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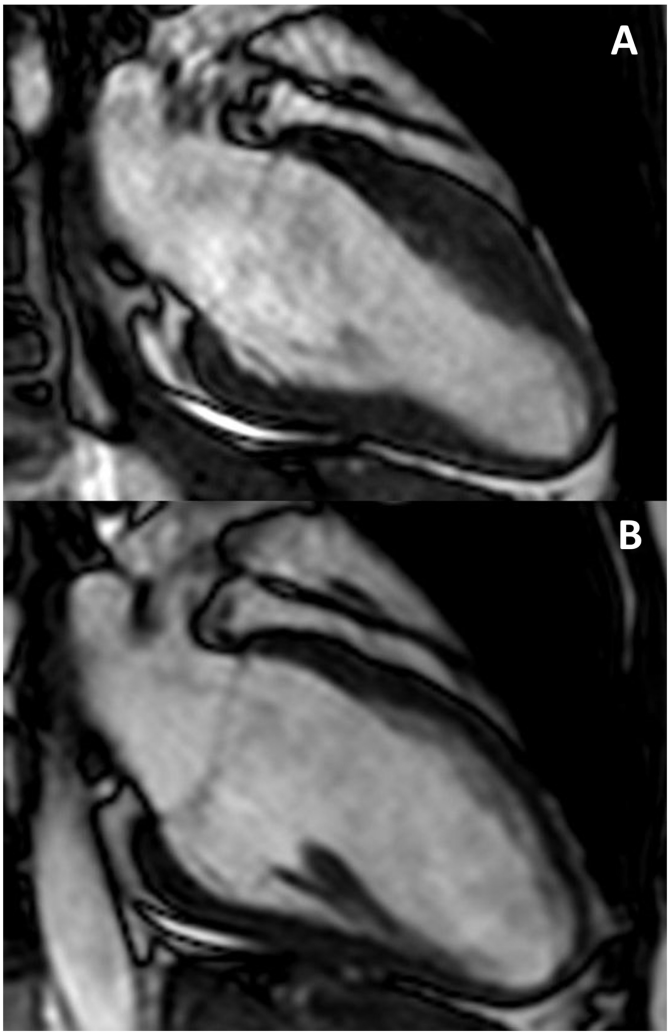

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Kiosk 9Q-TA-03
A Rare Genetic Cardiomyopathy Mimicking Cardiac Amyloidosis

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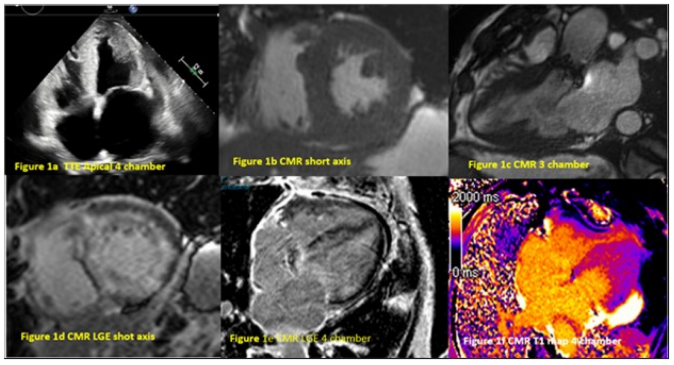
Description of Clinical Presentation: • A 78 –year-old male patient with history of chronic lymphocytic leukemia in remission, atrial fibrillation, bio-prosthetic aortic valve replacement for severe aortic insufficiency, hypertension, right hand carpal tunnel syndrome, peripheral neuropathy, pericardial effusion, presented with 2 months of worsening dyspnea on exertion, fatigue and lower extremity swelling. Clinical examination showed elevated jugular venous distension, irregularly irregular cardiac rhythm and lower extremity edema. Labs were remarkable for elevated Pro-BNP, elevated Hs-Troponin and abnormal immunoglobulins with elevated IGG level. Echocardiogram (Figure 1a) revealed moderate left ventricular (LV) hypertrophy with moderately reduced systolic function, bi-atrial enlargement and myocardial speckling pattern concerning for cardiac amyloidosis.

