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Case report

Atypical presentation of Kawasaki disease resembling a retropharyngeal abscess

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= Abstract =

Kawasaki disease is an acute systemic inflammatory disorder, the etiology of which has not yet been established. The clinical manifestations are non-specific and are common to many pediatric infectious and immunologic diseases. In 2 cases presenting fever, cervical lymphadenopathy, and retropharyngeal abscess-like lesions on the neck shown in a computerized tomography (CT) scan, the diagnosis of Kawasaki disease was delayed. The patients were not responsive to antibiotic therapy, and fluid collection was not found during retropharyngeal aspiration. They showed typical Kawasaki manifestations over time and atrioventricular valve regurgitation in the echocardiogram. A diagnosis of Kawasaki disease should be considered in similar cases of fever and cervical lymphadenopathy. A CT scan that shows a retropharyngeal abscess-like lesion on the neck can be a false positive finding, and other laboratory exams, including echocardiography, should be performed as early as possible to avoid unnecessary medical and surgical procedures. (Korean J Pediatr 2009;52:251-255)

Key Words : Mucocutaneous lymph node syndrome, Retropharyngeal abscess

Introduction

Kawasaki disease (KD) is an acute systemic vasculitis of unknown cause, that primarily affects children under 5 years of age. It has been reported worldwide and is now the leading cause of acquired heart disease in children in the developed world[1]. The clinical findings of KD are nonspecific and are commonly found in many pediatric infectious and immunologic diseases. Further complicating the diagnosis is the fact that these clinical features may be absent or may evolve over many days after the onset of fever[2]. Therefore, KD presents a diagnostic challenge, and a high index of suspicion is required for early diagnosis and treatment of KD. In the present report, we describe two cases of KD initially presenting as retropharyngeal abscess, with a review of the literature.

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Case report

Case 1

An 18-month-old Korean boy presented with a 2 day history of fever, irritability and postauricular swelling. A medical history of retropharyngeal abscess and Kawasaki disease had been recorded. At 3-months of age, he was admitted for fever and right cervical lymphadenopathy. The neck computed tomography (CT) scan showed a low density lesion in the retropharyngeal space suggesting an abscess, and multiple lymph node enlargements in the right posterior cervical space (Fig. 1). With antibiotics treatment, the fever subsided on the 7th hospital day, and the cervical lymphadenopathy was improved. At 8 months of age, he was admitted for typical KD and showed prompt improvement after treatment with intravenous immunoglobulin (IVIG) and aspirin. During the convalescent period, he developed peeling of the fingertips, and the mild dilation of the left main coronary was normalized.

At 18 months of age, on examination in the emergency department, the patient had a high temperature. He also had an erythematous rash on his trunk, dry lips, and an oropha-

ryngeal injection. Bilateral firm enlargement of the postauricular cervical lymph nodes was noted. Although tender to palpation and overlying erythema, none of the masses were fluctuant.

The white blood cell count was measured at 14,500/μL with 90% neutrophils and toxic granules. C-reactive protein was increased to 16.52 mg/dL. Liver enzymes, such as AST/ALT, were elevated to 186/204 IU/L. Blood and urine cultures were negative. Serology for Cytomegalovirs (CMV) and Epstein Barr virus (EBV) were negative. Neck CT scan revealed a low-density mass without wall-enhancement in the retropharyngeal space (Fig. 2). Though intravenous hydration and antibiotics such as augumentin, netilmycin and metronidazole were started, fever was persistent.

On the 3rd hospital day, he developed a skin rash, conjuntival injection, red lips, strawberry tongue, and swelling of the hands and feet. IVIG with a high dose of aspirin was prescribed, which resulted in prompt improvement. On the 5th hospital day, the fever reappeared with persistent Kawasaki manifestations. The neck CT showed improvement in the retropharyngeal lesion, but mild mitral regurgitation was noted on the echocardiogram. IVIG was administered again, but fever persisted untill the 8th hospital day, when the 3rd IVIG was given. On the 16th hospital day, desquamantion of the fingertips was noted.

