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■ Case report ■

Kawasaki disease presenting as retropharyngeal abscess

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= Abstract =

A group of patients with Kawasaki disease (KD) initially present with cervical lymphadenitis or deep neck infection. These unusual KD presentations lead to unnecessary antibiotic therapy or surgical intervention, thereby delaying intravenous immunoglobulin treatment and increasing the risk of coronary artery damage. We present four KD patients whose initial presentations mimicked a retropharyngeal abscess. Nonsuppurative cervical lymphadenitis or suspected neck abscess unresponsive to intravenous antibiotics could signal the possibility of KD. **(Korean J Pediatr 2008;51:1023-1027)**

Key Words : Kawasaki disease, Retropharyngeal abscess

Introduction

Kawasaki disease (KD) is an acute, multisystemic vasculitis of unknown cause that primarily affect infants and young children. Diagnosis is based on the clinical features, but there are many head and neck manifestations. The KD patients with multiple head and neck manifestations, such as cervical lymphadenopathy, stomatitis, and pharyngitis, usually visit or are referred to an otolaryngologist[1]. Involvement of deep cervical lymph nodes produces large, hypodense lesions on computerized tomography (CT) scan, which may lead to a misdiagnosis of deep neck abscess and subsequently unnecessary antibiotic therapy or surgery.

We describe here four KD patients who were initially diagnosed with retropharyngeal abscess and failed to respond to intravenous antibiotics, but were successfully treated after the correct diagnosis was established (Table 1).

Case Reports

Case 1

A 5-year-old boy was first seen by an otolaryngologist with a 4-day history of fever and a 3-day history of left neck swelling. He was febrile and complained of left neck pain and hoarseness. The neck mass was firm, nonfluctuating, and tender to palpation, measuring 6×5 cm. Laboratory measures are summarized in Table 2. A lateral neck X-ray showed widening of the retropharyngeal space. A contrast neck CT scan was obtained on hospital day 1. The scan revealed a 9.5-mm low density mass without wall enhancement, centrally located in the retropharyngeal space suggestive of retropharyngeal abscess (Fig. 1). Because of a lack of clinical improvement on intravenous antibiotics, surgical intervention was planned. On hospital day 3, he developed bilateral conjunctival injection and erythematous maculopapular rash on his trunk. His lips became dry and fissured, and he developed strawberry tongue and erythematous palms and soles on hospital day 4. On hospital day 5, under the diagnosis of KD, he was referred to us and was immediately given intravenous immunoglobulin (IVIG). Defervescence and clinical improvement occurred after IVIG treatment (Fig. 2). There was no coronary artery abnormality on echocardiography.

Case 2

A 7-year-old boy was referred to us after presenting with fever for 4 days and right neck swelling for 3 days. He was diagnosed with cervical lymphadenitis and oral cephalosporin was given at a primary care clinic, but his illness worsened. On admission, an erythematous tender cervical lymph node, measuring 5×5 cm, was noted on his right neck and several smaller lymph nodes on the left neck. Laboratory findings

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Table 1. Clinical Features of Four Kawasaki Disease Patients Presenting as Retropharyngeal Abscess

Patient	1	2	3	4
Age (yr)/Sex	5/M	7/M	2/M	9/M
Cervical mass				
Localization/Size (cm)	L/6×5	R/5×5	R/4×5	L/15×15
Erythema/Tenderness	−/+	+/+	−/+	+/+
Low density on neck CT				
Thickness (mm)	9.5	5.9	5.5	9.8
Laterality	−	−	−	−
Ring enhancement	−	−	−	−
Response to antibiotics	−	−	−	−
Time to appearance of other major symptom	HD 3	HD 3	HD 3	HD 4
Time to IVIG treatment	HD 5	HD 4, 8	HD 3	HD 5
Coronary artery lesion	−	+	−	+
Miscellaneous	Transferred from ENT on HD 5	IVIG− resistance		Transferred from ENT on HD 5

Abbreviations : CT, computerized tomography; ENT, otorhinolaryngology; HD, hospital day; IVIG, intravenous immunoglobulin; L, left; R, right

