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(CASE REPORT)

Atypical Kawasaki Disease in a 6-year-old Albanian child, Case report

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Abstract

Background: Kawasaki Disease is a vasculitis of medium size arteries of children less than 5 years. If untreated it results in dilatations of coronary arteries and may lead to aneurisms, heart failure, myocardial infarction and death.

Case summary: A 6-year-old child had prolonged fever of 7 days, mucosal involvement of throat and mouth and edema of limbs. After evaluation, the diagnosis of KD was made, and the child was treated with aspirin and corticosteroids.

Discussion: An atypical case of KD was made with only fever and two out of five criteria met. Follow-up revealed no complications.

Keywords: Kawasaki Disease; Coronary arterial aneurism; Echocardiography; Coronary artery aneurysm; Intravenous immunoglobulin; Vasculitis; Mucocutaneous lymph node syndrome

1. Introduction

Kawasaki disease (KD) is an acute medium-sized arteritis most commonly in ages 6 months to 5 years with prolonged fever of more than 5 days; as well as a) polymorphous exanthema of the trunk; b) changes of extremities and exfoliation of the fingertips, c) bilateral non-purulent conjunctivitis, d) non-purulent cervical adenopathy of more than 1.5 cm, and e) redness and swelling of mouth, lips, throat and tongue [1,3,4,10,12]. Reports suggest that complete KD criteria are only met in 50-64% of the cases [9].

The median age of children with KD is below 5 years with a median age of 3 years and a peak incidence in infants less than a year old, and mostly in boys [2,10,16]. KD is the leading cause of acquired heart disease in children in developed countries [3,12]. KD complications may develop in 25-30% of untreated children [5].

Treatment includes aspirin, steroids, and intravenous immunoglobulin [1,10,16]. If left untreated, the symptoms resolve in about 10 days [2]. In case of delayed treatment, missed diagnosis, or in treatment refractory cases aneurisms can result and cause severe sequelae [14].

The challenge is a timely diagnosis, and how to prevent cardiovascular complications. Unresponsiveness to intravenous IG is a major risk factor for the development of coronary artery lesions [5], as is the delayed administration of the IVIG 7-10 days after the onset of the disease [6]. The diagnosis of KD still relies on clinical criteria [12,16] and is one of exclusion [10].

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Special care should be given to identification of any implications to coronary arteries that come in the form of stenosis or dilatations [1], coronary artery fistula [14] and aneurism [16], myocardial infarction, arrythmias [2,5], myocarditis, pericarditis, congestive heart failure and death [10].

Less reported signs include redness around the BCG vaccination site, restlessness and painful joints that restrict movement [9]. Lab tests are marked with elevated ESR and CRP [9]. Follow-up of coronary arteries is recommended in 2 and 6 weeks, and if complications persist again up to 2 years.

2. Case summary/presentation

The child gets admitted to a pediatric clinic for continued fever and abdominal pain lasting for one week. Previously the child was seen by a PHC pediatrician and prescribed antibiotics and steroids. Lab and imaging tests were done at PHC level, and the lab results indicated urinary infection. The child was referred to the pediatric clinic as the situation was not improved, and the complaints of fever, abdominal pain and difficulties in walking persisted. Once presented to the pediatric clinic the child was admitted to the Cardiology unit for further investigations.

The female child was born in 2017 and is the second-borne of 4 children, delivered normally, with weight at birth at 3,350 gr, and vaccinated for BCG and Hepatitis B. The vaccination is completed as per schedule and Vit D3 is also supplemented regularly. The child has normal growth and reports no food or medication allergy, and appears conscious, pale, and subfebrile (37.5 °C), she weighs 24.5 kg.

Physical examination reveals a SpO_2=91-92% and heartrate 115 bpm, with a general appearance of someone who is suffering from a medium to severe illness, non-purulent conjunctivitis (Figure 1), pale and later reddish lips (Figure 2), wet tong, and erythematous throat. No palpable cervical lymph nodes, but edema in the lower extremities was noted. Later on desquamation of fingers (Figures 3 and 4) of both hands and feet was noted.

