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【 CASE REPORT 】

Adult Recurrence of Kawasaki Disease Mimicking Retropharyngeal Abscess

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Abstract:
Kawasaki disease (KD) is an acute, self-limited vasculitis of unknown etiology that occurs predominantly in young children (≤5 years of age). We herein report the case of an 18-year-old Japanese man with a history of incomplete KD during infancy; later, despite an initial diagnosis of retropharyngeal abscess, he was ultimately diagnosed with retropharyngeal edema associated with recurrent KD. Adult-onset or recurrent KD is an uncommon event, and retropharyngeal edema is a rare manifestation of this disease. Internists should be aware of the possibility of KD that mimics a retropharyngeal abscess, even in adult patients.

Key words: computed tomography, magnetic resonance imaging, retropharyngeal cellulitis, retropharyngeal edema, T2 shine through

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Introduction

Kawasaki disease (KD), or mucocutaneous lymph node syndrome, is an acute, self-limited systemic vasculitis of unknown etiology that occurs mainly in infants and young children (≤5 years of age) and predominantly affects individuals in Asian populations (1). KD is associated with various symptoms and is the major sequela of fatal coronary artery aneurysm, the risk of which can be decreased by administering oral aspirin and intravenous immunoglobulin therapy during the acute phase (1). However, patients with KD often develop unusual symptoms such as retropharyngeal edema, which has been recognized by pediatricians and otorhinolaryngologists (2-32).

In contrast to observations in children, first-onset and recurrent KD are rarely seen in adults (1). In the present report, we describe a case of recurrent KD in an adult patient; this condition, which initially mimicked a retropharyngeal abscess, was ultimately diagnosed as retropharyngeal edema.

Case Report

An 18-year-old Japanese man presented to the emergency department of our hospital with a 4-day history of a fever, neck pain, and painful swallowing. At 20 months of age, he had been diagnosed with incomplete KD based on the following findings: a persistent fever, polymorphous exanthema, unilateral cervical lymphadenopathy, and membranous desquamation of the fingertips during convalescence. At that time, he received neither aspirin nor intravenous immunoglobulin therapy. At 14 years of age, coronary computed tomography (CT) angiography revealed his coronary arteries to be normal, and his pediatric outpatient follow-up was terminated.

Upon admission to our hospital, the following data were recorded: temperature, 40.2℃; pulse rate, 96 beats/min; blood pressure, 116/64 mmHg; respiratory rate, 22 breaths per min; and oxygen saturation (digital pulse oximetry), 99% at rest while breathing ambient air. A physical examination revealed oral and pharyngeal mucositis and bilateral erythematous cervical lymphadenopathy. The laboratory findings (Table) revealed elevated levels of inflammatory markers, with no blood chemistry abnormalities, demonstrated by the following values: white blood cell count, 16,000/μL; neutrophil frequency, 89.6%; C-reactive protein level, 26.36 mg/dL (reference, <0.3 mg/dL); procalcitonin level, 1.52 ng/mL (reference, <0.05 ng/mL); soluble

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Table. **Laboratory Findings.**

Hematology		SIL2R	1,286 U/mL
WBC	16,000/μL	ANA	<40
Neu.	89.6%	ASO	46 IU/mL
RBC	485×10⁴/μL	Adenovirus Ab (CF)	(-)
Hb	14.7 g/dL	EBV-VCA-IgM	(-)
Plt	14.0×10³/μL	Cytomegalovirus-IgM	(-)
ESR	66 mm/h	Rubella virus-IgM	(-)
		Measles virus-IgM	(-)
Serology		HIV Ag/Ab Combo (convalescent phase)	(-)
CRP	26.36 mg/dL	Parvovirus B19-IgG (convalescent phase)	(-)
PCT	1.52 ng/mL		
IgG	905 mg/dL	Rapid antigen Group A Streptococcus	(-)
IgA	155 mg/dL		
IgM	138 mg/dL	Blood culture (two sets)	no growth