Case 2

A 6-year old Korean boy was admitted to the emergency room with fever and painful neck swelling. On physical examination, his tonsils were hypertrophied and his left infra-auricular lymph node had swollen to a 2×3 cm size. The lymph node was neither fixed nor fluctuant. The white blood cell count was 37,300/μL with 92% neutrophils. Hemoglobin was 12.6 g/dL and the platelet count was 251,000/μL. C-reactive protein was increased to 20.3 mg/dL. Liver enzymes were not elevated. Blood and urine cultures were negative. The neck CT scan revealed a low density lesion in the widened retropharyngeal space and multiple lymph node enlargements in the left infra-auricular area (Fig. 3). Intravenous hydration and antibiotics including augumentin, netilmicin and metronidazole were given at the Otolaryngology department, but the fever was persistent. On the 3rd hospital day, adenoidectomy and retropharyngeal aspiration were performed, but aspirate was absent. On the 8th hospital day, a generalized skin rash, non-purulent conjunctival injection, red lips, and strawberry tongue developed. A cardiac echocardiogram revealed slightly decreased fractional shortening (22%), mitral regurgitation of grade II-III/IV, tricuspid regurgitation of grade III-IV/IV, but the coronary arteries were not dilated. After IVIG infusion and aspirin prescription,

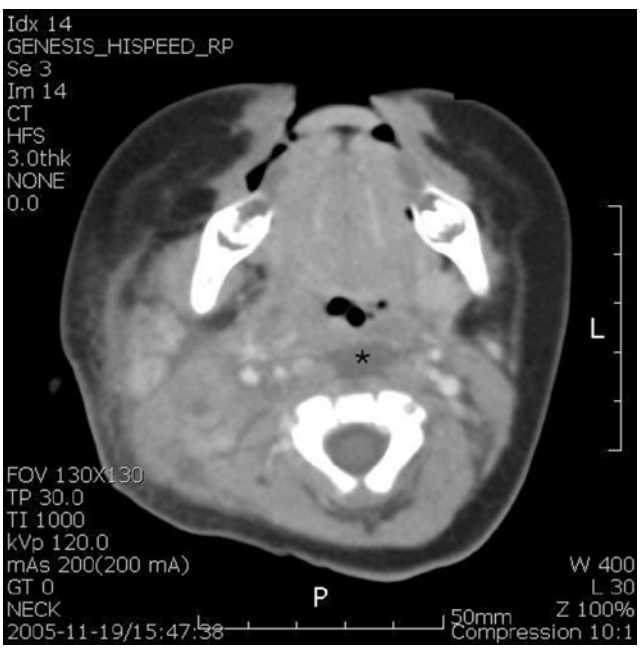

Fig. 1. Neck CT scan showing a low-density lesion in widened retropharyngeal space (*) and right cervical lymphadenopathy at 3 months of age in case 1.

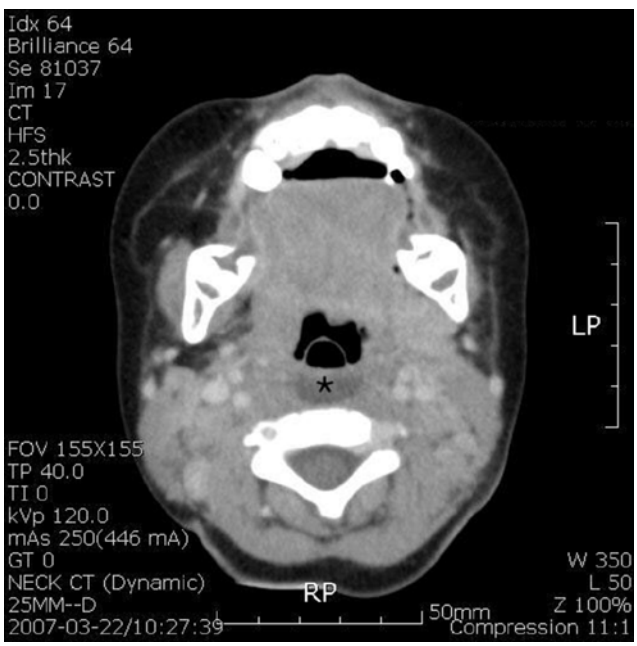

Fig. 2. Axial CT scan demonstrates a low-density lesion (*) in widened retropharyngeal space without contrast enhancement and bilateral cervical lymphadenopathy in case 1.

