Co-existing cardiomyopathy in the setting of congenital coronary artery anomalies: Further insights into pathogenetic and clinical aspects

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In clinical practice, coronary artery anomalies (CAAs) have been rarely encountered, and usually, they encompass a variety of congenital abnormalities in the origin, course, and termination of major coronary arteries [1–3]. Fortunately, most of these anomalies are clinically benign [2, 3]. However, certain CAAs (such as anomalous left coronary artery arising from the pulmonary artery [ALCAPA] or anomalous coronary arteries with an interarterial or intramural course) might be particularly associated with unfavorable outcomes including sudden cardiac death (SCD) [3]. In particular, emerging myocardial dysfunction significantly contributes to the adverse prognosis in the setting of CAAs [3]. The recent report by Silva et al. [1] describes a middle-aged male with an anomalous left coronary artery arising from the contralateral aortic sinus with diverse courses of its major branches [1]. Notably, the patient also had idiopathic left ventricular systolic dysfunction [1]. Accordingly, we would like to make further comments on potential implications of various cardiomyopathy patterns in the setting of CAAs.

First, certain CAAs (such as ALCAPA or myocardial bridge) might directly account for significant myocardial injury through induction of acute or chronic coronary syndromes (due to severe coronary hypoxemia, enhanced atherogenesis, vasospasm, etc.) [2, 3]. In the long term, a substantial amount of ischemic myocardial injury followed by a compensatory remodeling process might lead to ischemic cardiomyopathy in this context [3]. Even though, the anomalous coronary artery seems patent in such patients [1], it might potentially have subtle abnormalities such as an existing short intramural segment with an intermittent narrowing (that might have gone undetected) or episodes of coronary vasospasm (that might only be detected with a coronary vasoreactivity test). Given the absence of late gadolinium enhancement on cardiac MRI [1], extensive myocardial hibernation or stunning due to episodic ischemia might also serve as a potential trigger of cardiomyopathy in such patients (and, hence, indicate further tests for myocardial viability). We also wonder about other electrocardiographic (ECG) findings (Q waves, etc.), if they are available [1].

Second, myocardial dysfunction (manifesting as non-ischemic cardiomyopathy) and CAAs might independently arise as the major components of certain congenital cardiac anomalies such as tetralogy of Fallot (TOF) [2, 3]. Moreover, various combinations of these abnormalities (congenital cardiac defects, CAAs, and cardiomyopathy) might emerge as part of a systemic syndrome. In this regard, the patient might potentially harbor such a systemic syndrome due to his suspicious findings including mental, auditory, and visual deficits along with an existing horse-shoe kidney [1]. Accordingly, did the authors plan genetic counseling for further phenotypical analysis of the patient? Importantly, isolated non-ischemic cardiomyopathy (due to various triggers) might also arise coincidentally in those with CAAs.

Finally, and more subtly, certain forms of isolated familial cardiomyopathies and major CAAs might also co-exist in certain settings

[4]. Notably, there has been a particular co-existence of hypertrophic cardiomyopathy and anomalous origin of coronary arteries [4]. More specifically, a mitochondrial gene mutation (*MT-TK* gene encoding transfer RNA) was previously identified in a young patient with an anomalous left coronary artery arising from the right aortic sinus together with apical hypertrophic cardiomyopathy [4]. Notably, deafness and cardiomyopathy (hypertrophic or dilated) similar to the features of the patient described in [1] were previously interpreted as the manifestations of this gene mutation [4]. Therefore, the patient [1] might be in the late (burned-out) phase of hypertrophic cardiomyopathy primarily characterized by relative wall thinning and systolic dysfunction. Alternatively, he might have familial dilated cardiomyopathy primarily presenting with systolic dysfunction [1]. Importantly, certain forms of familial cardiomyopathy might have significantly worse outcomes including SCD [4], which potentially indicates the need for implementing preventive strategies including implantable cardioverter-defibrillator (ICD) therapy in the relatively early stages of the disease course. Taken together, genetic analysis (for gene mutations implicated in familial cardiomyopathies) together with subsequent family screening (with imaging modalities) might enable further risk-stratification and management of this patient along with an early diagnosis of CAAs and/or familial cardiomyopathy, if any, in his family members.

In conclusion, co-existing myocardial dysfunction in the setting of major CAAs seems to be a multifaceted phenomenon with important implications [1–4]. Importantly, differentiation between various cardiomyopathy patterns in patients with CAAs (largely through advanced imaging modalities, genetic analysis, etc.) might allow proper risk stratification and management of these patients and their family members.

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