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An Atypical Presentation of Incomplete Kawasaki Disease

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Background:
Kawasaki disease (KD) is a systemic panvasculitis that can cause coronary artery aneurysms (CAA) in up to 25% if left untreated. Rarely, aneurysms of other medium-sized arteries have also been reported to occur. The incidence of systemic artery aneurysms (SAA) with typical KD can be as high a 2.2%. Incomplete KD with SAA is not well described. We report a case of diffuse SAA in a 12-year-old boy with incomplete KD and giant CAA.
Case Presentation:
The patient presented with fever, malaise, abdominal pain, rash and cervical lymphadenopathy. Suspicion for the presence of a pericardial effusion on an abdominal CT scan prompted echocardiographic assessment. By echo, multiple giant CAA in all three coronary arteries was noted. Incidentally, on an aortogram after performing pericardiocentesis, it was noted that he had diffuse ectasia and aneurysms of every arterial branch off the aorta. Selective angiograms confirmed the presence of aneurysms in all medium-sized arteries throughout the body including pulmonary arteries. The patient was treated with IVIG, methylprednisolone and high dose aspirin. Incomplete KD was suspected. However, because of the systemic vasculitis, cyclophosphamide therapy was administered. Patient responded well to therapy with improvement in clinical symptoms. Anticoagulation with heparin was transitioned over to maintenance warfarin therapy. Patient remains asymptomatic 2 years later with large, but stable CAA.
Conclusions:
Incomplete KD can manifest with giant CAA and SAA. Immunosuppressant therapy may be necessary for systemic involvement. Anticoagulation is needed to prevent thromboembolic manifestations.

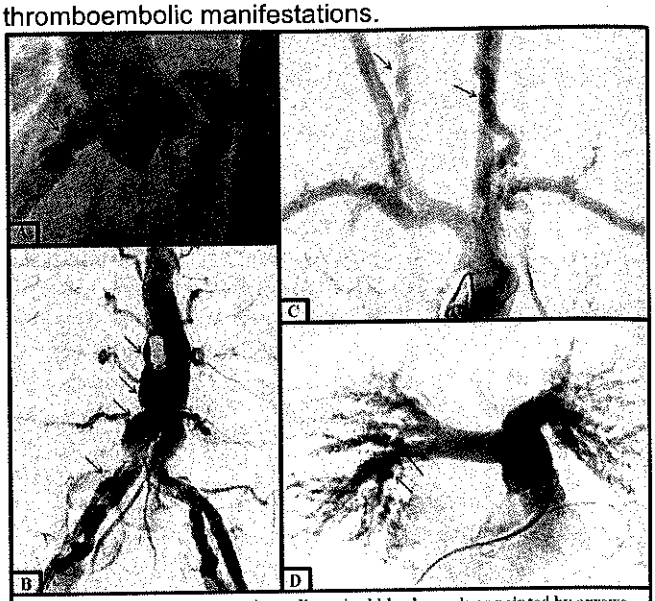

Figure illustrating aneurysms in medium sized blood vessels as pointed by arrows.

A. Right and Left coronary arteries
B. Descending Aorta, Iliac and Lumbar arteries
C. Vertebral arteries
D. Branch Pulmonary Arteries

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Is Lymph-Node-First Kawasaki Disease a High-risk Group for Coronary Aneurysm?

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Cervical lymphadenopathy (CLA) is the least common main feature in patients with Kawasaki disease (KD), comprising only 42%-65% of all diagnoses. Nonetheless, several studies have shown that KD patients who first present with remarkable CLA and fever (NF-KD) are older in age and exhibit stronger inflammation than that of typical KD (tKD) patients. Therefore, whether NF-KD patients are also a high-risk group for intravenous immunoglobulin resistance (rIVIG) and coronary arterial aneurysm (CAA), and whether tKD with CLA (tKD-CLA) is associated with higher inflammatory indices than tKD without CLA warrants investigation. Previous retrospective (R) and prospective (P) studies have shown varied results (Table). In this study, we reviewed 10 years of medical records from a tertiary referral hospital and identified 42 NF-KD patients. These NF-KD patients were then