OPEN ACCESS

Research Article

Infection control consequences – early Staphylococcal Scalded Skin Syndrome or Kawasaki Syndrome?

Frühes Staphylococcal Scalded Skin Syndrome (Dermatitis exfoliativa neonatorum) oder Kawasaki-Syndrom – Differenzialdiagnose mit Konsequenzen für die Infektionsprävention

Afshin Assadian[1]
Ojan Assadian[2]
Arne Simon[3]
Axel Kramer[4]

1 Department of General and Vascular Surgery, Wilhelminenspital, Vienna, Austria

2 Department for Hygiene and Medical Microbiology, Medical University of Vienna, Austria

3 Children's Hospital Medical Centre, University of Bonn, Germany

4 Institute for Hygiene and Environmental Medicine, Ernst Moritz Arndt University, Greifswald, Germany

Abstract

Childhood exanthemata are caused by a broad spectrum of common pathogens. Many exanthemata initially present very similarly, even though caused by different organisms, ranging from virus to bacteria and their respective toxins. In the majority of cases the diagnosis is only of academic value, since therapy does hardly differ. However, in some cases accurate and prompt diagnosis is paramount, since therapy and appropriate hygiene measures prevent morbidity and mortality. We present a case with two differential diagnoses, Staphylococcal Scalded Skin Syndrome and Kawasaki Syndrome, which demonstrates the importance of considering relatively rare conditions as the cause of a childhood exanthema and discuss differences in therapeutic and infection control management. From an infection control point of view, Staphylococcal Scalded Skin Syndrome is, in contrast to Kawasaki Syndrome, highly transmittable to other paediatric patients via the hands of the staff. Therefore maintaining correct hand hygiene as well as other infection control measures are of importance until the final diagnosis is established.

Keywords: Staphylococcal Scalded Skin Syndrome, dermatitis exfoliativa neonatorum, staphylogenic Lyell syndrome, Kawasaki syndrome, infection control measures

Zusammenfassung

Kindliche Exanthemata können durch ein breites Spektrum von Erregern verursacht werden. Oft präsentieren sie sich anfänglich ähnlich, obwohl sie von unterschiedlichen Erregern wie Viren, Bakterien und deren Toxine verursacht werden können. Zumeist ist die Diagnose von akademischer Bedeutung, da sich die kausale Therapie kaum unterscheidet. In einigen Fällen ist jedoch eine genaue und rasche Diagnose von höchster Bedeutung, da die korrekte Therapie und geeignete Hygienemaßnahmen Morbidität und Mortalität verhindern. Wir präsentieren einen Fall mit zwei Differentialdiagnosen, Staphylococcal Scalded Skin Syndrome (SSSS) und Kawasaki-Syndrom (KS), der die Notwendigkeit der Differenzierung von seltenen kindlichen Krankheiten aufzeigt. Unterschiede im therapeutischen und hygienischen Management werden diskutiert. Von krankenhaushygienischer Bedeutung ist, dass SSSS im Gegensatz zu KS auf andere pädiatrische Patienten über die Hände der Mitarbeiter übertragbar ist. Daher sind korrekte Händehygiene sowie weitere hygienische Maßnahmen dringend einzuhalten, bis die endgültige Diagnose vorliegt.

Schlüsselwörter: Staphylococcal Scalded Skin Syndrome, Dermatitis exfoliativa neonatorum, staphylogenes Lyell-Syndrom, Kawasaki-Syndrom, Infektionsprävention

Introduction

Childhood exanthemata are caused by a broad spectrum of common pathogens. Many exanthemata initially present very similarly, even though caused by different organisms – ranging from virus to bacteria – as well as having different rates of morbidity and mortality. In the majority of cases the diagnosis is only of academic value, since therapy does hardly differ. However, in some cases accurate and prompt diagnosis is paramount, since therapy and appropriate hygiene measures prevent added morbidity and mortality.

