1 IMAGING CARDIOVASCULAR FEATURES OF A FAMILY WITH TYPE 4 LOEYS-DIETZ SYNDROME

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Introduction Loeys-Dietz syndrome (LDS) is an inherited autosomal dominant disorder caused by pathogenic variants in the genes involved in TGF β signaling. There is a wide phenotypic spectrum of cardiovascular, skeletal, craniofacial and cutaneous manifestations. The hallmark of this rare connective tissue disorder is progressive aortic and peripheral arterial aneurysmal disease leading to acute events. We evaluated the range of cardiovascular abnormalities in a family with type 4 LDS. The background literature data and screening strategy is discussed.

Methods We retrospectively studied the radiological cardiovascular findings in 23 members of a family with features of LDS who were monitored with echocardiogram. 17 were investigated with MRI and 9 with CT. The head and neck arteries, the aorta and its visceral branches, iliac and femoral arteries were assessed. Size of the vessels, tortuosity, length and presence and the onset of arterial dissection were investigated.

Results 11/23 patients revealed arterial abnormalities, all with TGF β 2 variant. Two patients had type-A dissection at the age of 48 and 49, one treated surgically and the other medically. Three patients with aortic root dilatation underwent preventive personalized external aortic root support procedure (PEARS). The other abnormalities included: mild aortic root dilatation (3), infrarenal aortic aneurysm (1), tortuosity of carotid (2) and vertebral arteries (1), aneurysmal iliac arteries (1), dilated coeliac trunk and superior mesenteric arteries (1).

Conclusion Vascular abnormalities are common in patients with familial history of LDS and can predispose to potentially fatal events in later life. Assessing for cardiovascular risk in LDS is crucial.

2 RELATIVE VALUE OF CARDIAC MRI AND FDG-PET IN TREATMENT FOLLOW-UP FOR CARDIAC SARCOIDOSIS

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Introduction 18 F-FDG PET-CT and cardiac MR (CMR) are described in the diagnosis of cardiac sarcoidosis (CS), each identifying different aspects of the disease. We compared PET-CT and CMR in the follow-up of patients with suspected CS.

Methods 31 patients with proven extra-cardiac sarcoidosis and possible CS were identified from the local sarcoidosis registry. All had combined PET-CT and CMR on two occasions. Selected patients had not received immunosuppressants for at least 6 months before the initial combined study. 22 patients were treated with immunosuppressants for 3 months or longer. In 9, the clinician chose not to treat or the patient refused treatment. Follow-up PET-CT and CMR were done 102-770 days later (median 228).

Results Significant myocardial FDG uptake was shown on visit 1 in 17 treated patients (myocardial SUV $_{max}$ >3.6). Myocardial SUV $_{max}$ fell significantly on follow-up ($p < 0.01$) and was matched falls in FDG avid lung and node disease. Treated patients also showed improvement in left ventricular ejection

fraction (LVEF) ($p = 0.03$). 14 treated patients had late gadolinium enhancement (LGE) which was not necessarily matched by FDG. This was unchanged by visual assessment on follow-up. In 9 untreated patients (4 with FDG avid myocardium and 5 with LGE), there was no change in myocardial FDG, LGE or LVEF.

Conclusion Myocardial FDG uptake in CS represents active inflammation. When treated, this resolved or regressed on follow up with an improvement in LVEF and FDG avid thoracic disease. There was no change in extent of LGE. Untreated patients showed no change.

3 USPIO-ENHANCED MAGNETIC RESONANCE CORONARY ANGIOGRAPHY COMPARED TO COMPUTED TOMOGRAPHY CORONARY ANGIOGRAPHY

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Background Recent concerns regarding gadolinium has led to the investigation of alternatives for magnetic resonance coronary angiography (MRCA) including non-contrast imaging and contrast imaging with ultrasmall superparamagnetic particles of iron oxide (USPIO).

Methods Seven patients underwent USPIO-enhanced MRCA using a 3 Tesla PET/MRI scanner and computed tomography (CT) coronary angiography (CTCA) using a 128-multidetector computed tomography (CT) scanner. MRCA was performed using FLASH magnetic resonance angiography sequences with 3 to 4 mg/kg of intravenous ferumoxytol. Image analysis was performed by two observers blinded to other imaging. Per segment image quality was rated on a 4-point scale. Per vessel coronary assessment was performed, with segments classified as less than or greater than 50% luminal diameter.

Results 99 segments in 7 patients were assessed. Diagnostic image quality was observed in 80% (79/99) of segments with USPIO-enhanced MRCA compared to 99% (98/99) with CCTA. 85 segments in 21 vessels were available for the assessment of stenoses. On CCTA, 12 segments in 8 vessels had one or more >50% stenosis. Per vessel diagnostic accuracy of USPIO-enhanced MRCA was good with sensitivity 63%, specificity 77%, positive predictive value (PPV) 63%, negative predictive value (NPV) 77%, and accuracy 71%, and was better for proximal compared to distal segments.

Conclusion USPIO-enhanced MRCA has a good image quality for the identification of vessel segments and stenoses, particularly in proximal segments.

4 STUDYING CONTRAST ADMINISTRATION IN THE FONTAN CIRCULATION USING MR TIME RESOLVED ANGIOGRAPHY

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Introduction When patients with Fontan circulation require CT imaging, there are significant challenges in achieving adequate contrast opacification due to altered anatomical