Case report

Infantile cervical adenopathy and pyrexia with an unexpected sequel

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ABSTRACT

Introduction: Clinical presentation of cervical adenopathy and fever among infants albeit common remains a conundrum to many physicians notably, the otorhinolaryngologists. Kawasaki disease (KD) is an acute febrile systemic vasculitis which oftentimes is associated with cervical adenopathy.

Aim: We would like to highlight the diagnostic challenges faced along with management of KD and the importance of awareness amongst physicians in diagnosing this entity early.

Case study: Herein, we are reporting a case of a 10-month old who initially presented with a 3-day history of fever and unilateral cervical lymphadenopathy.

Results and discussion: This later turned out to be KD as the child progressively developed late onset of rashes, erythema of lips and tongue and edema of the hands and feet. Child successfully recovered with intravenous immunoglobulin and oral aspirin with no fatal complications.

Conclusions: Infants with fever and cervical adenopathy should not be taken frivolously as it may be a red herring of a more gruelling condition. As in our case, it turned out to be KD. Thus, high index of suspicion and collaboration between clinicians are prudent for early and accurate diagnosis and management as early treatment has been proven to improve the prognosis of patients.
1. INTRODUCTION

Kawasaki disease (KD), otherwise known as mucocutaneous lymph node syndrome, is an acute febrile systemic vasculitis of unknown etiology which manifests amongst children below five years of age. It usually affects medium-small sized vessels. KD is considered to be the leading cause of acquired cardiac malady amid developed countries as 20%–25% of cases are complicated with cardiac pathology. However, non-specific and sequential development of clinical features amongst KD patients may be misleading, causing delay in diagnosis. KD is common in children below 5 years of age and is rare amongst older children. In this case report, we highlight the importance of awareness of the sequential development of symptoms and management considering the devastating complications of this entity such as coronary artery aneurysm or thrombosis, myocarditis or pericarditis which may cause death.

2. AIM

We would like to highlight the diagnostic challenges faced along with management of KD and the importance of awareness amongst physicians in diagnosing this entity early.

3. CASE STUDY

A 10-month-old Malay infant presented to us with a one-day history of right neck swelling which was preceded by low-grade fever for the past 3 days. According to the infant’s mother, she noticed the right neck swelling to be increasing in size within a day and her child became lethargic with reduced oral intake. There was however, no other similar swelling elsewhere, no rashes, no skin changes, no vomiting, diarrhoea, fits or neck stiffness. There was also no accompanying nasal symptoms or aural symptoms, no cough or shortness of breath.

Informed consent was obtained from parents. Patient’s medical history revealed that he was just discharged home a week ago as he was admitted for pneumonia which resolved. His developmental milestone is according to his age. He had no allergies and his immunization is up-to-date. The infant lives at home with his parents with all the other members of his family well, none of whom with similar symptoms. The infant had no recent contact with any tuberculous or sick contacts or he did not travel outside the town recently.

Upon examination, patient was comfortable under room air, not tachypnoic or tachycardic with no audible stridor. Right-sided neck swelling was noted occupying level II, diffuse, soft, tender, not fixed to underlying skin, no punctum with inflamed overlying skin (Figure 1). The neck swelling caused mild torticollis. No other lymph node was palpable in the neck, axilla and inguinal with no evidence of hepatosplenomegaly. Intraoral examination, nasal and otoscopic were unremarkable. Review of his systems noted absence of rashes, gastrointestinal symptoms, weight loss, anorexia or other constitutional symptoms. Initial documented temperature was 37°C. Complete blood count revealed a white blood count of 14 × 10^9 cells/L and haemoglobin count was 10.0 g/dL. C-reactive protein done was raised with 20 mg/L and erythrocyte sedimentation rate was 35 mm/h. The remainder of his complete blood count, electrolytes and liver enzymes were within normal limits.

The infant was admitted for further investigation and was started on intravenous augmentin (15 g/kg b.m.) along with syrup paracetamol which was given a qid dosing. Subsequent day, the infant was noted to have spiking temperature ranging 37.5°C–38.5°C with reduced oral intake and few episodes of diarrhoea. The infant was started on intravenous fluid at once and was monitored stringently although his vital signs were stable. The 2nd day of admission revealed neck swelling over the left side of neck with rashes over trunk, swollen hands and feet with one episode of spiking temperature. The infant was referred to the paediatrics team for consultation and an ultrasonography of the neck was done which revealed

![Figure 1. Diffuse right neck swelling over the level II, III.](image)

![Figure 2. Right cervical lymphadenopathy.](image)
bilateral cervical lymphadenopathy (Figure 2). Upon the 3rd day of admission, there was signs of injected lips, tongue with bilateral non-purulent conjunctivitis. In the light of patient's clinical symptoms, a diagnosis of KD was made. Parents were informed regarding the diagnosis as well as the management and intravenous immunoglobulin was given followed by high-dose of oral aspirin. Echocardiography was performed to support diagnosis of KD following the intravenous immunoglobulin which exhibited mild trivial regurgitation with no signs of failure, coronary artery aneurysm or thrombosis.