Diagnostic Techniques and Their Most Important Findings: Cardiac MRI Figure (1b-f) showed moderate global left ventricular hypokinesis and severe concentric LV hypertrophy (septal thickness of 1.6 cm). T1 myocardial relaxation time (Figure 1 f) was elevated at 1180 msec with markedly elevated extracellular volume fraction (50%). Delayed enhancement images (Figure 1d, 1e) showed diffuse near transmural biventricular, bi-atrial, interatrial septal and pericardial late gadolinium enhancement with reversed myocardial nulling pattern on T1 scout sequence images. All these findings including the clinical history was strongly suggestive of cardiac AL amyloidosis. Both endomyocardial biopsy and aortic valve tissue pathology were both negative for cardiac amyloidosis. Genetic testing yielded a pathogenic variant of TTN gene and patient was diagnosed with Titin restrictive cardiomyopathy.

Learning Points from this Case: We present an unusual presentation of Titin (TTNtv) cardiomyopathy which mimicked cardiac amyloidosis. It is a rare form of genetic cardiomyopathy due to a truncating variant in the titin gene (TTNtv) and is associated with increased risk of atrial and ventricular arrhythmias. Multimodality cardiac imaging and cardiomyopathy genetic testing plays a vital role in early diagnosis and treatment of this rare condition.



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Kiosk 9Q-TA-04
Acute Myocardial Calcification Demonstrated on Multimodal Imaging

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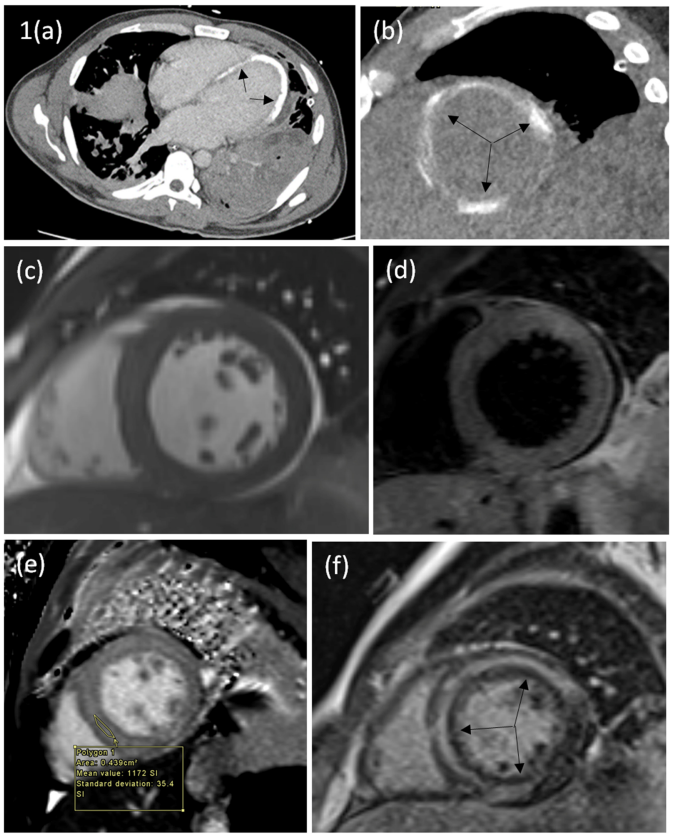
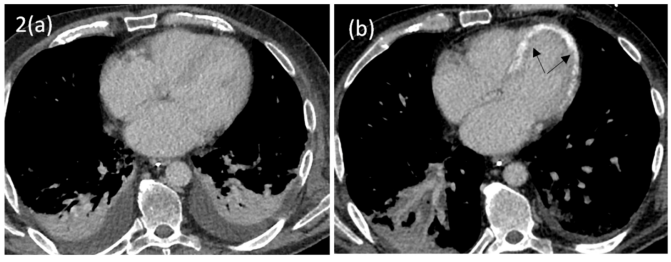
Description of Clinical Presentation: Three male patients with ages ranging between 30-50 and no previous cardiac history were

