CLINICAL VIGNETTE

Coronary Artery Aneurysms in a Young Woman due to Kawasaki Disease

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Introduction

Coronary Artery Aneurysms (CAA) are an abnormal dilatation of the coronary arteries of unknown etiology. CAA is defined as a localized dilation of more than 1.5 times the diameter of an adjacent artery. Most of the CAAs's are incidentally discovered on angiography. Symptomatic patients can present with unstable angina, myocardial infarction, arrhythmias or sudden cardiac death.[1]

Case Description

A 29-year-old female with no significant past medical history, presented to the emergency room with chest pain. The chest pain was acute, radiating to bilateral arms. She had associated heart burn, nausea and worsening shortness of breath. She had vomited on her way to the ER. The EKG on admission showed ST depression in the inferior leads, and ST elevation in the anterior leads. Troponins were greater than 6,000. Acute coronary angiogram revealed severe coronary arteries aneurysms with the left anterior descending coronary artery completely occluded with collaterals, and right coronary artery with 90% stenosis. She underwent urgent two vessel Coronary Artery Bypass Grafting did well post operatively. After discharge she presented to rheumatology for further management of presumed Kawasaki Disease.

Discussion

CAA's can be classified into three major groups based on the pathology findings, atherosclerotic, inflammatory and non-inflammatory. The most common cause of CAA's in adults is atherosclerosis, being responsible for almost half the cases.[2] Kawasaki disease (KD) is the major cause of CAA's in children and is the second most common cause of inflammatory CAAs in adults.[3] Table 1 presents a more detailed list of etiologies.

Causes of CAA's

Etiology	Examples
Congenital	Bicuspid Aortic Valve, Fibromuscular Dysplasia
Atherosclerotic	Atherosclerotic CAA's
Inflammatory	Kawasaki Disease, Takayasu's Arteritis, SLE, Bechet's Disease, Polyarteritis Nodosa, Rheumatoid Arthritis, IG-G4 Related disease
Infectious	Bacterial, mycotic aneurysm, syphilis, HIV, septic emboli, COVID
Drug Related	Cocaine, Protease Inhibitors, Amphetamines
Iatrogenic	Percutaneous Transluminal Coronary Angioplasty
Traumatic	Penetrating cardiac injury

Kawasaki Disease (KD)

KD is one of the most common vasculitides of childhood that rarely occurs in adults.[4] It is typically a self-limited condition, with fever and features of acute inflammation lasting for an average of 2 weeks without therapy.[5] However, the complication of CAA can lead to significant morbidity and mortality.

The diagnosis of KD requires the presence of fever lasting at least 5 days without any other explanation combined with at least 4 of the 5 following criteria: bilateral bulbar conjunctival injection, oral mucus membrane changes, peripheral extremity changes, polymorphous rash, and cervical lymphadenopathy (at least 1 lymph node >1.5 cm in diameter). Infection must be ruled out.[6] Of note, these findings are often not present at the same time. The cardiac findings are not part of the diagnostic criteria of KD but are important to support the diagnosis. Cardiac manifestations during the first week to 10 days of illness may include tachycardia out of proportion to the degree of fever with gallop on exam. Pericardial effusion is reported in up to 30% cases.[2] Approximately 30 % of children with KD have coronary artery dilatation at diagnosis, with frank CAA's usually not seen until after day ten of illness.[7] Infants are at a higher risk for CAA's partly because of a delay in diagnosis and treatment.

Adults more frequently present with cervical adenopathy, hepatitis, and arthralgia. In contrast to children, adults are less frequently affected by meningitis, thrombocytosis, and CAA's (5% vs. 18-25%).[8] Another review reported 26% of patients had CA vasculitis, 19 % had CAAs, and 9% had a myocardial infarction.[9]

Patients with KD, should have EKG and echocardiogram. Repeat echocardiograms are usually performed at one to two weeks and again four to six weeks after discharge. Patients without CAAs in the first month after onset of KD and without recurrent symptoms do not need further cardiac testing. Coronary computed tomography (CT) and MRA are useful options, with multimodal imaging often required to accurately evaluate and manage CAAs. Patients with coronary aneurysms undergo stress testing with myocardial perfusion imaging on a regular basis to evaluate for inducible ischemia.

Treatment with IVIG within the first ten days of illness reduces the risk of CAA's fivefold compared to patients not treated with IVIG.[10] It is important to diagnose KD as soon as possible to reduce the risk of CAA's.[5] The standard initial treatment for KD includes use of IVIG with aspirin. Steroids are recommended for high-risk patients. Statins, due to their pleitropic anti-inflammatory effects are used in most KD- induced CAA's.[11]

Conclusion

CAA's secondary to delayed diagnosis Kawasaki Disease are infrequent. Diagnosis can be challenging due to non-specific diagnostic criteria and lack of specific testing. Timely identification is important for effective treatment.

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