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Editorial

Diagnostic Applications for Clinical and Imaging Data in Kawasaki Disease with Lymphadenopathy-First-Presentation

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▸ See the article "Imaging and Clinical Data Distinguish Lymphadenopathy-First-Presenting Kawasaki Disease from Bacterial Cervical Lymphadenitis" in volume 26 on page 238.

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Kawasaki disease (KD) is an acute vasculitis in childhood that can lead to coronary artery abnormalities (CAA).[1] The incidence of KD has continuously increased during the past decade since 2000 in South Korea, and is up to 194.7 per 100,000 children younger than 5 years.[2] In the absence of specific diagnostic tools, the diagnosis relies on principal clinical features and ruling out other clinically similar diseases. Among the principal diagnostic criteria for KD, cervical lymphadenopathy is the least common, occurring in approximately 24-75% of KD patients.[3][4] A small subset of KD patients present only with fever and cervical lymphadenopathy before other clinical features appear, which is described in this study as cervical-lymphadenopathy-first-presenting KD (LKD).[3-6] Although LKD patients showed only fever and cervical lymphadenopathy on admission, the typical KD features developed subsequently, after which a complete KD diagnosis was made. However, LKD patients have significant risks for misdiagnosis such as bacterial cervical lymphadenitis (BCL) or other lymphadenopathies during the acute phase of KD, these patients also experience delays in early diagnosis, lack of prompt appropriate treatment, and can develop CAA due to the delay in diagnosis and unfamiliar clinical features. A delayed diagnosis of KD is an apparent risk factor for CAA, but whether LKD patients have a greater risk of CAA or intravenous immunoglobulin unresponsiveness is still debated.[3][7]

It is difficult to differentiate LKD from BCL at admission, even for clinicians that are familiar with this condition. Currently, there are two approaches for study design to overcome this problem. The first approach compares LKD patients with typical KD, based on clinical and laboratory findings. The other identifies clinical, laboratory, and radiologic features that could distinguish LKD from BCL patients. Details of those reviews in the literature are well described in this article.[8] From a clinical perspective, this issue is important for clinicians, and provides useful information for treating patients with KD. However, to our knowledge, all of the articles on this issue have similar limitations. This is because the number of LKD patients was small and there is currently a lack of detailed supportive evidence that can confirm whether patients with only a lymphadenopathy presentation were KD patients, and the condition was not caused by other bacterial or viral lymphadenopathy. Therefore, a multicenter study with a larger number of patients is needed to elucidate the exact clinical characteristics of LKD patients and to identify a useful diagnostic tool for the acute phase of KD.

To the best of my knowledge, Kanegaye et al.[3] reported the most informative article that prospectively evaluated clinical, laboratory and imaging data that distinguish 57 LKD and 78 BCL patients over 7 years. Other studies reported less than 50 LKD patients.[4-6,9-11] One of strengths of this article is that the number of LKD patients (51 patients) is not so small compared with other similar studies. The authors previously reported a comparison of 24 LKD and 259 KD patients between January 2012 and December 2014.[12] Therefore, combining the two patient populations provides combined clinical and imaging data that are more valuable and substantial. In addition, the frequency (5.5%) of incomplete KD patients in this study is smaller than recently reported from South Korea. In the 8th nationwide survey of KD in South Korea, 32.8% were found to have incomplete KD.[2] Therefore, an explanation for the differences can provide better insight for this patient population.

Also, I would like to make some suggestions for additional research about this topic. This study was limited because the ultrasound findings are limited by the retrospective nature of the study and by intrinsic problems, such as the operator-dependent technique. However, additional studies could more specifically assess parameters like margin, echogenicity or morphologic characteristics of enlarged lymph nodes. In addition, the definition of LKD and BCL should be further elucidated. There are currently no confirmative methods to diagnose KD, and, in practice, it is more difficult to differentiate between LKD and BCL patients. Therefore, detailed supportive evidence should be included in order to rule out other diseases that present with similar symptoms. Imaging studies can be helpful for differentiating LKD from BCL. LKD can be associated with deep neck inflammation which can lead to retropharyngeal edema and non-suppurative phlegmon.[1,3,13] The authors report that the incidence of abscesses in the LKD group was 7%, based on ultrasonography. This is an unusual finding because the nature of cervical lymphadenopathy in KD is usually non-suppurative. Therefore, this finding will need to be explained in detail. It is surprising that there are no follow-up descriptions except initial echocardiographic data. The coronary artery should be described in the text at least during the acute and subacute stages of the illness. Finally, the authors conclude that "LKD tends to have more elevated systemic and hepatobiliary inflammatory markers and pyuria than BCL". Therefore, identifying and providing specific cut-off values for these parameters could be used in clinical practice to more readily identify and diagnose these patients.

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