WBC: white blood cell, Neu.: neutrophils, RBC: red blood cell, Hb: hemoglobin, PLT: platelet, ESR: erythrocyte sedimentation rate, CRP: C-reactive protein, PCT: procalcitonin, Ig: immunoglobulin, SIL2R: soluble interleukin-2 receptor, ANA: antinuclear antibody, ASO: antistreptolysin O, Ab: anitibody, CF: complement fixation, EBV-VCA: Epstein-Barr virus viral capsid antigen, HIV: human immunodeficiency virus, Ag: antigen

interleukin-2 receptor level, 1,286 U/mL (reference, <499 U/mL); and erythrocyte sedimentation rate, 66 mm/h. The serological test results for adenovirus, Epstein-Barr virus, cytomegalovirus, rubella virus, measles virus, human immunodeficiency virus, and parvovirus B19 were negative.

Contrast-enhanced CT revealed multiple areas of cervical lymphadenopathy and low-density areas without ring enhancement in the retropharyngeal space, compatible with a diagnosis of retropharyngeal abscess (Fig. 1A). After admission, intravenous antibiotics were administered without clinical improvement; the negative results of antistreptolysin O titer serum testing and rapid antigen group A streptococcus throat swab testing and lack of bacterial growth on culturing of the patient's blood suggested the absence of a bacterial infection. On day 4 after admission, the patient gradually developed strawberry tongue, conjunctival injection, polymorphous exanthema, and erythema and edema on the hands. Magnetic resonance imaging (MRI) on day 4 revealed a retropharyngeal area that exhibited an isointense signal during a T1-weighted sequence and hyperintense signals during T2-weighted, apparent diffusion coefficient, and diffusion-weighted sequences, which was suggestive of inflammatory edematous changes (Fig. 1B-F). The platelet count increased from 14.0×10^4/μL at admission to a peak of 48.0×10^4/μL on day 11. Coronary CT angiography on day 12 demonstrated dilation of the right coronary and left anterior descending coronary arteries to 5.0 mm (Fig. 2).

Given these imaging findings, clinical features, and disease course, the patient met the diagnostic criteria for KD, and the ultimate diagnosis was an adult recurrence of KD with retropharyngeal edema. The patient had been using the anti-inflammatory drugs acetaminophen and loxoprophen for symptom relief and subsequently switched to a low dose of aspirin (100 mg daily) for 10 weeks because of the antiplatelet effect. During this treatment course, the inflammatory symptoms, laboratory abnormalities, and retropharyngeal edema appeared to resolve spontaneously. A follow-up evaluation at an outpatient clinic confirmed membranous desquamation of the fingertips and palms (Fig. 3), as well as recovery of the coronary arteries that had become dilated during the acute phase.

Discussion

In the present case, the diagnosis of KD was delayed, preventing us from administering intravenous immunoglobulin in a timely manner. As in the present case, early therapeutic intervention is sometimes difficult because there are no specific diagnostic tests for KD, and the diagnostic criteria are limited to clinical signs and symptoms such as i) polymorphous exanthema; ii) conjunctivitis; iii) cervical lymphoadenopathy; iv) oral changes, including an injected pharynx or lips, cracked or fissured lips, and strawberry tongue; and v) extremity changes that initiate with edema or erythema and progress to desquamation of the feet and hands. A diagnosis is possible if an individual with a prolonged (≥5 days) high fever meets at least 4 of the 5 criteria listed above (1). However, some individuals with suspected KD meet fewer than 4 of these criteria and are therefore diagnosed with incomplete KD, which is also associated with a risk of cardiovascular sequelae (1). The patient in the present case had a prolonged high fever, met all 5 of the above-mentioned diagnostic criteria, exhibited transient coronary artery dilation, and had a medical history of incomplete KD; therefore, an adult recurrence of KD was diagnosed.

