CASE REPORT

Acute peritonsillar swelling: a unique presentation for Kawasaki disease in adolescence

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Accepted 5 June 2018

SUMMARY

We report the case of a 14-year-old man with unilateral peritonsillar swelling, airway compromise and fever. On physical examination, the patient was pyrexia with trismus, dysphonia, uvula deviation, exudative unilateral peritonsillar swelling and unilateral cervical lymphadenopathy. Attempts at aspirating the prominent peritonsillar region were unsuccessful. CT head and neck identified a large inflammatory mass arising from the left palatine tonsil. The patient was treated for presumptive peritonsillar abscess. After failing to respond to intravenous antimicrobials, he progressively developed the classical sequelae of Kawasaki's Disease. Echocardiogram identified coronary arteritis. Intravenous immunoglobulin and high-dose aspirin were initiated and his clinical picture improved. Kawasaki's disease mimicking an acute infective process can pose a diagnostic dilemma. It is an uncommon differential in the adolescent population. Prompt recognition and initiation of appropriate therapy are imperative to minimise morbidity.

BACKGROUND

Kawasaki's disease (KD) is a medium vessel vasculitis, typically diagnosed under the age of 5.¹ It is the leading cause of acquired heart disease in the paediatric cohort, but is rarely seen in the adolescent or adult population.² Peritonsillar, retropharyngeal and parapharyngeal swelling are uncommonly reported manifestations of KD. Our case presents a rare clinical manifestation of a disease process not typically seen in the adolescent demographic. This is an important demonstration of the diverse clinical features of KD and its capacity to mimic acute infective pathologies. It is a critical reminder to consider KD as a differential early, even when confronted with atypical presentations in atypical populations. This case is unique, in that there are no documented cases of KD mimicking peritonsillar abscess in the adolescent population.

CASE PRESENTATION

We present the case of a healthy 14-year-old Caucasian man. He attended a tertiary paediatric emergency department with a 72 hours history of progressive odynophagia, drooling, neck swelling and dysphonia. He had been reviewed by a local medical officer and provided with a presumptive diagnosis of tonsillitis and commenced a course of antibiotics.

On admission he was pyrexia, tachycardia, dehydrated and drooling with limited mouth opening. He had a prominent cellulitic, exudative left peritonsillar swelling. His uvula was deviated to the right. He had left level II cervical lymphadenopathy and torticollis. He was intolerant of lying supine with oxygen desaturations. Flexible nasendoscopy showed a bulge in the lateral oropharyngeal wall. Attempts at aspirating the prominent peritonsillar swelling were unsuccessful. The patient was admitted to the paediatric high dependency unit for airway observation. He was commenced on intravenous dexamethasone 8 mg 8 hourly and intravenous piperacillin/tazobactam 4.5 g 6 hourly. He was consented for surgical intervention if his clinical picture did not improve.

After 72 hours, the patient had failed to progress with persistent fevers, lymphadenopathy and peritonsillar swelling. He progressively developed a new sequelae of clinical signs. Strawberry tongue, cracked lips, injected conjunctiva, polymorphous rash, oedematous hands and desquamation of the skin of his feet led to a revised diagnosis of KD.

INVESTIGATIONS

His biochemistry on admission showed leucocytosis (18.50) with neutrophilia (15.36). He had moderately elevated inflammatory markers (ESR 22, CRP 62). Peritonsillar exudate swab culture showed no growth. EBV and CMV serology were negative.

CT head and neck with contrast showed a 31×24 mm soft tissue mass arising from the left palatine tonsil fossa with an area of central low attenuation. There was surrounding mass effect with deviation of the upper airway. There was a large 3×3 cm lymph node anterior to the sternocleidomastoid muscle. Imaging was in keeping with a large inflammatory phlegmon arising from the left peritonsillar space. A repeat CT Neck after 72 hours reported persistent asymmetrical swelling of the left peritonsillar region (figure 1). The central area of low attenuation remained ill defined and was thought to reflect oedematous tissue rather than a defined abscess. Overall, the swelling was more prominent than previous imaging.

Repeat bloods taken when KD had become clinically obvious showed persistent leucocytosis (20.30) and neutrophilia (18.50). C reactive protein (CRP) had risen to 231 and erythrocyte sedimentation rate (ESR) to 59. Forty-eight hours after administration of intravenous immunoglobulin, his leucocytosis was subsiding (12.5) and absolute neutrophil count had returned to the

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To cite: Connell JT, Park JH. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2018-224441

Figure 1 Axial CT slice highlighting the left peritonsillar swelling (arrow).

normal range (9.25). ESR remained elevated at 104 while CRP was down trending (152).

The patient underwent serial echocardiograms once the diagnosis of KD had been made. In paediatric cardiology, coronary artery diameter range is adjusted for age, height, weight and body surface area. Deviations from normal range are measured using a Z score. Our patient's age appropriate range for right coronary artery (RCA) is 2.53–4.79 mm. His initial study

showed a prominent and irregular left main coronary artery within normal limitations for vessel calibre. The RCA was diffusely dilated and irregular, up to 5.4 mm with a Z Score of 3.01 (figure 2). Repeat echocardiogram after 1 week demonstrated a persistent diffusely dilated RCA up to 5 mm ($Z=2.32$). His third study performed 3 weeks later showed improvement in the prominence of his RCA measuring 4.2–4.6 mm. One vessel segment remained prominently dilated at 4.9 mm ($Z=2.15$). Another echocardiogram was performed after a further 4 weeks, demonstrating resolution of the diffuse RCA dilation towards normal. However, there remained a prominent region up to 5.1 mm suspicious for a saccular aneurysm. A coronary angiogram was ultimately performed 3 months following his initial presentation. No persistent aneurismal lesions were identified (figure 2).

