A 13-Year-Old Boy Who Has Kawasaki Disease Shock Syndrome Presents with Parotitis

Yilmaz Yozgat[1] Selcuk Uzuner[2] Aysegul Dogan Demir[2] Mustafa Ogur[2] Can Yilmaz Yozgat[3] Ozden Turel[4]

[1] Department of Pediatric Cardiology, Bezmialem Vakif University, Istanbul, Turkey
[2] Department of Pediatric Critical Care Medicine, Bezmialem Vakif University, Istanbul, Turkey
[3] Faculty of Medicine, Bezmialem Vakif University, Istanbul, Turkey
[4] Department of Pediatric Infectious Diseases, Bezmialem Vakif University, Istanbul, Turkey

Address for correspondence Yilmaz Yozgat, MBBS, MD, Division of Pediatric Cardiology, Department of Pediatrics, Bezmialem Vakif University, Adnan Menderes Bulvarı, Vatan Caddesi, 34093 Istanbul, Turkey (e-mail: yozgatyilmaz@yahoo.com).

J Pediatr Intensive Care 2020;9:60–63.

Abstract

We report a 13-year-old boy who (initially) had symptoms of toxic shock-like syndrome and mumps. Then, the patient was hospitalized in the pediatric intensive care unit (PICU) because of his ongoing hemodynamic instability (low blood pressure of 70/30 mm Hg and capillary refill time of >4 seconds). During his stay in the PICU, the patient was treated with fluid resuscitation and vasoactive infusion and at the same time was diagnosed with Kawasaki disease shock syndrome (KDSS), when giant right coronary artery aneurysms were detected on echocardiographic examination. This case illustrates the risk of KDSS in patient who carries both parotitis and toxic shock-like syndrome. The clinicians should be cautious about detecting any types of coronary artery aneurysms in such patients. This is the first case of KDSS associated with parotitis reported in the literature.

Keywords
- Kawasaki disease shock syndrome
- toxic shock syndrome
- parotitis

Introduction

Kawasaki disease (KD) is an acute self-limited systemic pediatric vasculitis that is characterized with high-grade fever for >4 days, bilateral nonpurulent conjunctivitis, changes in the lips or oral cavity, changes in the extremities, rash, and cervical lymphadenopathy. Recently, reaction at the site of administration of bacillus Calmette–Guerin (BCG) vaccine is a precious clinical finding in suspicious cases, and it might be as prevalent as cervical lymphadenopathy in KD.[1] Coronary artery aneurysms or ectasia develop in 15 to 25% all of the untreated children.[2,3] Toxic shock syndrome (TSS) is an exotoxin-mediated acute life-threatening illness, which is usually caused by superantigens which are being produced by either *Staphylococcus aureus* or group A *Streptococcus*. TSS is characterized by high fever, rash, hypotension, multiorgan failure (involving at least three or more organ systems), and desquamation, typically of palms and soles. KD and TSS have some overlapping clinical features, but coronary artery lesions are not evident in TSS, and the shock is not a feature of KD.[3,4] KD shock syndrome (KDSS) has recently been reported to occur secondary to KD. KDSS is associated with more severe laboratory markers of inflammation and a higher risk of coronary artery abnormalities, mitral regurgitation, prolonged myocardial dysfunction, and resistance to immunoglobulin therapy.[5] We report a patient who presented with features of mumps and TSS. Later, the patient was diagnosed with KDSS when coronary artery aneurysms, mitral regurgitation, and prolonged myocardial dysfunction were detected (subsequently) on an echocardiographic examination.

Case Report

Before the admission of the 13-year-old boy to our clinic, he was treated initially by a practitioner in family medicine for

received
May 13, 2019
accepted after revision
August 30, 2019
published online
October 9, 2019

DOI https://doi.org/10.1055/s-0039-1697978.
ISSN 2146-4618.