Figure 1 non-purulent conjunctivitis

Figure 2 Pale and later reddish lips

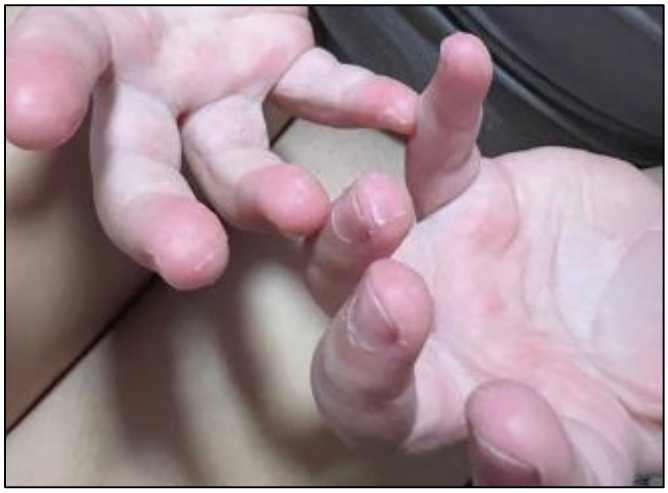

Figure 3 Fingers desquamation

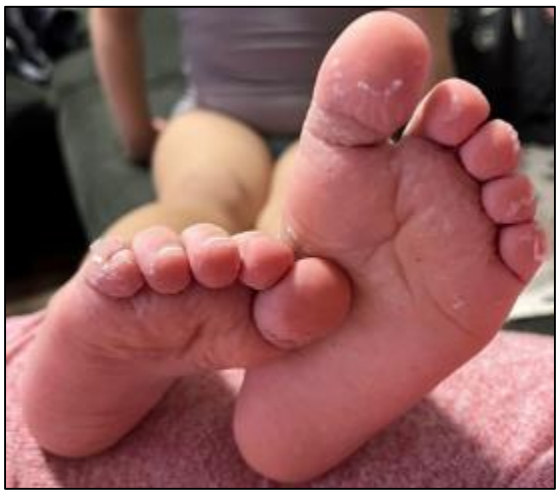

Figure 4 Desquamation of toes

Laboratory tests indicated elevated anti-streptolysin-O titers and positive C-reactive protein. Echography examination revealed dilatation of the coronary arteries in two consecutive days, prompting a request for an CT angiography that confirmed the findings (Figures 5-8). The radiological findings and the accompanying clinical findings (fever, oral mucosal changes, swelling of limbs) indicated a partial/incomplete KD diagnosis.

