A previously healthy 9 month old boy suffered an acute cardiac arrest at home after a few hours of irritability and inconsolability. His mother called 911 and initiated cardiopulmonary resuscitation (CPR). She continued CPR for 8-10 minutes until paramedics arrived. His initial rhythm was asystole. An intraosseous line was placed and 5 doses of epinephrine administered. Simultaneously, his trachea was intubated. Subsequently, he was found to be in ventricular fibrillation and defibrillation was provided twice. CPR was continued and he had pulseless electrical activity upon arrival in the Emergency Department. One additional dose of epinephrine was administered and spontaneous circulation returned. Total time without spontaneous circulation was at least 30 minutes. After initial stabilization, work up to determine the etiology of his cardiac arrest was initiated. ECG demonstrated deep Q waves in the anterior and lateral leads and ST elevation in the anterior leads. Troponin was markedly elevated at 84.5 ng/ml. Echocardiogram demonstrated severely depressed left ventricular systolic function with apical akinesis along with the findings depicted in Figure 1, specifically thrombosis within a giant left coronary aneurysm.

Further exploration of his history revealed that he had experienced an upper respiratory illness approximately 1 week prior to presentation. Two months prior to presentation, he had high fever for 3 days associated with conjunctivitis, redness of his eyes and cracked red lips.

A presumptive diagnosis of atypical Kawasaki disease (KD) was made and treatment was initiated with intravenous immunoglobulin (IVIG). He was anticoagulated with heparin and systemic thrombolysis with tissue plasminogen activator was instituted. Troponin peaked at 106.4 ng/ml.

Six days after presentation he underwent angiography which revealed complete occlusion of the left anterior descending coronary artery (LAD) proximal to the diagonal branch with collateralization from the right coronary artery (RCA) to the distal LAD and from the left circumflex artery (LCX) to the distal diagonal branch. Coronary angiography was repeated 3 days later, due to echocardiographic evidence of increasing left ventricular size and increasing mitral regurgitation, and demonstrated partial recanalization of the giant aneurysm with reflow to the LAD and diagonal branch.

One month after presentation, he demonstrated electrocardiographic and laboratory signs of new anterolateral wall ischemia. Repeat angiography demonstrated complete occlusion of the distal end of the giant aneurysm in the LAD distal to the diagonal branch. Coronary balloon angioplasty of the obliterated segment was performed without reflow but angiography the following day showed recanalization with flow in the LAD. However, akinesis of the apical, septal and lateral left ventricular wall segments persisted, resulting in listing for heart transplantation. Exactly 3 months after presentation, he underwent successful orthotopic heart transplantation. He was discharged to home 3.5 weeks later.

DISCUSSION

Coronary artery aneurysms (CAA) occur in about 15-25 % of untreated children with KD, and in about 5% despite appropriate treatment with IVIG in the first 10 days of illness1. Giant coronary artery aneurysms (GCAA), defined as aneurysms greater than or equal to 8 mm in diameter, are rare but dreaded sequelae of KD occurring in 0.3 -1% of cases despite appropriate treatment [1,2]. They are associated with the highest rate of progression to stenosis and the worst prognosis among patients with KD. These aneurysms are predisposed to thrombosis due to slowing of blood flow in combination with large dilated vessels and frequent occurrence of stenosis at either end of the aneurysm1.

Epidemiological studies have shown that specific risk factors associated with (but not causally linked to) the development of GCAA include age younger than 1 year or older than 5 years, additional administration of IVIG (beyond the first dose), total dosage of IVIG exceeding 2500 mg/kg, duration of IVIG administration greater than 3 days, and steroid administration with or without additional administration of IVIG [2]. Thrombus prevention with a combination of low dose aspirin and warfarin is recommended along with interval cardiac evaluation including echocardiography and cardiac catheterization with selective coronary angiography [1].

In a large Japanese study, cardiac event free rate and survival rate at 30 years after KD in patients with giant aneurysms was
found to be 36% and 90% respectively [3].

Kawasaki disease should be considered in the differential diagnosis of children presenting with evidence of myocardial infarction. Echocardiography with careful attention to the coronary arteries should be pursued. Our case highlights that GCAA can demonstrate reocclusion after initial recanalization, despite adequate anticoagulation, and thus require a high level of vigilance once detected.

REFERENCES


Cite this article