2 days. The practitioner diagnosed the patient as a case of tonsillopharyngitis and parotitis and prescribed cefuroxime therapy. During the first 2 days of treatment with cefuroxime, the patient continued to have a high-grade fever. A day after the treatment completion, the patient started to develop an itchy rash on his palms and his fingers, which extended throughout his body. Since there was no response to the treatment given so far, the patient was referred to our emergency department. During his admission to the emergency department, the patient was feeling nauseated and was febrile (40.3°C), tachycardia (120/min), mildly hypotensive (90/48 mm Hg), and poorly perfused (capillary refill of 3 seconds). His physical examination revealed a semiconscious state, with thick neck, tenderness in abdomen, swelling (2 × 2 cm) in the parotid gland (►**Fig. 1**), bilateral conjunctival hemorrhage (►**Fig. 2**), rash on his lower extremities and back (►**Fig. 3**), dorsal swelling of his right hand (►**Fig. 4**), and a strawberry-like tongue with cracked lips (►**Fig. 5**). BCG vaccine injection site reaction was not detected. Clinical characteristics criteria for complete KD include five different specified vasculitis findings. However, the present case only fulfilled three out of five specified vasculitis findings. These findings were changes in the lips or oral cavity, changes in the extremities, and skin rash. These findings could be confused with other types of vasculitis syndromes, so our team did not consider the patient to have KD initially. Laboratory investigations showed hemoglobin of

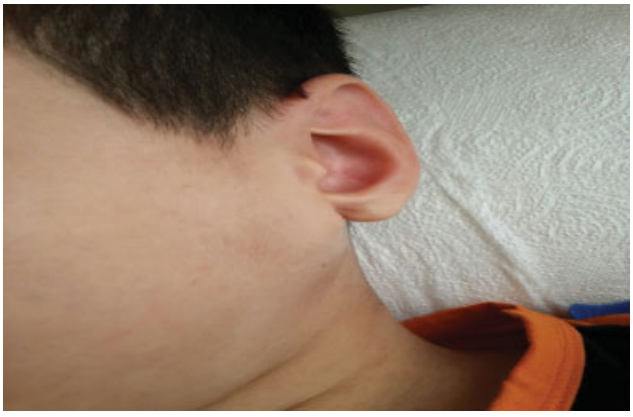

Fig. 1 Parotid swelling on the left side.

Fig. 2 Bilateral subconjunctival hemorrhage.

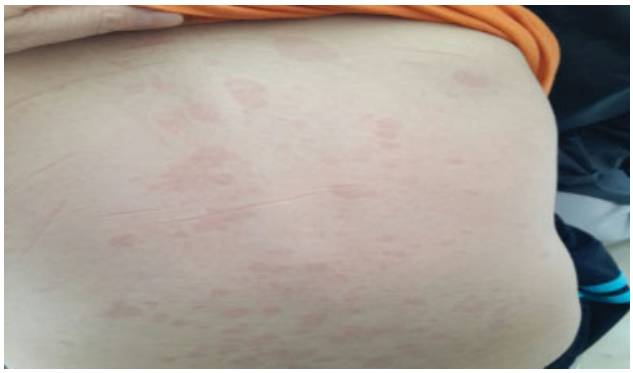

Fig. 3 Rash seen on the patient's back.

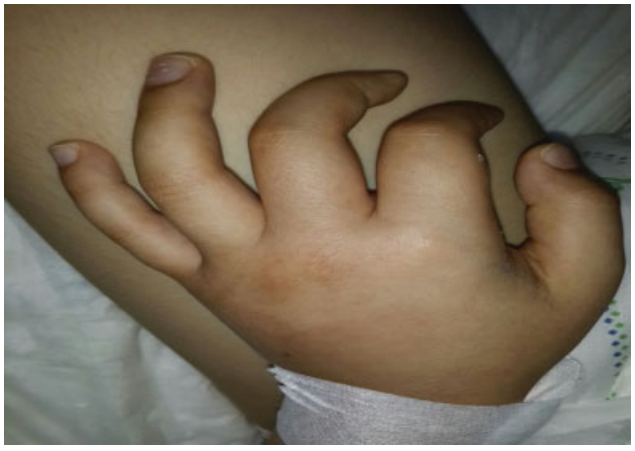

Fig. 4 Bilateral swelling of the patient's hand.

