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IMAGES IN CARDIOLOGY

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Echocardiographic clues to diagnosis of dystrophin related dilated cardiomyopathy

A 32-year-old man with a four-year history of congestive heart failure was referred to our institution. Two years previously, the patient had received a diagnosis of idiopathic dilated cardiomyopathy with severe secondary mitral insufficiency. Clinical reappraisal focused attention on the following features: (1) normal coronary arteries at angiography; (2) constantly elevated creatine phosphokinase (CPK) plasma values (300–450 mU/ml); (3) localised inferobasal left ventricular akinesia and thinning (arrow in panel A); (4) spongy appearance of the apicolateral left ventricular wall (arrow in panel B), suggesting ventricular non-compaction. It was also noted that the patient had been complaining of muscular asthenia (hitherto ascribed to heart failure), especially of the lower limbs. Suspected dystrophinopathy was confirmed after myocardial and peripheral muscle biopsies; immunostaining of myocardial tissue (panel C) for detection of N-terminal domain of dystrophin (monoclonal antibody NCL-DYSB, clone 34C5, Novocastra) revealed discontinuous and partially disrupted positive myocytes (of varying intensity) interspersed with negative myocytes. The final diagnosis was dystrophin related, severe dilated cardiomyopathy in a patient with neurologically mild Becker disease. Surgical mitral valve annuloplasty was attempted, but heart failure continued to worsen during the following two years. Heart transplantation was successfully performed. Macroscopic examination of the explanted organ confirmed the localised inferobasal left ventricular thinning (oval in panel D) and the presence of left ventricular non-compaction (rectangle in panel D).

In many cases dilated cardiomyopathy should nowadays no longer be considered a final diagnosis, but rather a starting point for further assessment. Molecular biology techniques increasingly allow identification of distinct aetiologies that share the dilated

cardiomyopathy phenotype. At each diagnostic step, the clinician needs to recognise highly specific (though not sensitive) signs that can be indicative of the true final diagnosis. In the present case, the echocardiogram contains two such “red flags”—namely, ventricular non-compaction and localised inferobasal left ventricular akinesia. Taken separately, either of these signs would raise a strong suspicion of dystrophinopathy. When combined, they are almost pathognomonic.

C Rapezzi
O Leone
E Biagini
F Coccolo
 crapezzi@aosp.bo.it