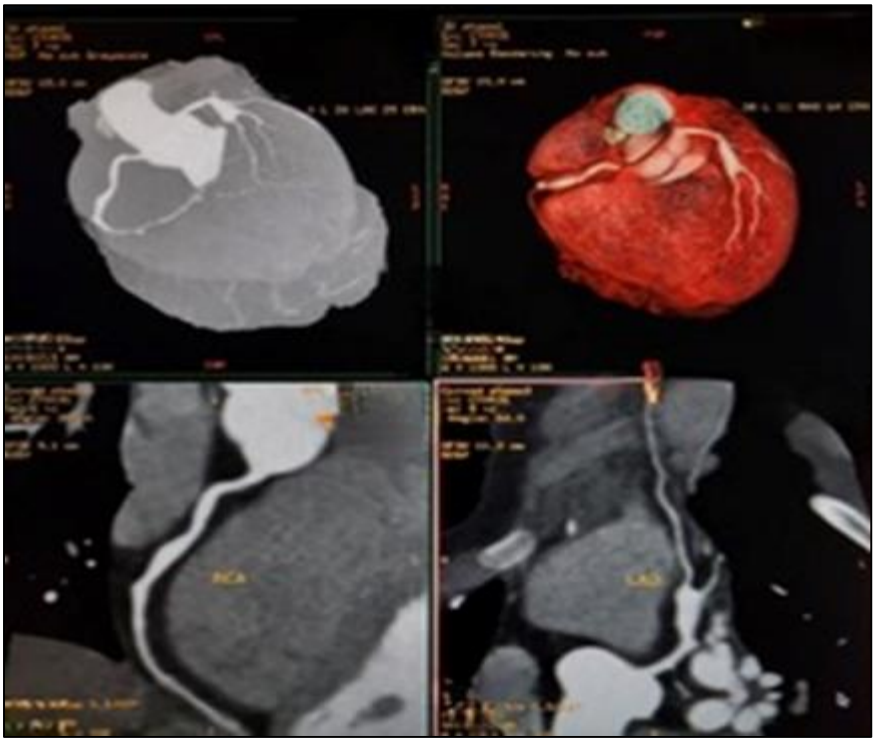

Figure 5 CT Angio refformatations ,MIP,VR

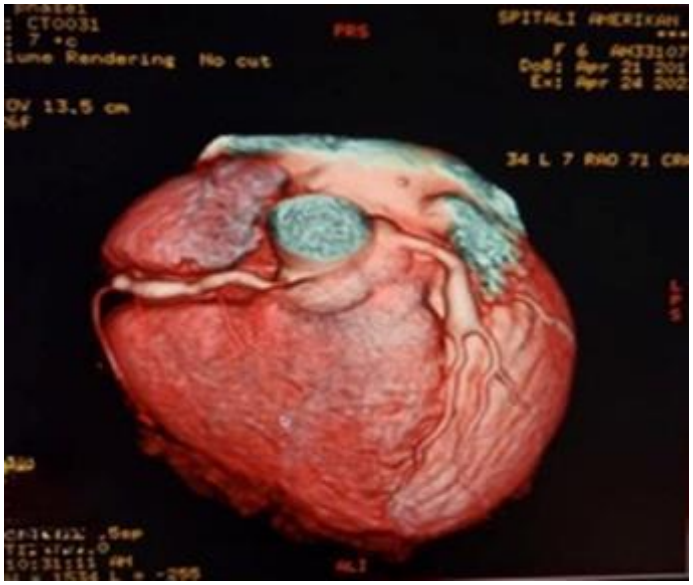

Figure 6 VR .Aneurizmal dilatation of RCA and LAD proximal

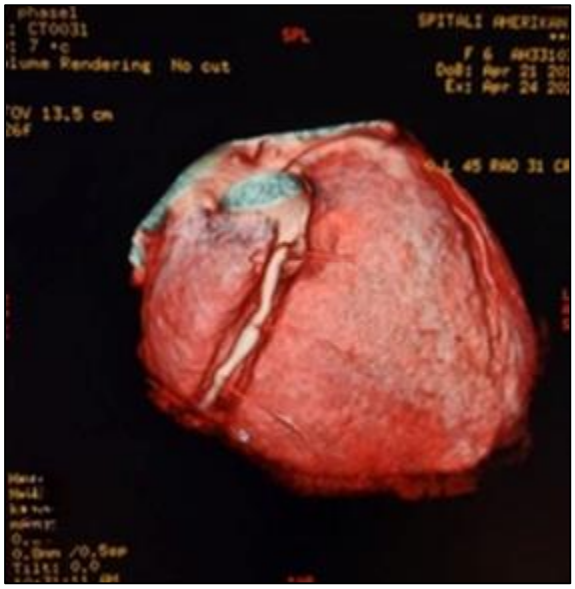

Figure 7 VR . Aneurismal dilatation of RCA

Treatment was based on high dose aspirin and steroids. Follow up after the treatment was continued for a period of 6 months with aspirin 100 mg daily. The check after 6 months was without any findings.

The table gives the timeline of presentation, examinations and findings.