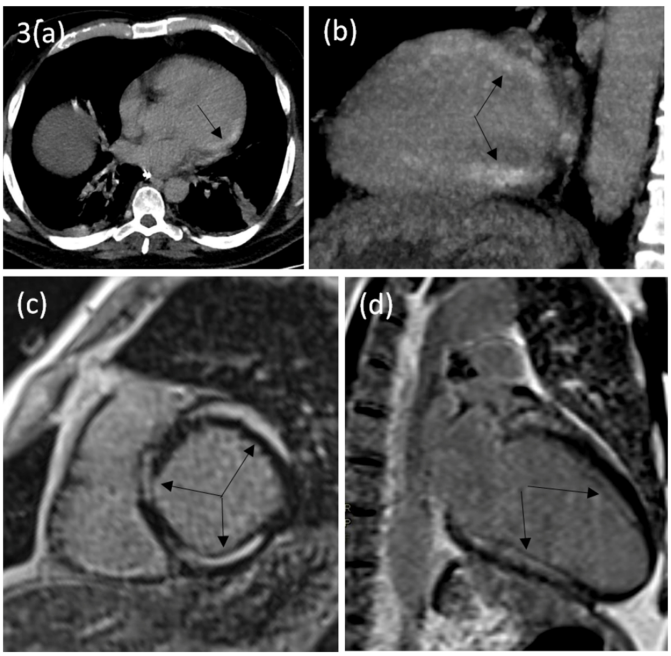
admitted to hospital, requiring prolonged intensive care admission for multiorgan failure secondary to septic shock. All patients had clear infective causes identified at presentation (peritonitis secondary to anastomotic leak post-elective small bowel resection, infective colitis and empyema), were intubated and ventilated and required emergency haemofiltration. Inpatient thoracic and abdominal computed tomography (CT) scans demonstrated new left ventricular (LV) myocardial calcification during the course of their illness. In two cases, these were diffuse and picked up by the general CT reporter. The other was more subtle and picked up during multidisciplinary team (MDT) review.

Diagnostic Techniques and Their Most Important Findings: All patients had CT Abdomen and pelvis scans (contrast and unenhanced) during the workup of their presenting conditions, which incidentally found new LV myocardial calcification. Subsequent bedside transthoracic echocardiography demonstrated mild-to-moderate LV impairment (Ejection fraction of between 40 – 45%) with regional wall motion abnormalities, and all had elevated blood troponin T levels. One patient's cardiac magnetic resonance (CMR) imaging demonstrated regional oedema on short tau inversion recovery (STIR) sequence but diffusely raised native T1 values with widespread myocardial fibrosis on late gadolinium enhancement (LGE), corresponding to CT changes. The second patient's CMR revealed mid wall enhancement of the basal to mid anterior, septal and inferior walls on LGE corresponding with regional calcification seen on CT scans. There was extensive, diffuse epicardial-mid wall LGE also seen in the third patient, similar to patterns seen in myocarditis and other non-ischaemic cardiomyopthies[1,2].

Learning Points from this Case: These cases all demonstrate presentations of severe sepsis, with LV impairment and renal failure in which the described pattern of myocardial calcification was incidentally identified over a relatively short period of time within our hospital Trust. The available literature indicates this is a rare entity with unknown long-term impact[3]. This is a condition that non-cardiac reporters may be unfamiliar with and may overlook. None of these cases were under specialist cardiology care highlighting the need for awareness from radiologists and other clinicians to ensure appropriate management and follow up for these patients is pursued. Myocardial calcification is not well seen on echocardiography or CMR, and therefore may be missed[4]. Further data needs to be gathered for significance on acute management as well as long term outcome for these patients[5]. A national database may also aid the development of knowledge in this matter.