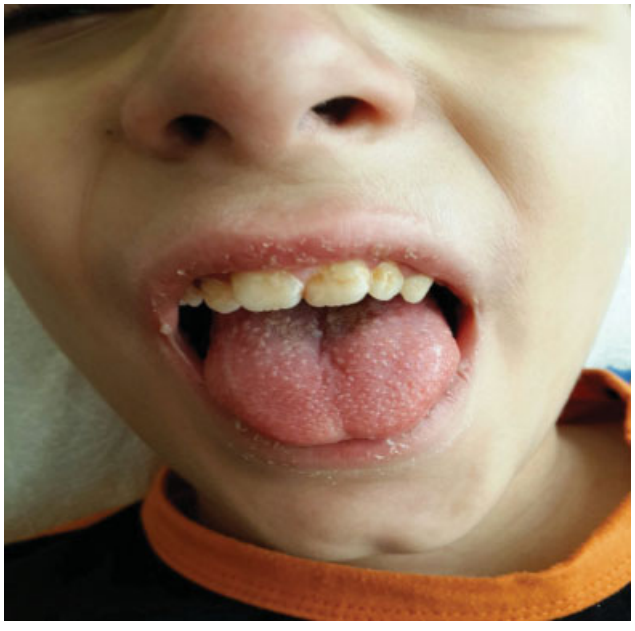

Fig. 5 A strawberry-like tongue with cracked lips.

10 g/dL, white blood cells of 11 × 103/μL, thrombocytopenia (109 × 103/μL), renal impairment (blood urea 105 mg/dL, serum creatinine 2.3 mg/dL), high C-reactive protein of 345 mg/L, and high erythrocyte sedimentation rate of 65 mm/h. The first sets of cardiac enzymes were troponin-I 3.5 IU/mL, creatine kinase (CK) 450 IU/mL, CK-MB isoenzyme 56 IU/L, amylase 185 g/dL, and albumin 2.8 g/dL. Mumps immunoglobulin M antibody was detected in the patient's blood analysis; hence, the patient was considered to have mumps. Rest of the laboratory investigations was regular. The computed tomography (CT) scan of the brain and the abdominal ultrasound examination were normal. A lumbar puncture test was performed to rule out the possibility of meningitis; the cerebrospinal fluid was normal. Microbial cultures were negative. Specifically, nasal swabs for *S. aureus* and *Streptococcus* were sent and were negative as well. As the patient was still sick and toxic, he was empirically treated with intravenous ceftriaxone (100 mg/kg) and vancomycin (40 mg/kg).

The patient was admitted to the pediatric intensive care unit (PICU) because of his ongoing hemodynamic instability (moderate hypotension with 70/30 mm Hg and poor perfusion with capillary bed capillary refill > 4 seconds). The initial management included administration of fluid resuscitation (60 mL/kg total), broad-spectrum intravenous antibiotics, and inotropic support. His hypotension persisted despite fluid resuscitation and vasoactive infusions (dopamine, dobutamine, and epinephrine). A cardiac echocardiographic examination performed on the third day in PICU revealed mild myocardial dysfunction (fractional shortening of 25%) and minimal mitral valve regurgitation, without coronary artery anomalies. On the sixth day of admission, the patient was still febrile with a temperature of 39.3°C. On the seventh day, another echocardiographic examination performed showed moderate mitral regurgitation, prolonged overt myocardial dysfunction (fractional shortening of 20%), and diffuse dilatation of right coronary artery (RCA) with the maximum diameter of the RCA being 8 mm. A CT angiography was performed to evaluate the coronary arteries. The CT coronary angiogram revealed a giant aneurysm of the RCA. The results of angiogram met the diagnostic criteria for KDSS. The patient was immediately started on intravenous immunoglobulin (IVIG) therapy (2 g/kg) and aspirin (75 mg/kg/d). An additional dose of IVIG was given on the 10th day for incessant fever. After the administration of the second dose of IVIG infusion, the fever resolved, and the rash disappeared 18 hours later. Serial laboratory results also showed signs of resolution of the inflammatory markers and gradual increase of his platelet count (810×10^3). An echocardiographic examination (on the 15th day) showed normal systolic function and giant aneurysm of the RCA. The patient was discharged on the 20th day on treatment with low-dose aspirin (5 mg/kg/d) and warfarin. During the second month of his illness, we performed a selective coronary angiogram, which revealed that there was no regression of the coronary artery dilatation (►**Video 1**). The large RCA had still not regressed in size; instead, it had stayed at the width of 10 mm.

