THE ANATOLIAN JOURNAL OF CARDIOLOGY

Official journal of the TURKISH SOCIETY OF CARDIOLOGY

Spontaneous Coronary Artery Dissection in the Setting of Duchenne Muscular Dystrophy: More Questions Than Answers

LETTER TO THE EDITOR

To the Editor,

Duchenne muscular dystrophy (DMD) has been widely recognized as an X-linked dystrophin-associated myopathy generally presenting with an unfavorable outcome.[1-3] The recent article by Öncül et al[1] has reported an interesting case of acute myocardial infarction (AMI) associated with spontaneous coronary artery dissection (SCAD) that was successfully managed with percutaneous coronary intervention in a boy with DMD. In this regard, we would like to further comment on the implications of SCAD in the setting of DMD.

In the clinical setting, DMD elicits a specific form of progressive skeletal muscle dysfunction along with cardiomyopathy (attributable to dystrophin deficiency) and generally ends up with mortality in early adulthood (largely due to respiratory complications, arrhythmias, etc.).[1,2] At the cellular level, dystrophin serves as an important constituent of dystrophin-associated protein complex that connects actin filaments to sarcolemma and extracellular matrix.[2,3] Mechanistically, dystrophin provides a strong sarcolemmal stability and integrity, and hence, myocyte protection against the repetitive strain generated by cyclic myocyte contraction might lead to eventual myocyte degeneration in the skeletal system and heart.[2,3] Muscular degeneration along with elastin fragmentation involving the vascular media layer (namely cystic medial necrosis) might serve as the primary trigger of SCAD evolution in a portion of cases.[4] However, it seems quite unusual to encounter degeneration due to dystrophin deficiency in myocellular components of the vascular structures including coronary arterial tree that is well known to demonstrate characteristic vasomotion pattern (and not a cyclic myocontractile pattern). In other terms, molecular aspects of muscular dystrophy fail to explain the potential vascular degeneration and associated complications (including SCAD) in patients with DMD.

Based on the aforementioned notions, SCAD evolution in the setting of DMD might be primarily attributable to the accompanying conditions (including enhanced endogenous stress, side effects of the drugs used, etc.) rather than the progressive coronary vasculopathy associated with dystrophin deficiency. It may be suggested that patients with DMD might potentially face an enormous level of emotional and physical stress due to the progressive and debilitating nature of the disease course, opportunistic infections, as well as extreme social isolation, etc. Accordingly, high levels of adrenergic discharge associated with endogenous stress were previously suggested as a potential trigger of SCAD.[4,5] Accordingly, we wonder about the presence and magnitude of potential stressors in the patient.[1]

Notably, potential side effects of the drugs used in the patient (including prednisolone and ataluren) might have also contributed to the evolution of SCAD in the patient. It is well known that steroids might potentially exert a detrimental impact on vascular structures leading to vascular frailty that might result in potential complications including coronary aneurysm formation in susceptible subjects.[6] Importantly, ataluren, the disease-modifying agent in the setting of DMD, might

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Cite this article as: Yalta K, Öztürk C, Yalta T. Spontaneous coronary artery dissection in the setting of duchenne muscular dystrophy: More questions than answers. *Anatol J Cardiol.* 2023;27(2):117-118.

DOI:10.14744/AnatolJCardiol.2022.2488

potentially enhance muscle dystrophin expression, yet might have potential side effects including vomiting, hypertension, etc.,[7] that all might have facilitated the evolution of SCAD (through mechanisms including Valsalva strain, direct vascular injury, etc.)[4] in the patient. Accordingly, did the patient[1] report any adverse effects associated with ataluren?

Finally, electrocardiogram (ECG) demonstrates typical signs of DMD-related cardiac involvement (augmented R/S ratio in V1-V3 and deep Q waves in V5-V6)[2] in the patient despite the reported normal ECG findings.[1] Similar to ECG changes, specific cardiac magnetic resonance imaging (MRI) findings (including subepicardial late gadolinium enhancement particularly involving the inferolateral walls) might emerge long before the evolution of overt wall motion abnormalities on ECG.[2] Therefore, we wonder about cardiac MRI findings in the patient. In this context, anterior AMI due to SCAD, on top of DMD-related myocardial involvement, might potentially accelerate adverse myocardial remodeling (even though the patient reportedly receives losartan) and hence transition to overt cardiomyopathy. This potentially warrants more frequent examination of cardiac functions in the patient for the timely initiation of guideline-directed heart failure therapy. Therefore, we wonder about the strategy of long-term surveillance (frequency of cardiac imaging and other tests including Holter monitoring, etc.) in the patient.

In summary, SCAD evolution in the setting of DMD might be regarded as an ill-defined and potentially multi-factorial phenomenon possibly attributable to the adverse impact of accompanying conditions (extreme levels of endogenous stress, drugs, etc.). Importantly, coronary complications (including SCAD) might significantly worsen the long-term cardiovascular prognosis and might necessitate close supervision of cardiac functions in patients with DMD.

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