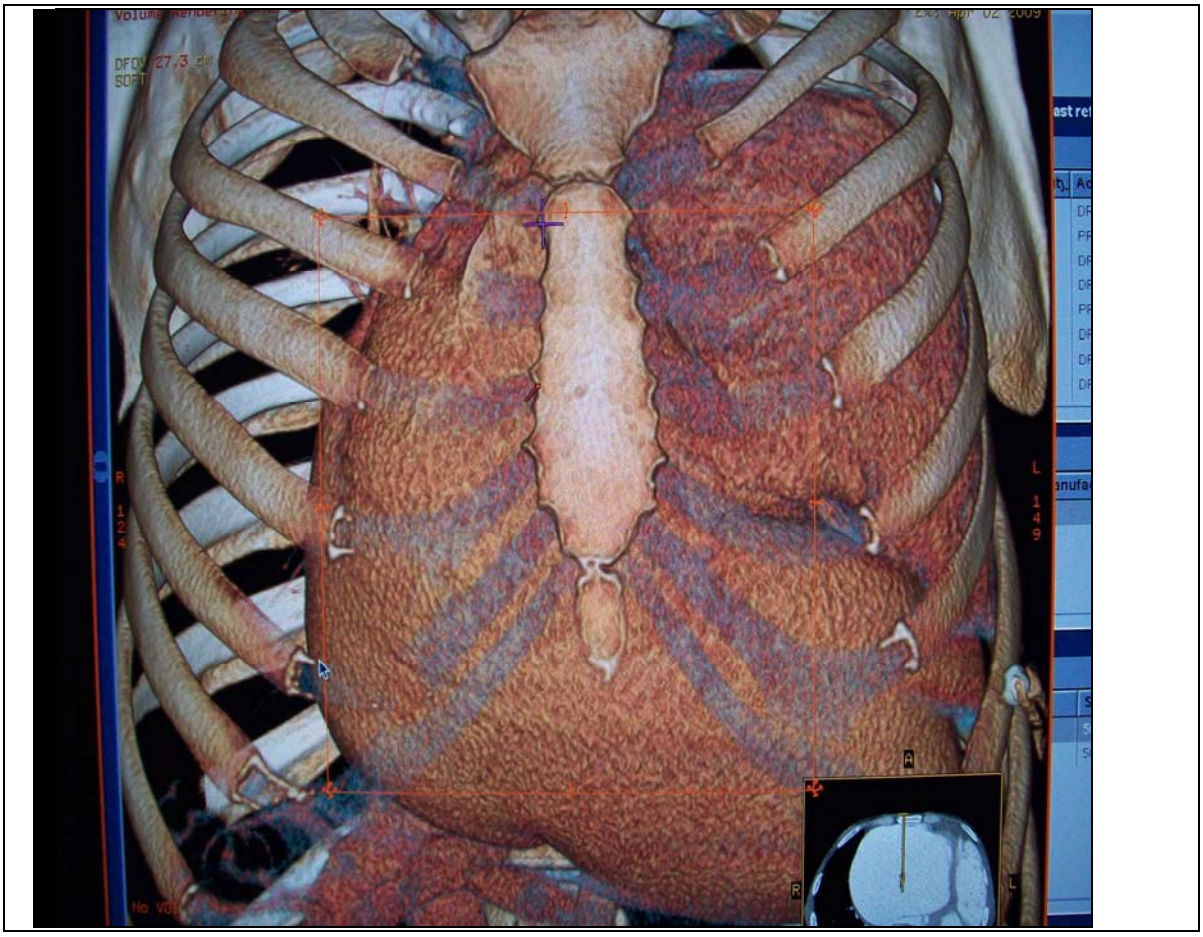






On clinical suspicion of pleural effusion and chest X ray showing a opaque hemi thorax there was a temptation to do a diagnostic pleural tap; but good audibility of heart sounds, a apex beat in mid axillary line and a convex upper border of left opacity with absence of meniscus sign on chest X ray prevented the treating team and a decision of going for a

CT thorax was done which revealed astonishing findings that the entire opacity was hugely dilated right atrium and right ventricle with a compressed left atrium and ventricle.



