Coronary Artery Aneurysms After Adult-Onset Kawasaki Disease

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In May 2012, we encountered a rare case of adult-onset Kawasaki disease (KD) with coronary artery abnormalities in a 24-year-old Japanese man. The patient presented with recent-onset chest pain that occurred when he was at rest and no other symptoms. His medical history included hospitalization 2 years before the present admission, at another hospital, because of a fever of unknown origin that lasted for 2 weeks, accompanied by polymorphic exanthema, cervical lymphadenopathy, nonpurulent conjunctivitis, strawberry tongue, and fingertip desquamation. At that time, the patient was diagnosed with adult-onset Still’s disease. On the basis of that diagnosis, he was treated with steroid pulse therapy alone, and his symptoms were alleviated. After 2 months, he received oral steroid therapy and was assumed to have been treated successfully; however, the patient was admitted to our hospital 20 months after concluding the oral steroid therapy.

On admission to our hospital, KD was suspected because of his past history of “strawberry tongue,” which is unique to this disease.1 The chest pain was associated with an abnormal ECG that showed an abnormal Q wave in the inferior (II, III, aVF) ECG zones (Figure 1). A cardiac 3-dimensional computed tomography scan showed an aneurysm of the proximal right coronary artery, with tight postaneurysmal stenosis. The scan also showed proximal aneurysms of the left anterior descending artery and the proximal circumflex artery (Figure 2). He was treated with percutaneous coronary intervention and discharged on day 28 without any complications.

Typical KD findings in both adults and children include fever, conjunctivitis, pharyngitis, skin erythema progressing to a desquamating rash on the palms and soles, and strawberry tongue. Adults more frequently present with cervical adenopathy (93% of adults versus 15% of children), hepatitis (65% versus 10%), and arthralgia (61% versus 24% to 38%). In contrast, adults are less frequently affected by meningitis (10% versus 34%), thrombocytosis (55% versus 100%), and coronary artery aneurysms (5% versus 18% to 25%).2

KD is a common vasculitis of childhood, but adult-onset KD is rare. No specific diagnostic tests are available for KD; the diagnosis is based on the presence of characteristic clinical findings. In addition, a consensus does not exist regarding the treatment of adult-onset KD. The treatment of KD with high-dose intravenous immunoglobulin within the first 10 days of illness reportedly decreases the prevalence of coronary artery aneurysms in childhood. Recently, the incidence of coronary artery abnormalities was reported to be significantly lower in pediatric patients receiving intravenous immunoglobulin plus prednisolone therapy than in those receiving only intravenous immunoglobulin therapy.3 Even patients with adult-onset KD have been reported to benefit from prompt intravenous immunoglobulin administration.4 However, according to one report that examined 91 patients, only 16.5% were diagnosed with adult-onset KD within 10 days of disease onset.4

The low diagnostic accuracy for adult-onset KD, in contrast to that in pediatric patients, can be attributed to the several differential diagnoses that are possible in adult cases.
including drug hypersensitivity reaction and toxic shock syndrome. Other potential diseases with similar symptomology include streptococcal infection (scarlet fever), leptospirosis, *Mycoplasma* and *Rickettsiae* diseases, adenoviral and other viral illnesses (eg, measles, rubella, Epstein-Barr virus infection, and fifth disease), toxoplasmosis, mercury poisoning (acrodynia), and rheumatologic disorders (Reiter syndrome, juvenile rheumatoid arthritis, and Still’s disease). Careful attention to clinical details, such as the nature of the conjunctivitis, the presence of strawberry tongue, and a clinical history of any febrile illness in which the fever persists for >5 days and for which there is no obvious explanation, are required to make a correct diagnosis. Complicating the ability to make a correct clinical diagnosis, a growing body of evidence suggests that a number of children with KD do not fulfill the classic criteria used for KD diagnosis and that a few adult-onset KD cases also do not fulfill the classic criteria.

Once a differential diagnosis excludes the possibility of rheumatologic disorders or autoimmune disease, steroid therapy is often administered; however, treatment with steroids alone might have adverse effects in KD patients, causing progression of coronary lesions. In the present case, only steroid therapy was administered to the patient at the time of his initial hospitalization, 2 years earlier, and this may have resulted in an exacerbation of the patient’s coronary artery abnormalities. Thus, the present report suggests that adult-onset KD should be considered as a differential diagnosis in cases presenting as rheumatologic disorders or autoimmune disease to prevent adverse effects caused by steroid treatment in adult-onset KD patients.

**Disclosures**

None.

**References**

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