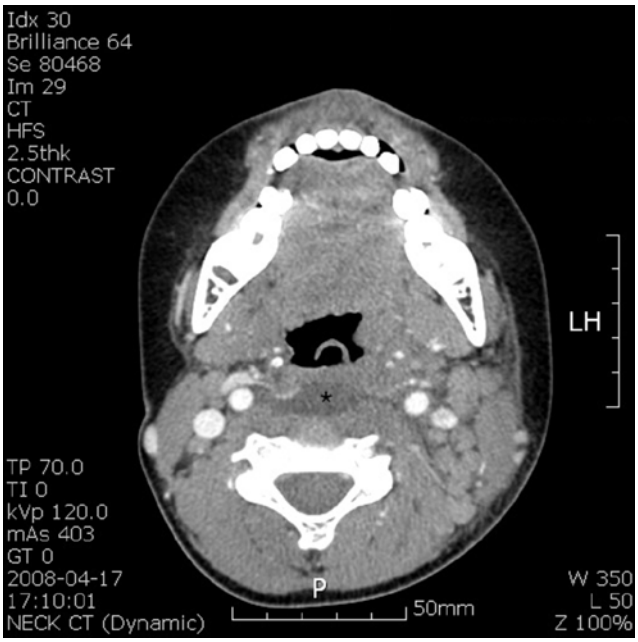

Fig. 3. Neck CT scan showing a low-density lesion (*) in widened retropharyngeal space and left cervical lymphadenopathy in case 2.

the fever subsided the following day. Four weeks later, a follow-up cardiac echocardiogram showed normal cardiac function and normal coronary artery size without valve regurgitation.

Discussion

Case 1 is peculiar in some aspects. Firstly, he suffered from retropharyngeal abscess and Kawasaki disease separately 5 months apart during infancy. Secondly, KD recurred almost 10 months later with the presentation of a retropharyngeal abscess on the CT scan. Thirdly, fever with cervical lymphadenopathy was the presenting symptom for each of the three illnesses. Case 2 was diagnosed after a week of illness because of the delayed appearance of Kawasaki manifestations.

Currently, diagnosis of KD is based on the same clinical criteria used to describe the disease over 30 years ago. However, non-specific clinical features, evolving presentations, and atypical or incomplete presentations make early diagnosis and timely treatment difficult. A recent nationwide study in Canada reported that delays in diagnosis and treatment, which occur more frequently in older children, are associated with an increased risk of coronary aneurysm, hence, high diagnostic suspicion and prompt referral are required to reduce the rates of cardiac complications[2]. There are some studies regarding the difficulties in clinical diagnosis of KD after the first week of illness. In an analysis of the diagnosis of KD after illness day 10, the delayed diagnosis group exhibited typical features dispersed over time, as opposed to the close clustering of symptoms in the early diagnosis group[3]. In addition, coronary aneurysms occurred more often in the delayed diagnosis group, for this reason, a high index of suspicion is needed in young children presenting with fever and rash illnesses[3]. From the retrospective analysis of 25 delayed diagnosis cases that were diagnosed in the second week, KD should be strongly considered in a child presenting with fever for more than one week, who looks "non-toxic" (not very sick), and who has non-purulent bulbar conjunctivitis and oral mucositis[4].

Case reports suggest that infants with KD have atypical presentations and high complication rates, therefore, criteria are too restrictive to allow early diagnosis and effective treatment[5]. Some authors advocated routine use of echocardiography for early detection of KD and initiation of early prophylaxis in patients with features suggestive of KD, even if they do not fulfill all of the criteria to be labeled KD[6].

In 12% of the KD with head and neck manifestations, cervical lymphadenopathy is the initial presenting symptom, which may cause a significant delay in diagnosis[7]. In the recent review of 5 KD cases presenting with acute tonsillitis, initial manifestations were polymorphous rash in 1 case, and cervical lymph nodes in 4 other cases. All of the patients were later diagnosed as KD on the 8th to 28th day of illness[8]. Otolaryngologists should maintain a high index of clinical suspicion for KD, because of its ability to mimic acute suppurative processes and the potential for cardiac complications with delayed diagnosis or unnecessary surgical intervention[9].

Among the head and neck manifestations, a deep neck space infection-like symptom occurred in 8 out of 155 KD cases including 17 atypical cases[7]. A Medline search revealed at least five reported cases of KD with a tentative diagnosis of a retropharyngeal abscess[9-13]. The clinical characteristics of previously reported cases along with those of our cases are summarized in Table 1. In all cases, the CT scan revealed hypodense lesions with or without enhancement, compatible with a retropharyngeal abscess. Thus all of the cases, including our cases were initially treated as a retropharyngeal abscess, but showed no improvement with antibiotics. A correct diagnosis was eventually made in all cases

Table 1. Clinical Characteristics of Kawasaki Disease Resembling Retropharyngeal Disease (Literature Review and Our Cases)