We present a case which demonstrates the importance of considering relatively rare conditions as the cause of a childhood exanthema. The one main differential diagnosis, Staphylococcal Scalded Skin Syndrome (SSSS), also known as dermatitis exfoliativa neonatorum or staphylogenic Lyell syndrome, is caused by group II coagulase-positive staphylococci, usually phage type 71, which elaborate exfoliatin (also called epidermolysin), a toxin that splits the upper part of the epidermis just beneath the granular cell layer [1]. The infection often begins during the first few days of life in the umbilical stump or diaper area; in older children, the face is the typical site. Toxin produced in these areas enters the circulation and affects the entire skin. The differential diagnosis includes drug hypersensitivity, viral exanthemas, scarlet fever, thermal burns, genetic bullous diseases (e.g. some types of epidermolysis bullosa), acquired bullous diseases (e.g. pemphigus vulgaris, bullous pemphigoid), toxic epidermal necrolysis and rare the Kawasaki syndrome. Kawasaki syndrome (KS) or Kawasaki disease is an acute febrile illness of unknown etiology that primarily affects children younger than 5 years of age. KS is characterized by fever, rash, swelling of the hands and feet, irritation and redness of the whites of the eyes, swollen lymph glands in the neck, irritation and inflammation of the mouth, lips, and throat. Serious complications include coronary artery dilatations and aneurysms. The standard treatment with intravenous immunoglobulin and aspirin substantially decreases the development of these coronary artery abnormalities [2]. For epidemiologic surveillance, CDC defines a case of KS as illness in a patient with fever of 5 or more days duration (or fever until the date of administration of intravenous immunoglobulin if it is given before the fifth day of fever), and the presence of at least 4 of the following 5 clinical signs [3]:

- rash
- cervical lymphadenopathy (at least 1.5 cm in diameter)
- bilateral conjuctival injection
- oral mucosal changes
- peripheral extremity changes.

Because SSSS is, in contrast to Kawasaki Syndrome, highly transmissible to other paediatric patients via the hands of the staff, isolation of cases is recommended, whereas for KS isolation of cases is not required.

Case report

A 15 month old boy presented with generalised rash for two weeks, runny nose for one week and off feeds. No fever was noted by his mother. One day prior to admission the family's general practitioner prescribed 250 mg Flucloxacillin, 8 hourly, of which only one dose was taken. The patient was known to suffer from atopy. His asthma was treated with Salbutamol 100 µg inhalers for the last 4 months, his eczema with hydrocortisone cream. No allergies were known. The little boy has had his Pertussis, Tetanus, Diphtheria, Haemophillus influenza B and oral Polio vaccinations at 2, 3 and 4 months of age. There were no recent vaccinations.

On inspection the patient was irritable and crying, a generalised erythematous rash was present with mild exfoliation in the axilla, the back of the neck and ears. A perioral and periorbital rash with fissured, cracked lips was also present. The pulse was 120 beats per minute, the systolic blood pressure 90 mm Hg, the respiration rate 30 per minute, the temperature 37.0°C, the capillary return was 1 second, the oxygen saturation 98% in air. His pupils were equally reacting to light, and normal fundi and a marked conjungtival injection were noted. The patient had a red tongue and a slightly inflamed throat. A few markedly enlarged cervical lymph nodes were also present. The other systems did not reveal any abnormalities.

Clinical impression and differential diagnoses

One of the differential diagnoses was SSSS because of generalized erythematous rash, exfoliation of the skin and conjunctival injection. In SSSS a scarlatiniform erythema develops diffusely and is accentuated in flexural and periorificial areas. The inflamed conjunctivae occasionally become purulent. Characteristically, circumoral erythema is prominent as is radial crusting and fissuring around the eyes, mouth and nose. At this stage the patient may be Nikolski positive. The desquamative phase usually begins after 2–5 days of cutaneous erythema. The healing process occurs in 10–14 days without scarring. SSSS occurs predominantly in children under 5 years of age and includes a range of different skin manifestations, ranging from localized bullous impetigo to generalized cutaneous involvement with systemic illness ([4], p. 1891-2).

Even though no pyrexia was observed, there was a strong suspicion of KS, since four of the six criteria for diagnosing KS were present. The diagnosis of KS depends on the identification of clinical criteria, which are the following: (1) fever for at least 5 days, (2) nonexudative conjunctival injection, (3) erythema of the oropharynx and lips (with cracking), strawberry tongue, (4) acute no purulent lymphadenopathy (one or more nodes of at least 1.5 cm in diameter), (5) polymorphous erythematous exanthema, (6) at least one of the following: erythema of the palms

and soles, oedema of the palms and soles, desquamation of the tips of the digits and around the nails [5]. KS is diagnosed if 5 or more of the above named criteria are present. There are also some distinctive, however non-diagnostic, laboratory features in KS: elevated Erythrocyte Sedimentation Rate (ESR), leukocytosis, thrombocytosis, sterile pyuria, moderately elevated transaminase levels and elevated level of acute phase proteins [6].
Another quite common cause for a similar presentation is Toxic Epidermal Necrolysis (TEN), which appears to involve a hypersensitiviy reaction that results in damage to the basal cell layer of the epidermis. It is triggered by a variety of drugs and vaccinations such as Diphtheria and Measles. The criteria for diagnosing TEN are: (1) widespread blister formation and morbilliform or confluent erythema, associated with skin tenderness, (2) absence of target lesions, (3) sudden onset and generalisation within 24 hours, (4) the histological finding of a full thickness epidermal necrosis and minimal to absent dermal infiltrate. In TEN the Nikolsky sign is only positive at the site of erythema, the desquamation does not occur over macroscopically unaffected skin ([4], p. 1852).