Adult-onset KD is rare, and adult recurrences of KD are extremely rare. A recent review of the English-language literature from 2015 described only 100 cases of adult onset KD (2) and only 2 cases of recurrence in an adult after childhood presentation among a total of 70 cases of adult

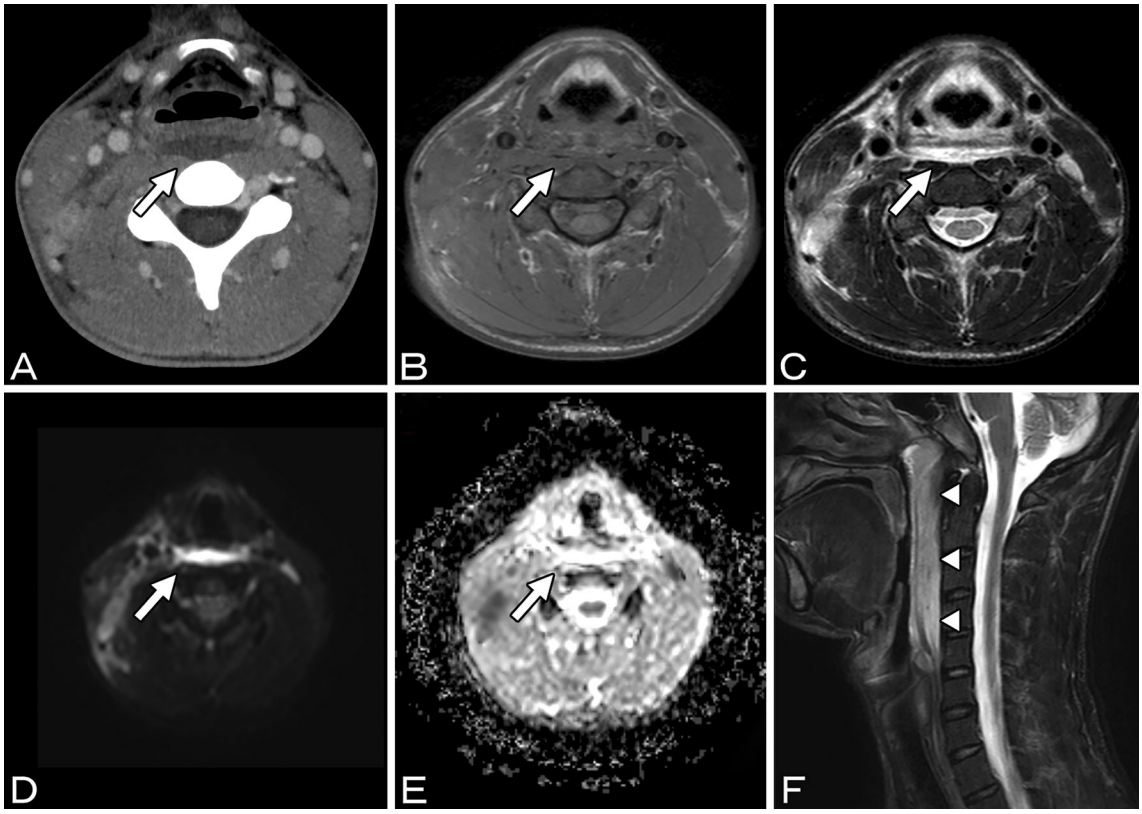

Figure 1. **Axial contrast-enhanced computed tomography reveals a low-density area without ring enhancement in the retropharyngeal space (A, arrow). This retropharyngeal space exhibited isointensity at the C4-5 vertebral level during a T1-weighted MRI sequence (B, arrow) and hyperintensity during T2-weighted (C, arrow), apparent diffusion coefficient (D, arrow), and diffusion-weighted MRI sequences (E, arrow). A sagittal T2-weighted short-tau inversion recovery sequence shows the lesion as a hyperintense area extending from the C1 to C5 vertebral levels (F, arrowheads). MRI: magnetic resonance imaging**

