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Kawasaki disease with retropharyngeal edema: case series from a single center experience

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To the Editor: Kawasaki disease (KD) and its classical diagnostic criteria are well known to pediatricians nowadays. However, KD can present with substantial overlap of clinical picture with many infectious diseases. Recently it is increasingly recognized that KD can present as pyogenic abscess-like formation with retropharyngeal edema (RPE). As estimated by Yoskivitch et al.,[1] 5% of KD presented with some forms of deep neck infection. They could be in various forms, such as peritonsillar abscess, parapharyngeal or retropharyngeal abscess-like edema or acute mastoiditis. Not only do they mimic retropharyngeal abscess clinically, some with incomplete presentation may also contribute to delayed diagnosis and hence timely treatment. In this case series, we reported our experience in managing 24 cases of KD presenting with RPE.

From January 1, 2007 to December 31, 2018, as one of the local tertiary referral centers for Paediatric Intensive Care, we encountered 24 cases of KD presenting with RPE. None of them were able to fulfill the diagnostic criteria for KD on presentation. They presented as fever with a variety of neck complaints clinically indistinguishable from cervical lymphadenitis or deep neck abscesses, including neck pain, tender neck mass, torticollis, and limited in range of neck motions. None of them had any signs of upper airway obstruction such as stridor. All of them were initially managed with empirical antibiotics but were all unresponsive to treatment.

As clinical course progressed, more KD signs evolved and eventually 12 of the 24 cases fulfilled the complete KD criteria at the time of initiation of intra-venous immunoglobulin (IVIG). The remaining 12 cases, however, remained to exhibit only fever plus presence of 2 to 3 KD criteria. In these cases, the diagnosis of KD was established by ancillary findings such as grossly elevated inflammatory markers, hypoalbuminemia, normochromic normocytic anemia, thrombocytosis after day 7 of fever, elevated alanine transaminase (ALT), or positive echocardiographic findings. Upon reviewing all the 24 cases, more than 50% had hypoaalbuminemia <35 g/L, sky-high C-reactive protein >1000 mmol/L, thrombocytosis >500,000/mL, and normochromic normocytic anemia. Five of them had positive echocardiogram findings of coronary abnormalities. These findings helped consolidate the diagnosis of KD and the decision for initiation of IVIG, coherent with the latest American Heart Association recommended approach for diagnosing incomplete KD.[2]

For all the 24 cases, ear-nose-throat surgeon was consulted for opinion. Imaging studies of the neck were arranged as suggested. Twenty-two had computed tomography (CT) scan of and two had ultrasonography of the neck. The choice of imaging modalities was selected based on availability as liaised with radiologists. The radiological findings were summarized in Supplementary Table 1, http://links.lww.com/CM9/A56. Most of them showed findings of RPE or fluids. The extent of retropharyngeal phlegmon was typically long-segment with no definite rim-enhancing collections. Generalized cervical lymphadenopathy was also evident, commonly involving level II (upper internal jugular deep cervical chain) and sparing level III-IV lymph nodes (middle and lower internal jugular deep cervical chains). These findings were consistent with the retrospective series reported by Nosaki et al and Kato et al.[3,4] Larger-scale prospective studies will be needed to establish the clear role and recommendation for the choice of ultrasonography and CT in differentiation of KD with RPE vs. infectious cervical lymphadenitis or genuine retropharyngeal abscesses.

One case eventually underwent fine-needle aspiration of the suspicious abscess, which only yielded 1.5 mL necrotic content, subsequent bacterial culture of the aspirated content was negative.[5] All 24 cases eventually responded satisfactorily to IVIG. This further reinforced the correct diagnosis of KD especially in those cases with incomplete
presentation, minimizing them from undergoing further invasive surgical interventions. One case suffered from the complication of coronary aneurysm and is currently still on warfarin, the remaining 23 cases recovered well with no long-term coronary sequelae.

In conclusion, KD with RPE can present with predominantly neck complaints and cervical lymphadenopathy mimicking deep neck infections. Incomplete presentation may further add to the diagnostic challenges and delay initiation of IVIG. Unresponsiveness to broad-spectrum empirical antibiotics, emergence of KD features, supportive ancillary laboratory findings (such as anemia, thrombocytosis, hypoalbuminaemia, elevated ALT) should raise suspicion of KD. Echocardiogram should be performed if expertise is available, as finding of coronary abnormalities is diagnostic. Involvement of RPE is commonly long-segment without definite rim-enhancement on CT scan. Early suspicion of KD and subsequent satisfactory response to IVIG may avoid the need for unnecessary invasive procedures such as fine needle aspiration or surgical explorations.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients’ parents/guardians have given their consent for the images and other clinical information to be reported in the journal. The patients’ parents/guardians understand that the names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflicts of interest
None.

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