Diagnosis

While awaiting the results of the investigations, the patient's skin suddenly started to peel dramatically and developed a generalized positive Nikolsky sign.
Due to the absence of pyrexia, a normal CRP, a normal platelet count, no recent history of vaccination, a prolonged onset of more than 24 hours and the massive desquamation, the admitting team excluded KS and TEN. The clinical diagnosis of SSSS was established. As a consequence further investigations such as Electro Cardiogram (ECG) and echocardiography were not performed. The blood cultures were negative after 48 hours and the skin swabs grew heavy mixed commensal skin flora.

Discussion

In Japan and the United States of America KS has become the most common cause of acquired heart disease in children [7]. The incidence of KS in Great Britain is 3.4 per 100.000 children under 5 years [8] as compared to 172.2 per 100.000 children under 5 years in Japan [9]. The serious complications of KS are systemic vasculitis resulting in coronary artery aneurysmata (15–30%) [10] and myocardial infarction or aneurysm rupture. The case fatality rate of 3.7% observed in KS patients in Great Britain in 1990 [8] is unfavourably high compared to 0.1% in Japan [9]. These figures may be a direct reflection of the fact that British doctors are not that commonly exposed to patients with KS and hence do not include KS in their differential diagnosis. It is also very important to consider KS if less than 5 criteria are present, as occurring in the well described atypical KS. In these patients it is recommended to perform serial echocardiography including 2 examinations in the first 3 weeks after onset [11] the treatment of choice is 30 mg/kg acetylsalicylic acid daily until the fever settles as well as intravenous immunoglobulins, 2 g/kg, as a single infusion within 10 days of onset. This significantly decreases the incidence and severity of aneurysm formation as well as relieving symptoms [2]. There is no clear evidence of the infectious etiology of this condition and infection control measures do not appear to be necessary.
However, therapy for SSSS requires intravenous antibiotic treatment, directed towards eradication of the exfoliating toxin producing Staphylococcus aureus, supportive skin care, and attention to the fluid and electrolyte balance. Furthermore it is important not to hesitate in commencing analgesia, since the patients suffer from marked skin tenderness. From an infection control point of view, SSSS is, in contrast to KS, highly transmittable to other paediatric patients via the hands of the staff. Therefore correct hand hygiene as well as barrier nursing is important until the diagnosis is established. Diagnosis is either clinical or via skin biopsy, which shows subcorneal granular layer split and as a characteristic feature the absence of inflammatory infiltrate [3].
In TEN it is paramount to appreciate etiologic factors, particularly when the symptoms are drug induced. In those circumstances the administration of the drug must be discontinued immediately. Management is directed towards correcting fluid and electrolyte imbalances as well as treating secondary skin infections. Analgesia therapy is also very important since marked skin tenderness is present.
SSSS is, in contrast to KS, highly transmittable to other patients [12]. In this situation, strict application of infection control measures and the importance of prompt isolation of all infectious patients and those with presumed or possible infection until an infectious etiology has definitely ruled out cannot be overemphasised.
In staphylococcal infections the most important route of transmission is direct by contact from one patient to another and indirect by the hands of the staff. Therefore, by application of general infection control measures and barrier nursing, as long as they are perfectly practised, strict isolation in a single room is not warranted. However, implementation of infection control in children is aggravated by the normal behaviour of the children themselves. Close contact of young children is almost constant unless children are specifically segregated. Hence, to facilitate infection control measures in paediatric patients infected or colonised with pathogenic organisms, contact isolation in a private room should be practised.
When a private room is not available, special emphasis has to be placed on selecting the roommates for a possibly infectious patient. Immunocompromised patients and patients who are about to undergo extensive surgery with insertion of prosthetic devices should not be chosen as roommates for children with suspected SSSS. Also, patients with impaired kidney function may also be at high risk of acquiring complications after colonisation with exotoxin producing strains of Staphylococcus aureus,

since the exotoxin, which is produced by a localised infection and released into the blood stream, is primarily eliminated through the kidneys.