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Kiosk 9Q-TA-05

Central Conducting Lymphatic Anomaly: A Rare Cause of Chylopericardium in a Child Diagnosed on Magnetic Resonance Lymphangiography

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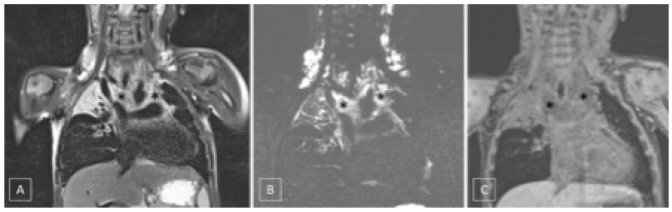
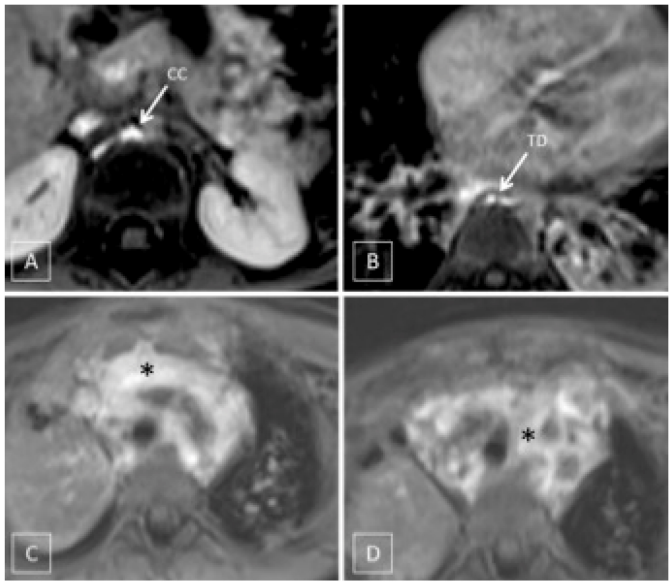
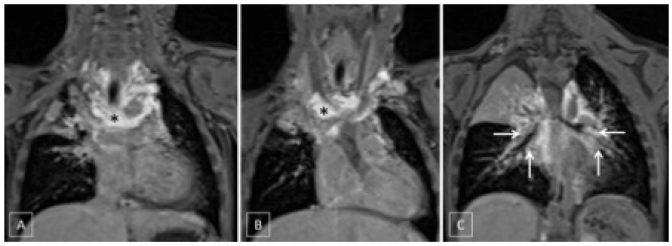
Description of Clinical Presentation: A two-year-old boy presented with fever and respiratory distress. Transthoracic echocardiography revealed presence of massive pericardial effusion. The effusion was drained; the drained fluid was milky white in appearance with biochemical analysis suggestive of chylopericardium. Tubercular work up was negative.

Diagnostic Techniques and Their Most Important Findings: MR lymphangiography was performed to further evaluate the cause of chylopericardium. An ill-defined soft tissue was seen extending from lower neck till subcarinal region draping along the aortic arch, arch vessels, left brachiocephalic vein, trachea and abutting the pericardial reflections. This soft tissue was also seen extending along bilateral peri-bronchial interstitial tissue. **(Figure 1)** It appeared slightly hyperintense on T2-weighted images and hypo-to-isointense on T1-weighted images. Under USG guidance, a inguinal lymph node on either side was cannulated using a 25-gauge needle. Dynamic contrast-enhanced (DCE) MR lymphangiography was performed. The thoracic duct was visualized in left paramedian location only till the subcarinal region with the superior part of thoracic duct and its venous confluence not seen. There was presence of reflux into the lung parenchyma along peri-bronchial interstitial tissue suggestive of pulmonary lymphatic perfusion syndrome. The reflux was also seen in mediastinal soft tissue described previously which showed homogenous enhancement. **(Figure 2,3)** The was no evidence of bony, splenic or hepatic involvement. In view of presence of ill-defined mediastinal soft tissue, non-visualization of superior thoracic duct and presence of lymphatic reflux, a likely diagnosis of central conducting lymphatic anomaly was made. The child was started on Sirolimus and there was resolution of pericardial effusion and lymphatic mass at 6 months follow up.

Learning Points from this Case: MR lymphangiography is an encouraging, minimally invasive imaging method for the diagnosis of lymphatic abnormalities. Central conducting lymphatic anomaly are rare group of disorders with overlapping imaging features and its imperative to be familiar with the imaging characteristics so that timely diagnosis can be made and appropriate management instituted. Sirolimus is an emerging effective treatment option in rare complex lymphatic anomalies.







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