Table 1 The timeline of presentation, examinations, and findings

Time	Presentation	Investigation	Findings	Management
D1	Fever, abdominal pain	Lab tests	Urinary infection, RF=14.7 IU/mL CRP=70.6 mg/L ASLO=941.7IU/mL	Antibiotics, steroids
D7	Fever, abdominal pain	ECHO	LCA dilatation (d=3.8 mm) LCx aneurism (12x7.5 mm)	W
D8		ECHO	LCA (0.3-0.5 mm) aneurism Requested CA examination	
D12		Lab tests	Urea (8.43 mmol/L) GGT (43.86 U/L) ASTO 575.4 IU/mL	
D19		CT angiography	Medial RCA aneurism (2.7 mm proximally and 4 mm distal to marginal acute artery) Medial LAD aneurism (4.5 mm) RCA dominance	Aspirin, Prednol, Neacet(?)

3. Discussion

Kawasaki disease has an unknown etiology [2]. Some of the potential causes might include autoimmune, genetic or respiratory infections [3,10,12,16]. Destruction of the elastic lamina, inflammatory cells infiltrate and smooth muscle death result in a coronary aneurism 6-8 weeks of the acute episode [1]. Despite the recovery through myo-intimal proliferation there is a continued risk of stenosis and occlusion of these vessels. In less than 1% of the patients a giant aneurysm (>8 mm) may develop, leading to coronary stenosis, obstruction, or acute thrombosis [1]. It is important to rapidly treat complications, conduct a differential diagnosis, prepare a follow-up plan and discharge training [7].

Echocardiography has a high sensitivity and specificity for the detection of abnormalities of the proximal coronary arteries [1]. Advanced cases manifest with LV dysfunction (20%) and/or myocardial inflammation (50%) [1]. The CT angiography in these ages is performed rarely and has particular characteristics that merit special attention. It is a delicate procedure that requires a combination of the spatial, temporal and contrast resolutions, in a short time while patient is holding their breath. Treatment with IVIG within 10 days from onset reduces the risks of complications [10,11].

Alternative diagnosis might include multisystem inflammatory syndrome in children (MIS-C). Some of the similar signs between KD and MIS-C include fever, rash, oropharynx congestion and red eyes [2]. Other characteristics have been reported as a rash at the site of the BCG vaccination [11] or arthritis of small joints [14]. Other publications indicate that the differential diagnosis might include non-bacterial infections, autoimmune and neoplasms in the differential diagnosis [11].

Differential diagnoses should also consider acute respiratory syndrome coronavirus type 2 SARS-CoV-2[8], due to either as similarity with upper respiratory signs; or as an infectious trigger of the immune response; or a concurrent infection of COVID-19 and KD [9]; or infection with measles, group A beta-hemolytic streptococcal infection [12] or EBV [13]. Fever after broad spectrum antibiotic, and redness at the BCG site [11] – is a suggestion for diagnosis of infants with KD, together with leukocytosis and thrombocytosis [11]. Several studies reported irritability of the patient [12]. In this case the child was diagnosed with urinary infection, and high WBC [11].

Atypical/incomplete KD reported in some studies [13]. A partial/atypical KD is when the case does not present all the signs as listed (fever and at least 4 of the 5 criteria) [11].

Treatment includes administration of high dose aspirin until the fever subsides, IVIG before the first 10 days from onset, and sometimes corticosteroids [3]. When the case is confirmed, vaccinations are delayed for one year [14]. Some studies also reported lack of IVIG for the treatment of KD [12] which was true in this case as well. Follow-up is recommended with a treatment of aspirin for 8 weeks [11], and ECHO follow-up from 6 weeks to 2 years depending on the complications [13].

4. Conclusion

This case of atypical Kawasaki Disease was diagnosed in a patient 6 years old. Only two out of the five criteria for KD were met, and after a treatment with aspirin and corticosteroids the patient recovered completely. This case highlights the necessity of clinicians to pay careful attention to similar presentations in children under 6 years old and diagnose KD swiftly to prevent complications (x aneurisms, heart failure, myocardial infarction) and death.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study through their parents.

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