Hand disinfection remains the single most important means of preventing nosocomial infections. Barriers should be used as needed when soiling of clothing, skin or mucous membranes is anticipated. Gloves should be used for anticipated contact with mucous membranes, intact and non-intact skin and all secretions. Meticulous hand disinfection has to be performed after gloves are removed and before taking care of another patient.

Gowns are worn only when patient contact is likely to lead to direct soiling of garments with infective material. The likelihood of contamination or soiling usually determines whether a health-care worker needs to wear a gown or other barriers, such as plastic aprons.

The use of masks is eventually indicated for those who care in a close contact for the patient and droplet transmission by inducing aerosol during skin-manipulations can not be excluded. Linen and contaminated dressings must not be handled with fingers but should immediately be placed, with forceps or gloves, into a container or bag which is immediately closed and sealed. Double bagging is not needed unless the outside of the bag is soiled.

In general, high standards of cleaning, avoiding overcrowding, minimising inter-ward transfers and maintaining adequate levels of trained ward staff should be maintained.

References

1. Resnick SD. Staphylococcal toxin mediated syndromes in childhood. Semin Dermatol. 1992;11(1):11-8.
2. Newburger JW, Takahashi M, Beiser AS, Burns JC, Bastian J, Chung KJ, Colan SD, Duffy CE, Fulton DR, Glode MP, et al. A single intravenous infusion of gamma globulin as compared with 4 infusions in the treatment of Acute Kawasaki Syndrome. N Engl J Med. 1991;324(23):1633-9.
3. Rowley AH, Shulman ST. Kawasaki syndrome. Clin Microbiol Rev. 1998;11(3):405-14.
4. Behrman RE, Kliegman RM, Arvin AM, editors. Nelson Textbook of Pediatrics. 15th ed. Philadelphia: Saunders; 1997.
5. American Heart Association Commitee on Rheumatic Fever, Endocarditis, and Kawasaki Disease. Diagnostic Guidelines for Kawasaki Disease. Am J Dis Child. 1990;144(11):1218-9.
6. Rowley AH, Gonzales Cruzi F, Shulman ST. Kawasaki Syndrome. Curr Probl Pediatr. 1992;21(9):387-405. DOI: 10.1016/0045-9380(91)90008-9
7. Leung DY, Cody Meisner H, Fulton DR, Murray DL, Kotzin BL, Schlievert PM. Toxic shock syndrome toxin secreting Staphylococcus aureus in Kawasaki syndrome. Lancet. 1993;342(8884):1355-8. DOI: 10.1016/0140-6736(93)92752-F
8. Dhillon R, Newton L, Rudd PT, Hall SM. Management of Kawasaki Disease in the British Isles. Arch Dis Child. 1993;69:631-66. DOI: 10.1136/adc.69.6.631
9. Kawasaki T. Kawasaki Disease. Asian Med J. 1989;32:497-506.
10. Kato H, Koike S, Yamamoto M, Ito Y, Yano E. Coronary aneurysms in infants and young children with acute mucocutaneous lymph node syndrome. J Pediatrics. 1975;86(6):892-8. DOI: 10.1016/S0022-3476(75)80220-4
11. Management of Kawasaki Syndrome: A consensus statement prepared by North American participants of the Third International Kawasaki Symposium, Tokyo, Japan, Dec 1988. Pediatr Infect Dis J. 1989;8(10):663-7.
12. Department of Health and Human Services, Centers for Disease Control and Prevention (CDC). Kawasaki Syndrome Case Report. Atlanta, Georgia: CDC; 2003. Available from: http://www.cdc.gov/kawasaki/docs/ks_case_report-print.pdf

Corresponding author:
Ojan Assadian, MD, DTMH
Department for Hygiene and Medical Microbiology, Medical University of Vienna, Währinger Gürtel 18-20, 1090 Vienna, Austria, Fax: +43-1-40400-1907
ojan.assadian@meduniwien.ac.at

Please cite as
Assadian A, Assadian O, Simon A, Kramer A. Infection control consequences – early Staphylococcal Scalded Skin Syndrome or Kawasaki Syndrome? GMS Krankenhaushyg Interdiszip. 2009;4(2):Doc05.

This article is freely available from
http://www.egms.de/en/journals/dgkh/2009-4/dgkh000130.shtml

Published: *2009-12-16*