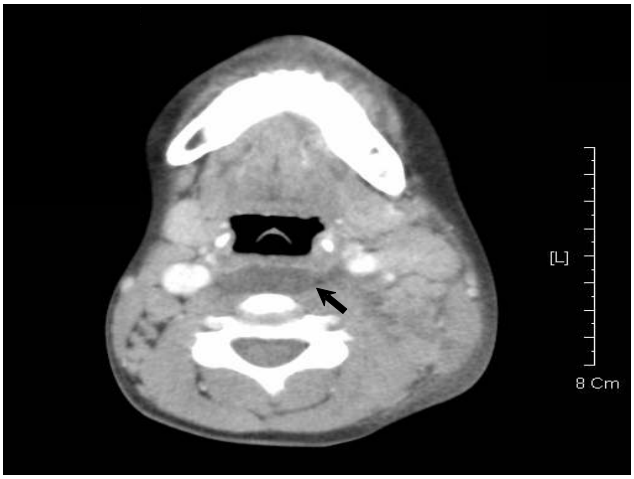

Fig. 1. Axial CT scan demonstrating bilateral cervical adenopathy greater on the left side and a 9.5-mm hypodense retropharyngeal lesion without peripheral enhancement (arrow), suggestive of a retropharyngeal abscess in the case 1 patient.

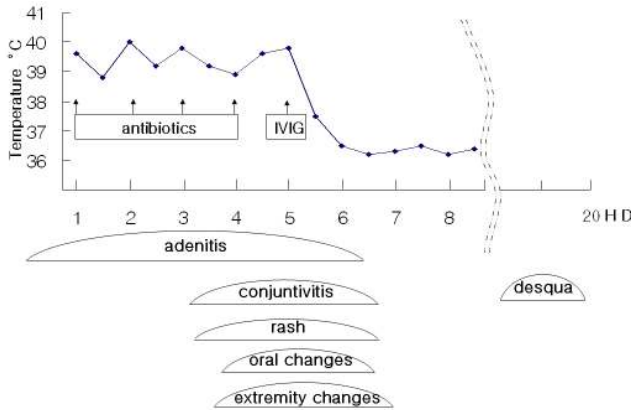

Fig. 2. Changes in body temperature during treatment and symptoms shown time-sequentially in the diagnostic criteria of Kawasaki disease in the case 1 patient. Unilateral neck mass mimicking deep neck infection resistant to antibiotics is the initial presentation. Other clinical features develop gradually at hospital day (HD) 3 or 4. Clinical improvement with defervescence occurs after intravenous immunoglobulin (IVIG) administration.

Table 2. Laboratory Measures of Four Kawasaki Disease Patients at Initial Presentation

Patient	1	2	3	4
Hemoglobin (g/dL)	10.9	12	12.6	12.9
WBC (/mm^3)	15,500	18,700	21,200	25,500
% neutrophil	78	71	85	76
Platelet (/mm^3)	274	293	202	316
CRP (mg/dL)	13.2	18.5	8.4	22.4
ESR (mm/hr)	50	57	60	84
AST/ALT (IU/L)	7/19	22/36	22/27	28/30

Abbreviations : ALT, alanine aminotransferase; AST, aspartate aminotransferase; CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; WBC, white blood cell

are shown in Table 2. The CT scan revealed a hypodense, centrally located retropharyngeal lesion without ring enhancement. He was diagnosed with deep neck infection. Fever persisted despite treatment with intravenous antibiotics for three days. Moreover, he developed erythematous maculopapular rash on whole body, conjunctival injection and erythematous lips on hospital day 3. Anterior uveitis was present on slit-lamp examination. A diagnosis of KD was then established. He was treated with IVIG on hospital day 4, but was resistant, and required a second treatment of IVIG on hospital day 8 and additionally pulse steroid therapy.

Echocardiography revealed a small saccular aneurysm of the left anterior descending artery and ectasia of the right coronary artery. He was maintained on aspirin for one year. The coronary artery lesions eventually regressed to normal size on follow-up.

Case 3

A 2-year-old boy presented with a 3-day history of fever and a swollen lymph node on his right neck. The neck mass, measuring 4×5 cm, was tender to palpation without overlying erythema. Laboratory measures are presented in Table 2. Intravenous cephazolin was started under a diagnosis of cervical lymphadenitis. Due to the absence of clinical improvement, a contrast CT scan of the neck was obtained on hospital day 2. The scan revealed a low density retropharyngeal abscess without wall enhancement. Antibiotic coverage was changed to clindamycin to include anaerobic organisms. However, on hospital day 3, his lips turned red and erythematous maculopapular rash appeared on his whole body. In addition, his palms and soles became swollen and erythematous. He was started on high dose aspirin and IVIG on hospital day 3. After IVIG infusion, his clinical condition improved dramatically with defervescence. No coronary artery lesion was noted on echocardiography.