Source	Age (yr)/ Sex	Initial Sx	Neck CT	Days for Dx of KD	Surgical finding	Echo finding
Park[9] 1997	4.5/M	Fever for a week, treated as otitis	Left RP mass extending laterally into neck, several small areas of hypolucency within the mass and associated ring enhancement	9	Not done	Normal
McLaughlin[10] 1998	4/M	Irritability for 2 days, fever, neck swelling, decreased neck motion	Extensive RP edema crossing the midline and extending from the level of nasopharynx inferior to thoracic inlet	5	Not done	Normal
Homicz[11] 2000	6/F	Fever for 2 days, neck pain, torticollis, odynophagia	Low density mass without enhancement in RP space	4	Aspiration with no abscess	Normal
Gross[12] 2001	9/M	Fever for 7 days, torticollis, cervical lymphadenopathy	RP soft tissue swelling	22	Incision with no fluid collection	6-8 mm dilated and ectatic coronary a
Hung[13] 2006	2.4/M	Fever for 2 days, neck swelling	Low density lesion in RP space	3	Not done	Normal
Case 1	1.5/M	Fever for 2 days, irritability, postauricular swelling	Low-density lesion without wall-enhancement in RP space	5	Not done	MR
Case 2	6.0/M	Fever for 1day, neck swelling	Low density lesion in RP space	9	Aspiration with no abscess	MR, TR

Abbreviations : Dx, diagnosis; KD, Kawasaki disease; MR, mitral regurgitation; RP, retropharyngeal; TR, tricuspid regurgitation

when additional signs and symptoms of KD developed. In four cases diagnosed within a week of onset of symptoms, coronary problems were absent. However, in one case, a patient was diagnosed on the 22nd day of illness and coronary aneurysm was detected[12]. Surgical intervention was performed in three cases, including case 2 in the present report, but there was no abscess or fluid collection. It appears that the CT scan cannot differentiate abscess from inflammation. There is still no reported coexistence of KD and a retropharyngeal abscess in the previous reports. Once a patient is diagnosed with KD, the abscess-like lesion on the CT scan should be viewed as a type of inflammation rather than as an infection[13]. Although KD mimicking a deep neck infection appears to be rare, the true incidence of retropharyngeal pathology is unknown because of the lack of routine neck imaging. Hence, the likely prevalence of unreported cases is also unknown[11].

Case 1 was very resistant to IVIG, so IVIG was administered three times. For cases with IVIG-resistance, that have a persistent or recrudescent fever, rescue therapies include multiple courses of IVIG, pulse dose methylprednisolone as well as Infliximab[14, 15]. Infliximab, a TNF-α antagonist was successfully used for a KD patient with a giant aneurysm unresponsive to multiple doses of IVIG and methylprednisolone[16]. In the present case 1, even though the patient was diagnosed as KD, we could not use steroids because of the possibility that a retropharyngeal abscess might coexist.

In conclusion, KD may be initially misdiagnosed as a retropharyngeal abscess by neck CT scan in a febrile child with cervical lymphadenopathy. If the fever is refractory to antibiotic treatment in such a patient, the possibility of KD should be considered, Echocardiography should be performed as early as possible and unnecessary surgical intervention should be avoided.

한 글 요 약

인두후부의 농양과 유사하게 표현된 비전형적인 가와사끼병

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가와사끼병은 급성 전신적 염증성 질환으로, 그 원인은 아직 밝혀지지 않았고, 임상양상은 비특이적으로 많은 소아의 감염성 질환이나 면역학적 질환에서도 볼 수 있다. 저자들은 발열과 경부 임파선병증을 보인 두 증례에서 경부 전산화단층촬영상 인두후부의 농양과 유사한 소견을 보였으나 항생제 치료에 반응하지 않으며 인두 후부의 흡인술에서도 농양이 없던 경우로, 시간이 지나면서 전형적인 가와사끼 증상의 출현과 함께 심초음파에서 방실 판막의 역류를 보여 늦게 진단할 수 있었다. 본 증례에서처럼 발열과 경부임파선병증으로 나타나는 경우에도 가와사끼병을 의심해야 하며, 이 경우 전산화단층촬영에서 인두후부의 농양 유사 병변은 위양성 소견으로 볼 수 있다. 경부임파선병증이 적절한 항생제치료에도 불구하고 발열 등의 증상이 지속되는 경우에는, 심초음파를 포함한 검사를 조기에 시행하여 불필요한 내과적,

외과적 치료를 줄여야 할 필요가 있겠다.

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