Video 1

Right coronary angiography revealing severe diffuse aneurysmal coronary artery. Online content including video sequences viewable at: https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0039-1697978.

Discussion

KD is an acute self-limited systemic pediatric vasculitis, which is characterized by high-grade fever for more than 4 days, bilateral nonpurulent conjunctivitis, changes to the lips or oral cavity, changes in the extremities, rash, and cervical lymphadenopathy. Coronary artery aneurysms or ectasia usually develop in 15 to 25% of all untreated children.[1] The recent description of KDSS shows a positive association between more severe laboratory markers of inflammation and a higher risk of coronary artery abnormalities, mitral regurgitation, inadequate response to IVIG, and prolonged myocardial dysfunction.[5,6]

KDSS is distinguished from KD by most overt systolic and diastolic dysfunction and shock, thrombocytopenia, more severe laboratory markers of inflammation, and having an inadequate response to IVIG and a higher rate of coronary artery dilation and aneurysm formation. Although the overt systolic dysfunction resolved with vasoactive infusions in our patient, the abnormal ventricular diastolic function persisted into the chronic phase of the illness.[5–7] TSS is an exotoxin-mediated acute life-threatening illness which is usually caused by superantigens which are being produced by either *S. aureus* or group A *Streptococcus*. TSS is characterized by high-grade fever, rash, hypotension, multiorgan failure (involving at least three or more organ systems), and desquamation, typically of palms and soles. This syndrome includes severe myalgia, vomiting, diarrhea, headache, and neurologic abnormalities.[3,4] Hypotension and systolic dysfunction are common features of TSS, but coronary artery dilatation has not been reported in any cases in the literature as yet.

There are several reports in the literature that present similar pediatric patients who have had a diagnosis like our case. All the examples which are given below also had fulfilled case criteria for both TSS and KD, and they were ultimately diagnosis with KD.[8–10] Davies et al described a case of an adolescent male who was initially treated for TSS, and later, an echocardiographic examination revealed coronary artery aneurysm.[11] Another case was a 7-month-old infant who had the same symptoms and findings of TSS; however, his autopsy report suggested that the infant had coronary artery vasculitis.[8] Our case presented with similar clinical features of TSS, but our case does not satisfy the criteria for TSS. In the literature, there have been no reports about the correlation between the mumps virus and TSS. Our case had the clinical features of KDSS, which is usually associated with more severe laboratory markers of

inflammation and an earlier onset of more severe coronary artery dilatation, mitral regurgitation, and prolonged myocardial dysfunction.

We have reviewed 15 different cases of KD, which are also presented with parotitis in the literature.[9] In all of these 15 cases, none developed the shock syndrome. The difference from our case is that our case was diagnosed with KDSS instead of KD. There has not been any other case in the literature that had the presence of KDSS with parotitis. Due to the characteristics of KD, a potential infectious cause might trigger the disease.[10] Even though it is still unknown as to how infectious agents play a role in causing vasculitis syndrome, it is known that infectious agents play an integral role to cascade inflammation in susceptible children with KD.[12] In our study, the mumps virus triggered the cascade of inflammation. Maybe because of his gender and his older age, the case was diagnosed with KDSS instead of KD.

Conclusion

In this case, we distinguished KDSS from KD by evaluating most overt systolic and diastolic dysfunction and shock, thrombocytopenia, more severe laboratory markers of inflammation, inadequate response to IVIG, and a higher rate of coronary artery dilation and aneurysm formation. KDSS should be considered in all patients who have parotitis and toxic shock-like syndrome; clinicians should be cautious about detecting coronary artery aneurysms in such patients.

Ethical Approval

This case report does not contain any studies with human participants or animals performed by any of the authors. Institutional Review Board approval was not required for this case report.

Authors' Contributions

All authors participated in creating the content of the case report, editing, and providing final approval for submission. No undisclosed authors contributed to the case report.

Funding

This research received no specific grant from any funding agency or commercial or not-for-profit sectors.

Conflict of Interest

None declared.

Acknowledgments

The authors would like to thank the parents and patient for granting permission to publish the case report.

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