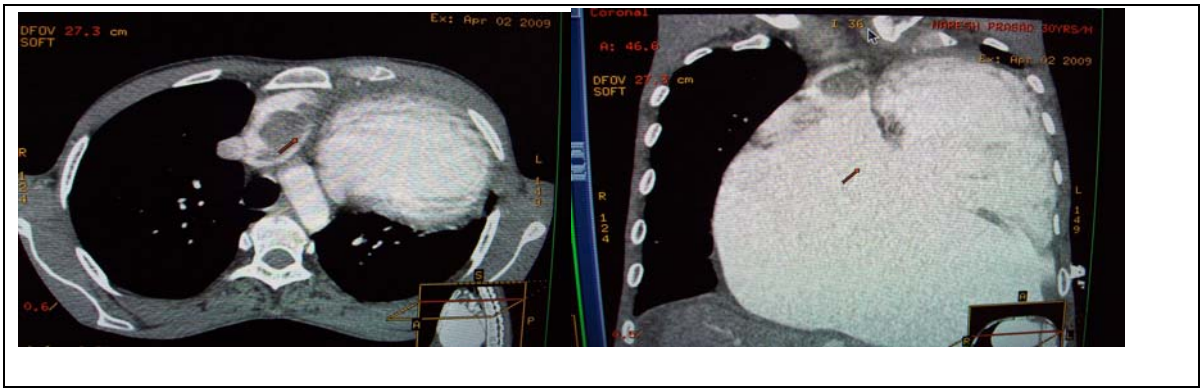
Following this a CT angiography of heart was done which revealed that right atrium was dilated with dimensions of 12 cm (vertical) x 10 cm (AP) with a filling defect of 2.5 cm x 1.9 cm attached to its posterior wall with homogenous attenuation without enhancement s/o thrombus. SVC was normal (1.5 cm) but the IVC was dilated (2.6 cm) with dilated hepatic veins. Right ventricle measured 17 cm x 14 cm with a enlarged pulmonary outflow tract. The wall of the right atrium and ventricle was so thinned out that with a 64 slice CT scan the exact thickness could not be measured.

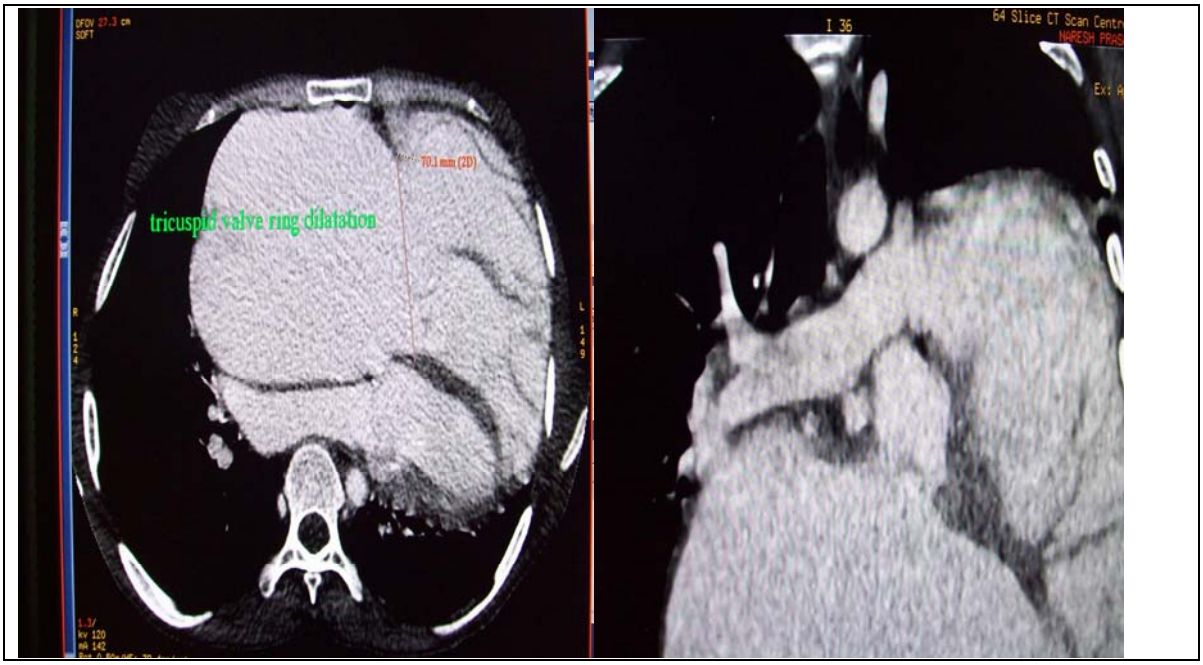
The pulmonary vasculature was normal with pulmonary artery caliber 2 cm at the origin and right descending artery diameter of 1.5 cm. Left atrium and left ventricle were compressed with left atrium measuring 1.5x 3.8 cm and ventricle measuring 4.5 x 8 cm with normal wall thickness of about 6 mm.

Echocardiographic findings confirmed the anatomical findings of CT scan and showed normal left ventricular function with Ejection fraction of 65 % and a severe tricuspid regurgitation and severe compromise of right ventricular systolic function.

CT abdomen was done to rule out any cause of chylous ascitis; which showed altered liver echotexture with gross ascitis.

