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Sarcoidosis: Hiding in Plain Sight

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Association of ANK2 Mutation and Massive Myocardial Calcification: A Case Report

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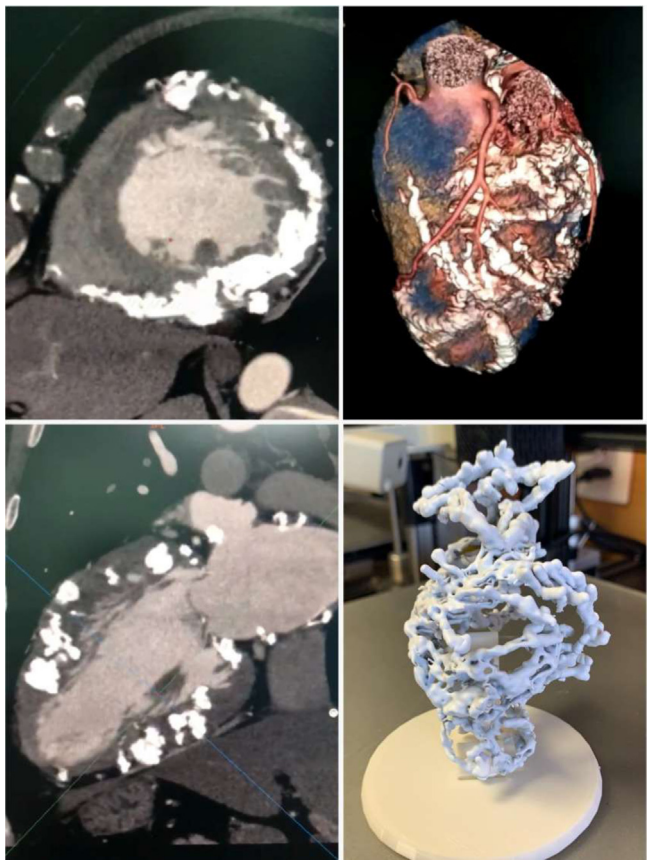
Introduction: A 35-year-old man with no prior history of cardiovascular diseases was admitted with progressive exertional chest discomfort. Physical examination revealed irregular heart rhythm and an ejection murmur at the aortic area. EKG showed atrial fibrillation, and chest X-ray, calcification within the cardiac silhouette. Laboratory tests were all within normal ranges, including serological and inflammatory tests.
Case Report: An echocardiogram demonstrated diffuse myocardium calcification and intense mitral- aortic fibrocalcification, causing severe stenosis and moderate regurgitation, with a dilated left ventricle (LV) and moderate systolic dysfunction, akinesia of the inferolateral walls and hypokinesia of the anterior walls. A 128-row multidetector CT showed massive calcification affecting the aortic and mitral valves, the right ventricle, the left atrium wall and all the segments of the LV, preserving only the basal septum. Cardiovascular magnetic resonance imaging 1.5T revealed diffuse myocardial thickening, matching the areas seen on CT (figure). These areas were hypointense on the SSFP cine images and on T1- and T2-weighted images and had no late gadolinium enhancement (LGE). However, there was LGE in the underlying myocardium, in a non-ischemic distribution. Pericardium and coronary arteries were preserved. Genetic analysis identified a rare heterozygous variant of uncertain significance, c.10702C>T(p.Arg3568Trp), in ANK2 gene.
Summary: Ankyrin-B, also known as Ankyrin-2, is encoded by the ANK2 gene, which plays critical roles in the localization and stabilization of ion transporters and ion channels in cardiomyocytes. The Ankyrin-B syndrome has been associated to long QT syndrome and torsades depointes, but there are no reports of myocardial calcification. The patient was found to be a candidate for cardiac transplantation and is currently on the waiting list.



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Sarcoidosis: Hiding in Plain Sight

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Introduction: Sarcoidosis is a systemic disease that can masquerade as many conditions. Due to its often patchy distribution, myocardial biopsy is only ~30% sensitive. Herein we present a case of sarcoidosis diagnosed from an extra-cardiac biopsy years after the onset of cardiomyopathy.
Case Report: A 54 year old man initially presented with dyspnea and fatigue due to new onset HFrEF of 20%. He had a significant family history of SCD. His workup revealed non-obstructive CAD, a myxomatous mitral valve with bileaflet prolapse and moderate regurgitation, and a cystic structure attached to the tricuspid valve. Given his frequent ventricular ectopy, he was discharged on guideline therapy for HFrEF and a wearable cardioverter defibrillator. Genetic testing for dilated cardiomyopathies revealed EYA4 and TTN variants of unknown significance. A cardiac MRI a few months later demonstrated no abnormal late gadolinium enhancement and persistent systolic dysfunction, so he underwent AICD implant. His course was further complicated by recurrent admissions for supraventricular and ventricular tachyarrhythmias. A cardiopulmonary exercise stress test demonstrated low-risk results. Further evaluation noted prolonged AV conduction. A RHC revealed mild post-capillary pulmonary hypertension and a mildly decreased cardiac index at 2.01 L/min/m^2. Months later, he underwent CRT-D upgrade. He started improving functionally until he developed left eyelid swelling, initially thought to be lymphoma. An orbital biopsy revealed non-caseating granulomas. A PET/CT demonstrated FDG uptake within subcarinal lymph nodes but no cardiac uptake. Initially he declined immunomodulatory therapy, but as his symptoms worsened, he started corticosteroids and transitioned to mycophenolate mofetil due to intolerance of methotrexate. Due to disease progression on repeat FDG-PET, he was switched to infliximab.
Summary: The diagnosis of sarcoidosis can be difficult due to its variable presentation, including heart block, heart failure, ventricular arrhythmias and sudden cardiac death. Cardiac involvement occurs in 25% of cases. Our case was complicated by a cMRI that was negative for cardiac involvement, which emphasizes the importance of complimentary inflammatory imaging with FDG-PET. A multidisciplinary approach is needed that engages sarcoid specialists for earlier diagnosis to ensure rapid initiation of therapy to reduce end-organ dysfunction.

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WITHDRAWN

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Successful Pregnancy with HeartMate 3 (Abbott) Left Ventricular Assist Device

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Introduction: Left ventricular assist devices (LVADs) are an evolving therapy for patients with end-stage heart failure (HF). Due to high maternal and fetal risk from hemodynamic variation and need for anticoagulation, LVADs are a relative contraindication to pregnancy. Successful pregnancies have been observed with axial-flow LVADs as well as the continuous-flow HeartWareTM (Medtronic) LVAD, but the safety of the continuous-flow HeartMate 3TM (HM3) LVAD (Abbott) with pregnancy is unknown. Herein we present a case of a successful delivery in a patient with a HM3 LVAD.
Case Report: A 39-year-old female with peripartum cardiomyopathy underwent HM3 implantation as destination therapy due to severely elevated panel-reactive antibodies. She followed routinely with HF clinic. Two years after implantation, she presented to HF clinic at 15 weeks gestation despite oral birth control. She received counseling on the risk of pregnancy