Case 4

A 9-year-old boy was hospitalized to the otolaryngology due to high fever for 2 days, drooling, and torticollis caused by left neck swelling. The neck lesion, which measured 15 ×15 cm, was diffusely erythematous swollen, hard and fixed with severe tenderness and heating sensation. The white blood cell count was 25,500/mm^3 with C-reactive protein (CRP) of 22.4 mg/dL (Table 2). Neck CT showed a hypodense retropharyngeal abscess without peripheral enhancement and multiple bilateral cervical lymphadenopathy. Intravenous antibiotics was immediately started. Needle aspiration was done in the posterior pharyngeal wall, but no abscess cavity was identified, and no microorganism was cultured from aspirates. Furthermore, no clinical improvement occurred and CRP increased to 26.1 mg/dL despite antibiotic therapy. On hospital day 4, he developed conjunctival injection, redness of lips, a whole body erythematous maculopapular rash, and abdominal pain on right upper quadrant area. Abdominal CT revealed gallbladder empyema, acute cholecystitis, and a tiny splenic abscess. He was transferred to us on hospital day 5, and IVIG was immediately infused. Echocardiography showed dilatation of the right coronary artery, measuring 5 mm and a medium-sized aneurysm of the left anterior descending artery, measuring 6 mm. Following IVIG treatment, fever subsided subsequently, the cervical adenopathy significantly decreased in size, and his torticollis slowly resolved. However, repeat echocardiography on hospital day 20 revealed that the left anterior descending artery lesion changed to a giant fusiform aneurysm measuring 20 mm (Fig. 3). The patient was discharged on aspirin on hospital day 25. He was doing well but was lost to follow up 5 months later.

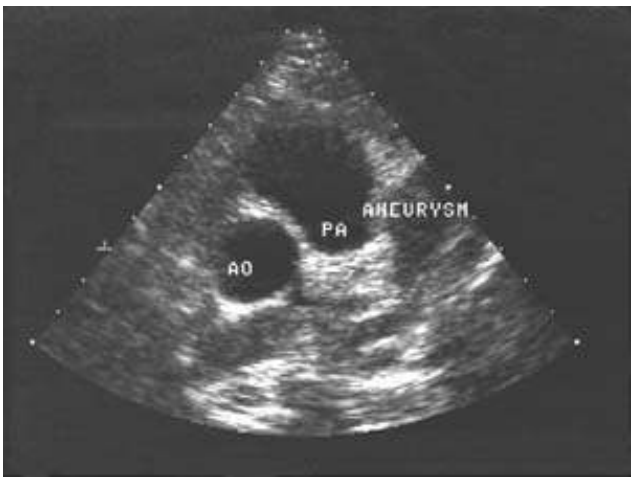

Fig. 3. Echocardiography showing a giant fusiform aneurysm of the left anterior descending artery measuring 20 mm in the case 4 patient. Abbreviations : AO, aorta; PA, pulmonary artery.

Discussion

KD is an acute, multisystemic vasculitis of unknown cause that is usually encountered in children younger than 5 years of age. No diagnostic test exists for KD, and thus, diagnosis is based on its clinical features. However, not all of the diagnostic features can be observed at initial presentation[2]. Moreover, due to the slow and variable evolution of the disease, the initial diagnosis are often incorrect[3]. Although the disorder is not familiar to the otolaryngologist, some frequently occurring head and neck symptoms including cervical lymphadenopathy and oropharyngeal injection may allow otolaryngologists to involve in patient management at early stage of the disease[4, 5]. The propensity of KD to mimic common infectious processes of the head and neck frequently delays correct diagnosis[4]. Common erroneous diagnoses are tonsillitis, upper respiratory infection,

otitis media, and cervical adenitis[4]. In particular, massively enlarged and tender cervical nodes may mimic a suppurative process[6]. Moreover, the clinical course of KD-associated cervical adenopathy may closely resemble the course of a retropharyngeal abscess[6]. Although cervical adenopathy is the least commonly encountered (50-75%) of the five diagnostic criteria of KD and is usually of minor significance[7], it occasionally (approximately 12%) becomes the most prominent and initially presented symptom, especially in older children[8, 9].