Other biochemical investigations revealed hemoglobin of 80g/dl, a total bilirubin of 4 mg/dl and direct fraction of 2.5 mg/dl. AST & ALT levels were 200/130 IU respectively. Serum total protein/ albumin were 4.4/2 g/dl. Renal function was within normal limits. A PT INR was done with value of 7.8. These findings suggested cardiac cirrhosis in this patient.





Our patient however expired a 4 days after admission and there was nothing much to offer in such advanced disease patients. A post mortem examination could not be done for lack of consent.

Discussion

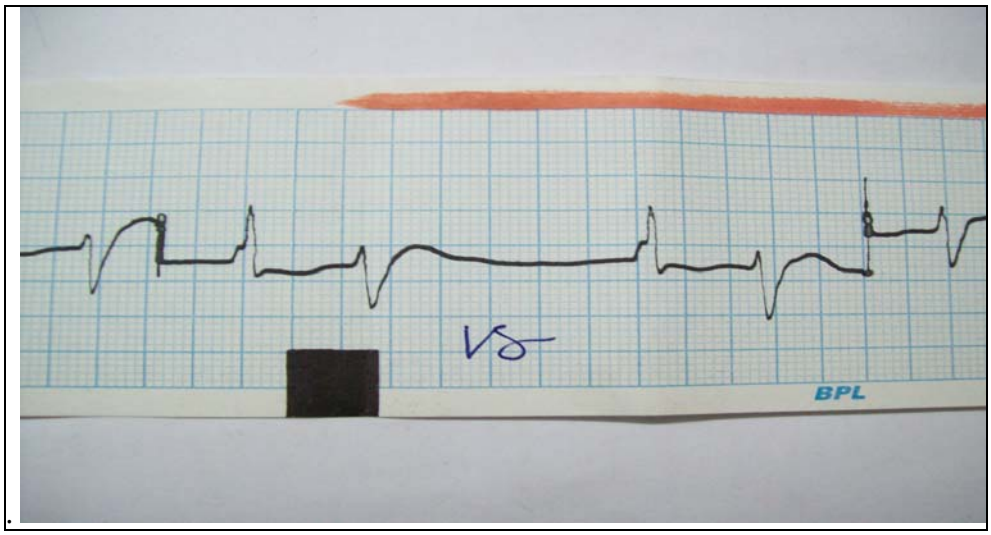
A final diagnosis of lone right sided cardiomyopathy with cardiac cirrhosis; leading to chylous ascitis, and a right atrial thrombosis with arrhythmias was reached. Two types of lone right sided cardiomyopathies are compatible with this presentation arrhythmogenic right ventricular dysplasia (ARVD) and UHl's anomaly (parchment heart syndrome). Right ventricular dysplasia is a muscle disease of unknown cause that is often familial; characterised by adipose or fibroadipose infiltration of the right ventricular myocardium [1]. It is generally regarded as a selective disorder of the right ventricle. It is now often cited as the common cause of sudden death (due to arrhythmias) in young athletes in countries where screening in athletes for hypertrophic cardiomyopathy has begun; especially high incidence from Italy.

The ECG findings commonly include a RBBB pattern with epsilon wave; described as a terminal notch in the QRS complex, due to slowed intraventricular conduction. Ventricular

ectopics in ARVD have LBBB morphology, with a QRS axis of -90 to +110 degrees. Most commonly the ectopics arise from the right ventricular outflow.

On the other hand UHL's anomaly or parchment heart syndrome is said to be almost complete absence of right ventricular myocardium resulting in extremely thin ventricular wall. Some people do not differentiate the two disorders. However several studies have studied the differential features of the two disorders. UHL's anomaly being distinguished from ARVD by more extensive loss of myocardium, early age of presentation (neonatal); As opposed to ARVD which presents after 20 years and patchy involvement of myocardium [2].

Our case presented late but had almost complete absence of the myocardium. Also ECG showed ectopics with LBBB morphology; although it was debated that arrhythmias may have been due to digitalis toxicity as patient's arrhythmias and clinical status improved when digitalis was stopped due to bradycardia; as patient's heart rate was 40/min on admission.



However there have been a few reports of non invasive recognition of this UHL's anomaly even to the age of 50 years [3].

Management of both these disorders is mainly preventive by anticoagulation and anti-arrhthymic therapy including intra cardiac defibrillators.

Our case was also important as it also gives a learning message to students not to try a blind pleural tap whenever feasible, had someone tried it in our patient it would have been disastrous. Also our case had chylous ascitis which occurs in only about 0.5 % cases of cirrhosis [4] and in cases of cardiac disorder there are only few reports of chylous ascitis with chronic constrictive pericarditis[5]; but we could not lay hands on any other report on this phenomenon in any other type of right heart failure. Chylous ascitis commonly occurs following trauma, surgery, malignancy or tuberculosis.

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