A rare cause for exertional shortness of breath and acute coronary syndrome in a 21 year old postpartum mother

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Abstract

21 year old postpartum mother presented with worsening exertional dyspnoea 6 weeks after uncomplicated vaginal delivery of her 1st child. She admitted with ischemic type chest pain associate with autonomic symptoms for 2 days duration. ECG on admission showed ST segment depressions in lead I, AVL, V3, V4, V5, V6. Her troponin I titer was initially 0.73 and rose to 1.32 ng/mL(reference range below 0.12 ng/mL). Her full blood count, inflammatory markers, liver and renal function tests were normal.

Her 2D echocardiogram revealed dilated left and right coronary arteries and peculiar feature of multiple colour Doppler flows inside left ventricular muscle. She had preserved Left ventricular ejection fraction with no regional wall motion abnormalities. She underwent coronary angiography which revealed abnormal left main coronary artery originating from main pulmonary artery where the left system fills retrogradely via collaterals from right coronary artery. It was confirmed by CT coronary angiogram. There was no angiographic evidence of obstructive coronary lesions.

The patient was referred to cardiothoracic surgeon for corrective surgery and advised to avoid pregnancy until corrective surgery is performed. This case illustrates the importance of vigilant workup of patients presenting with acute coronary syndrome at young age as congenital anomalies of coronary arteries may present for the first time in adult life.

Individuals with childhood-onset coronary artery anomalies are at increased risk of lifelong complications. Although pregnancy is thought to confer additional risk, a few data are available regarding outcomes in this group of women. We sought to define outcomes of pregnancy in this unique population.

We performed a retrospective survey of women with paediatric-onset coronary anomalies and pregnancy in our institution, combined with a systematic review of published cases. We defined paediatric-onset coronary artery anomalies as congenital coronary anomalies and inflammatory arteriopathies of childhood that cause coronary aneurysms. Major cardiovascular events were defined as pulmonary oedema, sustained arrhythmia requiring treatment, stroke, myocardial infarction, cardiac arrest, or death.

A total of 25 surveys were mailed, and 20 were returned (80% response rate). We included 46 articles from the literature, which described cardiovascular outcomes in 82 women (138 pregnancies). These data were amalgamated for a total of 102 women and 194 pregnancies; 59% of women were known to have paediatric-onset coronary artery anomalies before pregnancy.

In 23%, the anomaly was unmasked during or shortly after pregnancy. The remainder, 18%, was diagnosed later in life. Major cardiovascular events occurred in 14 women (14%) and included heart failure (n=5, 5%), myocardial infarction (n=7, 7%), maternal death (n=2, 2%), cardiac arrest secondary to ventricular fibrillation (n=1, 1%), and stroke (n=1, 1%). The majority of maternal events (13/14, 93%) occurred in women with no previous diagnosis of coronary disease.

The anomalous origin of the left coronary artery from the pulmonary artery - known as Bland-White-Garland syndrome - is a rare congenital malformation that affects 1 in 300,000 live births. Most patients die in infancy without any surgical treatment. Some patients who survive past childhood often have varying symptoms such as myocardial ischemia, impaired left ventricular function, mitral regurgitation, and progressive heart failure, depending on the development collateral circulation. In the present report, we describe a procedure wherein the left coronary artery ostium was translocated through the transverse sinus of the pericardium in a 43-year-old mother with Bland-White-Garland syndrome and concomitant mitral regurgitation and report on the associated midterm results.

Women with paediatric-onset coronary artery anomalies have a 14% risk of adverse cardiovascular events in pregnancy, indicating the need for careful assessment and close follow-up. Prospective, multicentre studies are required to better define risk and predictors of complications during pregnancy.

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