Patient subsequently recovered well and was discharged home with low-dose aspirin and a 1-week appointment upon which child exhibited complete recovery.

4. RESULTS AND DISCUSSION

Doktor Tomisaku Kawasaki was the first who described KD in a 4-year-old Japanese boy in 1967, which subsequently became prevalent amidst Japanese descendants in Hawaii. Although highest incidence of this disease are seen among Asians, many authors have reported that KD may occur in any ethnicity. Male to female ratio have been shown to be 1.5 : 1.0 with peak age of incidence between 18–24 months although cases among adults have been reported. It is the second most common vasculitis amongst children following Henoch Schonlein. Our patient mentioned above is a 10-month-old Malay boy.

Although the etiologic factor of KD remains unknown; microbial infection, immune response or genetic susceptibility has been postulated to cause KD. In our case, unresolved infection could be the culprit as the child had recent hospital admission for pneumonia.

Diagnosis is made based on clinical presentation as there is no specific diagnostic test or pathognomic clinical features which oftentimes causes difficulty and delay in diagnosis. Having said that, diagnostic guideline have been made to assist physicians and has been regarded as the cornerstone of diagnosis. Clinical criteria which has been established based on the guideline includes unexplained fever for 5 days or longer and at least 4 of the following: (1) non-exudative conjunctival injection; (2) oral involvement, including strawberry tongue, mucosal hyperemia, and cracked or erythematous lips; (3) changes in peripheral extremities, including edema or desquamation; (4) polymorphous rash; and (5) acute cervical lymphadenopathy greater than 1.5 cm, which oftentimes may not present simultaneously and resolve without any treatment which makes the diagnosis more challenging.

Diagnosing KD causes conundrum all the criteria are not present simultaneously. As for cases with prolonged fever of more than 5 days with any of 2 to 3 clinical criteria's mentioned above, they are categorised as 'Incomplete KD'; which mostly is confirmed by an echocardiography. At times, patients may turn up to the hospital with a cardiac sequelae years after having KD without being able to recall such illness during childhood. Our patient discussed above exhibited fever along with all five criteria which is highly suggestive of KD.

Apart from clinical criteria, a few laboratory investigations which can support the diagnosis includes elevated Erythrocyte sedimentation rate, C-reactive protein and platelet count accompanied by negative rheumatoid factor and antinuclear antibodies. Echocardiography is another important investigation for patients presenting with prolonged fever as to rule out KD or atypical KD.

Differential diagnosis of infants presenting with pyrexia and cervical adenopathy includes: (1) infection which may be bacterial, viral, mycobacterial or granulomatous and fungal; (2) rheumatological; (3) neoplastic; (4) immunological; (5) vasculitic disease (6) storage disorder. Oftentimes, infants presenting with fever and lymphadenopathy requires a wide range of workup to obtain correct diagnosis. This however, may cause frustration amongst parents who are usually anxious to know the diagnosis and management of their little ones. Presence of cervical lymphadenopathy in children below 5 years of age are almost always linked with viral illness as compared to adolescent whereby lymphoma should be ruled out. Lymphadenopathy that persists beyond six to twelve weeks warrants a fine-needle aspiration for cytology or biopsy.

Management of KD is focused on reducing the inflammation around coronary arterial walls as well as myocardium especially during the acute phase of disease. Intravenous immunoglobulin (IVIG) is the first line of treatment, commonly given in a single dose of 2 g/kg b.m. given over 10–12 hours which is aimed to prevent or reduce aneurysm formation as well as symptomatic relief for acute illness and has been proven beneficial especially if administered in the early phase. High-dose of oral aspirin is then given for its anti-inflammatory effects (80–100 mg/kg b.m. a day in four divided dose) for 14 days or until patient becomes afebrile for 48–72 hours followed by low-dose aspirin for its antiplatelet effect (3–5 mg/kg b.m. a day) up to 8 weeks. However, untreated cases has been reported to resolve spontaneously within weeks and it’s often regarded as ‘self-limiting.’

As for the follow-up of these patients, one the baseline echocardiography is normal, a repeat echocardiography should be done after 1-month of treatment and subsequently after 1 year is essential. It is prudent to take note that recurrence has been reported amongst KD patients and mostly within 2 years from the initial diagnosis. Hence, this warrants awareness among clinicians as with prompt treatment, good prognosis have been advocated regarding this condition.

5. CONCLUSIONS

Infants presenting with fever and cervical adenopathy should not be taken lightly by attending physicians as it may be an early sign of a more gruelling condition. As in our patient, it turned out to be KD. Albeit having all five criteria of KD, sequential progress of symptoms delayed the diagnosis. Thus, high index of suspicion and collaboration
between clinicians are prudent for early and accurate diagnosis and management as early treatment has been proven to improve the prognosis of patients.

Conflict of interest
There is no conflict of interest.

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