With regards to the diagnosis of a retropharyngeal abscess, CT may be a sensitive tool (sensitivity, 90%) for the detection of deep neck lesions[2]. CT scan provides the most accurate information on the extent and exact anatomical location of the lesion[10]. However, its specificity is too low (60%) to differentiate abscess from cellulitis[11]. Furthermore, there is no significant difference in Hounsfield unit between abscess and cellulitis in the retropharyngeal region[10]. In the presence of acute febrile illness, lymph nodes in the retropharyngeal space can become enlarged and appear as an enhancing capsule with a hypodense core on CT scan which could be interpreted as an abscess[12]. In addition, if obtained very early in the disease process, its accuracy can be even lower[12]. Moreover, radiologists may be overaggressive in the diagnosis of abscess[2]. Partially or thinly enhancing rims are presented in both early abscess and cellulitis, and occasionally in some abscess cases the rims are absent[10]. Thus, a hypoattenuating rim-enhancing retropharyngeal collection on CT scan is predictive, but not diagnostic of abscess[13]. Therefore, the decision to perform surgical procedure should be based on the clinical picture including antibiotic responsiveness rather than CT findings alone[11].

In this study, all four KD patients presented with chief complaints of fever and neck mass. The symptoms and signs of deep neck infection preceded and overrode other clinical features of KD, which gradually developed 3 to 4 days after hospitalization. All patients eventually fulfilled the diagnostic criteria. Mean duration from disease onset to diagnosis was 7.3 days (range, 6 to 9 days). Contrast CT scans of the neck showed a low density retropharyngeal lesion in all cases, which ranged in size from 5.5 mm to 9.8 mm. Very recently, Oyama et al.[14] compared the characteristic findings of retropharyngeal low density in KD patients with those observed in non-Kawasaki patients: it is characterized by 1) occurrence at an older age (5.3±1.8 yr vs 2.7±1.8 yr, P= 0.031), 2) no laterality (0% vs 50%), and 3) no ring enhancement (0% vs 67%). These findings are compatible with ours, i.e., 3 of our 4 patients aged 5 years and older, all hypodense lesions on CT were centrally located in the retropharyngeal area, and no lesion showed wall enhancement.

The coexistence of a retropharyngeal abscess and KD cannot be confirmed. It is, however, notable that no report has been issued on the coexistence of these two diseases[2]. The retropharyngeal space consists of adipose tissue and lymphatic vessels in a space surrounded by alar fascia and prevertebral fascia[11]. Other than retropharyngeal abscess, CT hypodensity in the retropharyngeal space can be detected in various pathogenesis associated with cervical lymph node swelling[12]. In KD, swelling of the cervical lymph node is one of the major clinical findings, whereas accumulation of lymph fluid in the retropharyngeal space may produce a low density lesion without wall enhancement[14]. Thus, an abscess-like low density lesion on CT may be a type of non-suppurative inflammation rather than an infection[2]. It may be caused by the reactive inflammation of lymphatic tissue which is considered to be the pathogenesis of cervical lymph node swelling in KD[14].

In conclusion, KD may be initially misdiagnosed as retropharyngeal abscess. Nonsuppurative cervical adenitis or suspected neck abscess resistant to intravenous antibiotics should signal the possibility of KD although the diagnostic criteria are not fulfilled.

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가와사끼병은 아직 원인이 밝혀지지 않은 급성 발열성 혈관염으로 진단을 위한 검사 소견이 없기 때문에 진단은 임상적 증상에 의존한다. 초기에 발열과 경부 림프절 종창 소견만을 보이고 가와사끼병의 다른 주 증상은 며칠 후에 서서히 나타나는 경우는 단순히 경부 감염성 질환으로 오인할 수 있다. 저자들은 질병 초기에 CT 검사로 후인두부 농양으로 진단받았으나 항생제 치료에 반응이 없었고 3-4일 후에 가와사끼병의 다른 증상이 뒤늦게 발현되어 정맥면역글로블린 투여 후에 해열과 증상의 호전을 보인 4명의 가와사끼병 환아를 보고하고자 한다. 발열과 림프절 종창의 주 증상으로 인해 경부 림프절염 또는 CT 검사 후 후인두부 농양으로 진단받은 환자들에서 항생제 치료의 효과가 없을 경우에는 가와사끼병의 가능성을 항상 염두에 두어야 한다.

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