DIFFERENTIAL DIAGNOSIS

The clinical presentation was initially most consistent with a peritonsillar or retropharyngeal abscess. CT localised the swelling to the peritonsillar tissue. After failing to respond to intravenous antibiotics, atypical infective organisms were also considered. Once the additional clinical signs of KD emerged, the diagnosis clearly shifted to KD.

TREATMENT

Once KD was suspected, the patient was administered 160 g (2 g/kg) of intravenous immunoglobulins (IG.) The dose was repeated 48 hours later. He was commenced on oral antiplatelet therapy. He was discharged after his fever, rash and oropharyngeal swelling had subsided. His discharge therapy was 5 mg/kg/day of aspirin for 4–6 weeks. He was planned for continued cardiac surveillance and internal medicine/immunology follow-up.

Figure 2 Echocardiogram highlighting a 5.4 mm right coronary artery dilated segment.

OUTCOME AND FOLLOW-UP

Two days after being discharged, the patient represented with recurrence of classical KD symptoms. He was administered intravenous methylprednisolone (30 mg/kg/day) for 3 days. Symptoms subsided and he was discharged on a weaning course of oral prednisolone. One week later, he represented with melaena. He required two transfusions of packed red blood cells. Upper gastrointestinal (GI) endoscopy diagnosed two duodenal ulcers likely related to prednisolone and aspirin despite taking a regular proton pump inhibitor. He ultimately recovered with KD-related symptom resolution and return to normal activities. The coronary aneurysm diagnosis was downgraded on coronary angiography.

DISCUSSION

KD is a medium-vessel vasculitis characterised by systemic and mucocutaneous inflammation. The diagnosis of classical KD requires persistent fever for ≥ 5 days in addition to four out of five typical clinical features. Typical features criteria include polymorphous exanthema, cervical lymphadenopathy, conjunctival injection, skin changes to extremities and mucosal changes to oral cavity and lips. Atypical KD requires < 4 of these symptoms if coronary vessel changes are observed.¹

KD is most commonly seen under the age of 5. It is rare in adolescents with only 2%–6% of cases presenting over the age of 9. Cases diagnosed in adolescent years are associated with greater diagnosis delays and higher incidence of cardiac complications.² This is, at least in part, secondary to KD being considered as a less probable differential diagnosis, leading to delayed recognition and treatment initiation.³ KD is the leading cause of acquired cardiac disease in the paediatric population, underpinning the importance of prompt diagnosis to minimise cardiac complications.⁴

KD has a tendency to mimic a diverse range of differential diagnoses on presentation including viral exanthems, streptococcal infections and acute lymphadenitis.¹ Tonsillar, peritonsillar and parapharyngeal manifestations of KD have been reported sparingly and typically in a younger demographic. Ravi and Brooks reported the case of a 7-year-old child with a clinically suspected peritonsillar abscess. Symptoms persisted despite unilateral tonsillectomy prompting the revised diagnosis of KD.⁵ Cai *et al* reported a 3-year-old child with parapharyngeal swelling secondary to KD, unresponsive to antibiotics and surgical drainage.⁶ Yap *et al* presented a 10-year-old child with exudative bilateral tonsillitis who subsequently developed coronary complications.⁷ Typical to all of these cases was that diagnosis was delayed due to the initial absence of archetypal features of KD.

Cases of retropharyngeal space manifestations have been more widely documented.^{8–13} Hung *et al* reported KD with retropharyngeal swelling incorrectly reported as abscess on CT.⁸ A literature review by Hung *et al* of five similar case reports concluded that CT may over diagnosis inflammatory nodes as abscess. Ganesh *et al* and Fang *et al* reported cases of retropharyngeal swelling secondary to KD that led to airway compromise requiring intubation.^{9–10} Wood *et al* reported a case of atlantoaxial subluxation secondary to KD-related retropharyngeal oedema.¹³ All of these cases demonstrate the severity of symptoms that deep neck space manifestations of KD can herald.

Our case is a critical learning tool due to its unique demographics. Newly diagnosed KD in this age demographic is rare, particularly in the Caucasian population where incidence is 3.6–3.7 per 100 000.¹⁴ The incidence in Caucasian populations

has been estimated to be up to 10-fold less common than in Asian populations.¹⁵

This case also highlights the capacity for KD to mimic acute infective differential diagnoses. Our case was hallmarked by a peritonsillar focus of swelling, which is a scarcely documented anatomical site for manifestations of KD. The coronary aneurysm identified on the echocardiogram highlights the potential sinister complications of KD and reiterates how fundamental prompt diagnosis is to minimise cardiac morbidity. Finally, this case is a timely reminder of the potential complications of medical therapy, demonstrated by the unfortunate development of duodenal ulcers, anaemia and transfusion requirement secondary to high-dose antiplatelet and steroid therapy. KD can present as a diagnostic dilemma with a spectrum of clinical manifestations. Its diverse presentations can compromise diagnosis and initiation of appropriate management. Prompt diagnosis can minimise cardiac morbidity.

Learning points

- Kawasaki's disease (KD) can present with a range of clinical manifestations.
- KD should be considered early, particularly when suspected infective foci are not responsive to traditional therapy.
- KD can occur in adult or adolescents. Older demographics may incur worse clinical outcomes secondary to diagnostic delays.
- Delayed diagnosis of KD can contribute to cardiac morbidity.

Contributors The patient and their family were integral contributors through their consent, cooperation and ongoing provision of collateral history.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent Parental/guardian consent obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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