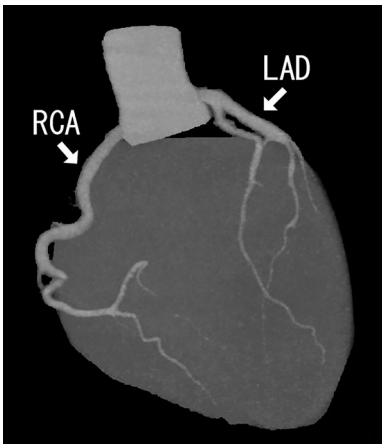

Figure 2. **Coronary CT angiography on day 12 after admission demonstrates dilation of the right coronary and left anterior descending coronary arteries to 5.0 mm. CT: computed tomography, RCA: right coronary artery, LAD: left anterior descending coronary artery**

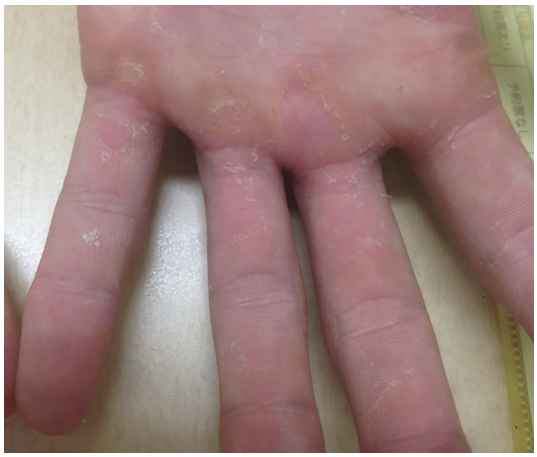

Figure 3. **The patient's left fingertips and palm exhibit membranous desquamation during the convalescent phase.**

KD (3). Furthermore, the frequencies of cardiac involvement and complications are higher among patients with adult-onset KD than among children with KD; such complications affect 44% of adult cases, and 26% develop coronary vasculitis (4).

Retropharyngeal edema, which often mimics retropharyngeal abscess, is a rare complication of KD with various differential diagnoses, including infection of the spaces surrounding the retropharyngeal space, trauma, neoplasm, post-

radiation therapy complications, internal jugular venous thrombosis, systemic fluid overload, or inflammation (e.g. acute calcific longus coil tendinitis and crown dense syndrome) (5). In the present case, all other previously known causes of retropharyngeal edema were ruled out, and this condition was attributed to KD. A review of the English-language literature indicated that this complication, which is generally diagnosed based on CT findings, was first described by Pontell et al. in 1994 (6). Subsequently, the details of similar cases were presented sporadically as case reports. Some recent publications have discussed retrospective studies of neck CT findings in patients with KD, including many cases of KD with retropharyngeal edema. We identified 30 reports involving a total of 157 cases of KD with retropharyngeal edema (7-36). Almost all cases were reported by pediatricians or otorhinolaryngologists, which we attribute to age-related and anatomic factors. All cases occurred in patients younger than 10 years of age, except for a single 14-year-old patient, and the average patient age was 4.8 years (7-36). To our knowledge, no similar reports have described cases in adults, who tend to visit adult primary care physicians such as internal medicine clinicians. Therefore, the present case is the first adult case reported worldwide.

The pathophysiology of KD-associated retropharyngeal edema is unknown. However, inflammation and edema associated with systemic vasculitis and increased microvascular permeability have been suggested as primary mechanisms (37). In the present case, MRI revealed T2 shine-through, or a high signal on DWI images caused by a high T2 signal rather than by restricted diffusion. This phenomenon is most often observed with vasogenic edema. Accordingly, the MRI findings of the present case were compatible with the hypothesized pathophysiology.

In conclusion, adult-onset or recurrent KD is an uncommon entity, and retropharyngeal edema is a rare manifestation of KD. To prevent coronary artery disease and ensure the administration of intravenous immunoglobulin in a timely manner, internists should be aware that KD can mimic a retropharyngeal abscess, even in an adult.

The authors state that they have no Conflict of Interest (COI).

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