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Case report

Retropharyngeal involvement in Kawasaki disease—A report of four patients with retropharyngeal edema verified by magnetic resonance imaging

Laura Puhakka [a,*], Riste Saat [b], Tuomas Klockars [c], Lauri Kajosaari [c], Eeva Salo [a], Tea Nieminen [a]

[a] Children's Hospital, Helsinki University Central Hospital and University of Helsinki, Stenbäckinkatu 11, PL 281, 00029 HUS, Finland
[b] Helsinki and Uusimaa Hospital District Medical Imaging Centre, Helsinki University Central Hospital and University of Helsinki, Finland
[c] Department of Otorhinolaryngology and Head and Neck Surgery, Helsinki University Central Hospital and University of Helsinki, Finland

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ABSTRACT

Kawasaki disease is an acute systemic vasculitis of childhood. The diagnosis is based on clinical criteria. Prognosis with adequate treatment is favorable. Untreated patients, however, may develop coronary manifestations predisposing to acute myocardial infarction. Retropharyngeal edema is a rare but known manifestation of Kawasaki disease. We present a case series of four Kawasaki patients presenting with clinical findings for retropharyngeal abscess and the magnetic resonance imaging findings of these patients, diagnosed during a six week period. To our knowledge, this is the first systematic report of cervical MRI findings of Kawasaki patients.

1. Introduction

Kawasaki disease (KD), also known as mucocutaneous lymph node syndrome, is an acute febrile systemic vasculitis of childhood. The syndrome was first described in 1967 in Japan [1]. KD typically affects young children. The etiology remains unknown, though there are clues to possible infectious etiology. The syndrome tends to occur in clusters and the clinical presentation with rash, fever and lymph node enlargement mimics infectious diseases [2]. About 20% of untreated patients will develop coronary manifestations due to vasculitis. Coronary artery aneurysms are a feared complication that may lead to acute myocardial infarction and death. High dose intravenous immunoglobulin (IVIG) 2 g/kg, and high dose acetylsalicylic acid (ASA) 80–100 mg/kg divided into 4 doses are the treatment of choice for KD [3,4]. Prompt treatment, preferentially within the first week of illness, is effective in reducing the risk of coronary aneurysms in most cases [5].

There are no specific diagnostic tests for Kawasaki disease. The diagnosis is based on the presence of fever ≥5 days and ≥4 out of five principal criteria: (1) changes in the extremities including edema or desquamation, (2) polymorphous exanthema, (3) bilateral nonexcudative conjunctival injection, (4) changes in the lips and oral cavity including fissuring of the lips, strawberry tongue and/or mucosal hyperemia, and (5) cervical lymphadenopathy over 1.5 cm in diameter. Other diseases causing the symptoms must be excluded [6].

Almost half of Kawasaki patients are reported to have arthritis [7]. Early onset arthritis typically affects multiple joints including large and small joints. Arthritis or arthralgia developing after 10th day of illness affects mainly large joints, especially knees and ankles. Other symptoms include diarrhea, vomiting and abdominal pain, pyuria caused by aseptic urethritis, aseptic meningitis, transient peripheral facial nerve palsy, hepatic enlargement and jaundice, and distension of gallbladder. Erythema and induration at the site of bacille Calmette-Guérin (BCG) scar can be seen [6].

As many of the manifestations occur in the head and neck, an otorhinolaryngologist may be the first medical professional to see a patient with KD. Kawasaki disease may begin with fever and cervical adenopathy with other clinical signs appearing later. In a series of 340 patients in California 16% had lymph node presentation as the first manifestation of Kawasaki disease [8]. Retropharyngeal edema is a rare but known presentation of KD and it may be misdiagnosed as a retropharyngeal abscess. There are several reports of Kawasaki disease presenting with symptoms

* Corresponding author. Tel.: +358 50 4288006.
E-mail address: laura.puhakka@hus.fi (L. Puhakka).

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resembling deep neck infection. CT findings of these patients are well reported but systematic information about MRI findings in patients with Kawasaki disease mimicking retropharyngeal abscess is scarce [9–26].

In this article we review the literature of KD resembling retropharyngeal abscess, report the MRI findings in a series of four KD patients with retropharyngeal edema and discuss the role of MRI in Kawasaki disease imaging.

2. Case series

2.1. Patient 1

A 4 y 8 month old boy of Congolese descent presented in the end of January 2013 with one day history of fever (39.1 °C) and neck pain. On the day of admission he had progressive torticollis, decreased neck movements and trismus. No enlarged lymph nodes were palpable in the neck. CRP was elevated 188 mg/L and white blood cell count was normal 10,000 μL^{-1}. Rapid antigen test as well as culture for group A streptococcus from the throat swab was negative.

In MRI of the neck a retropharyngeal rim-enhancing fluid collection from C1 to C3 (4 cm × 2 cm × 0.5 cm) was suspected, suggestive of retropharyngeal abscess. An enlarged lymph node with rim enhancement was found in left parapharyngeal space.

The treatment was initially conservative with iv cefuroxime and metronidazole, but on the 5th day of illness operative measures were initiated due to lack of clinical improvement and progression of trismus. Tonsillectomy, adenoidectomy, and exploration of retropharyngeal space were performed, but no abscess was found. Bacterial, fungal and tuberculosis samples were collected. Only scarce growth of acinetobacter species was detected in retropharyngeal space samples. This was considered contamination with no clinical significance.

Typical Kawasaki symptoms evolved during the next few days (days 4–8). In addition to high fever (40.5 × °C highest) the patient developed nonpurulent conjunctivitis, fissured lips, mild edema of the palms and mild erythematous rash on his upper trunk. Uveitis was confirmed on ophthalmologic examination. KD diagnosis was set on 8th day of illness and the patient received high dose IVIG and high dose ASA. The symptoms resolved dramatically after IVIG infusion. Fever subsided and other clinical symptoms resolved. In cardiologic follow-up a mild dilatation in the LCA (3 mm) was observed two weeks after the onset of the disease, but in later follow-ups normal status was regained.

2.2. Patient 2

The second patient attended the clinic only two and a half weeks after the first patient, in mid-February 2013. A 1 y 2 month old Finnish boy presented with one day history of fever and torticollis. On arrival he had fever of 38.9 °C, left sided acute otitis media, and cervical lymphadenitis. Erythema in the pharynx and mild exanthema on his upper body was seen. The white blood cell count was 20,600 μL^{-1} and CRP 72 mg/L. Rapid antigen test for group A streptococcus and culture from throat swabs were negative.

Lymphadenopathy was confirmed by ultrasound performed on the day of admission. No abscess was seen. MRI was performed on 5th day of illness and revealed retropharyngeal edema and enhancement suspicious for a developing abscess, shown in Fig. 1. Cervical lymphadenopathy was seen on both sides.

Bacterial lymphadenitis was initially suspected and treated with iv cefuroxime. In spite of combining clindamycin to the treatment, the patient remained febrile (highest 40.2 °C) and his CRP value increased to 135 mg/L. On the 5th day of the illness when inducing general anesthesia for the MRI scan, it was observed that the patient could only be ventilated with his head laterally tilted. The posture-dependent obstruction of the airways was due to retropharyngeal edema. After MRI the patient was transferred to intensive care unit. The edematous area suspected to be an abscess was aspirated with a needle the following day, but no fluid collection was found. The patient was subsequently extubated with no further breathing difficulties.

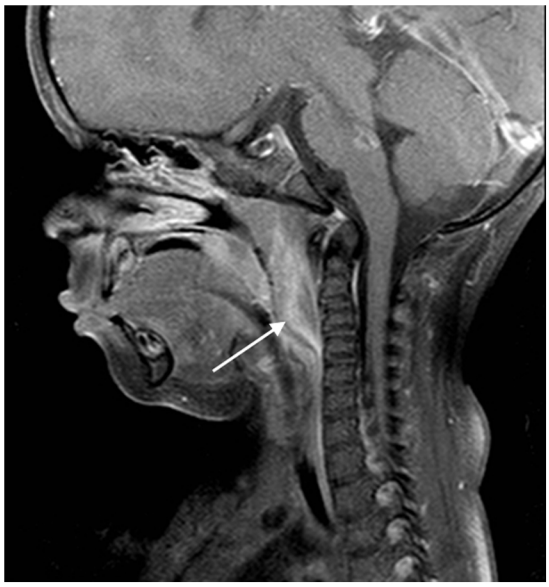

Fig. 1. Sagittal post-Gd T1fs MR-image of patient 2 shows a prominent retropharyngeal fluid collection with peripheral enhancement (arrow) obstructing the airways. The imaging finding is identical to that of a retropharyngeal abscess.

The patient developed classic Kawasaki symptoms: on the 6th day of illness he had in addition to lymphadenitis and fever also erythematous rash, nonpurulent conjunctivitis, red fissured lips and strawberry tongue, and edema in the fingertips. CRP increased to 207 mg/L and erythrocyte sedimentation rate was 93 mm/h. Echocardiogram was normal. The patient received high dose IVIG and ASA followed by rapid clinical improvement.

2.3. Patient 3

The third patient, a 6-y-old Finnish boy, was referred to our clinic in the beginning of March 2013 with a 6-day history of neck pain and a 5-day history of mild fever. Oral antibiotics for cervical lymphadenitis had been prescribed, but the symptoms progressed and a tender mass developed in the neck. On arrival the patient had mild fever (38.1 °C), torticollis, cervical lymphadenopathy and trismus. The history and clinical findings raised a suspicion of a deep neck infection. CRP at presentation was 68 mg/L and white blood cell count was 10,600 μL^{-1}.

Reactive enlarged lymph nodes with no abscess formation were seen on ultrasound. MRI of the neck on 6th day of fever revealed cervical lymphadenopathy and retropharyngeal edema susceptive for a bacterial infection (Fig. 2).

The patient received intravenous cefuroxime and clindamycin, but continued to have fever spikes up to 38.5 °C. The lymphadenopathy in the cervical area persisted and progressive erythema developed on the cervical skin.

On the 8th day of fever the patient fulfilled the remaining Kawasaki criteria: nonpurulent conjunctivitis, erythematous rash and swollen lips. Uveitis was diagnosed by ophthalmologic examination. After KD diagnosis was confirmed he received high