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Ritter's Disease in Child – A Case Report

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ABSTRACT

Ritter's disease which is medically known as Staphylococcal Scalded Skin Syndrome (SSSS), is a skin condition induced by exfoliative toxins produced by group II coagulase-positive staphylococci. This disease is most common in children and neonates which has high chances to be fatal if treatment is not initiated as early as possible. In this report, a case of a 2 year old child is being presented, who came with the complaints of erythematous lesions over lower jaw and neck for four days after which the lesions progressed to the right eye. The lesions were associated with peeling of skin indicating positive Nikolsky's sign. Based on clinical examinations a diagnosis of Staphylococcal Scalded Skin Syndrome was established. The patient responded favorably to the treatment which included i.v. antibiotics (ampicillin and cloxacillin), analgesics (syrup Paracetamol) and other supportive measures. Patient was discharged after six days on complete resolution of symptoms.

KEYWORDS: *Ritter's disease, Staphylococcal Scalded Skin Syndrome, exfoliative toxins, Nikolsky's sign, antibiotic treatment*

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INTRODUCTION

Ritter's disease which is medically known as Staphylococcal Scalded Skin Syndrome (SSSS), is an acute epidermolysis caused by exfoliative toxins produced by 5% of staphylococcus aureus. Group II coagulase-positive staphylococci, is the major organism involved [1]. This organism elaborates two types of exfoliatins, ETA and ETB. The exact mechanism by which the toxin produces exfoliation is still unknown; however several studies suggest that it follows a proteolytic mechanism. The crystal structures of both ETA and ETB were similar to the serine proteases of the chymotrypsin family and specifically to that of the glutamic-acid-specific proteases [2].The molecular target of these toxins is the Desmoglein-1 protein, which is expressed mainly in the superficial upper layers of the skin epidermis. Desmoglein 1 is a desmosomal cadherin responsible for the integrity of the cell-to-cell adhesive structures. The cleavage of Dsg-1 results in the destruction of desmosomal cell-cell attachments in the superficial layer of the skin. Macroscopically, this manifests as epidermal detachment, the primary symptom of SSSS [3]. Bullae forms and diffuse sheet like desquamation occurs. Two types of SSSS are thought to exist: a localized form, in which there is only a real involvement of the epidermis, and a generalized form, in which significant areas are involved, remote from the initial site of infection [4].The primary infection in neonates 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. Staphylococcal scalded skin syndrome is most common in children and neonates due to their poorly defined immune system [5]. The diagnosis of SSSS is by clinical examination and by collecting medical history. The tests of skin biopsy and culture examination may also be performed to confirm the disease condition. Treatment for SSSS remains supportive care and anti staphylococcal antibiotics despite the condition being toxin-mediated [6].

CASE REPORT

A 2 year old female child came with the complaints of erythematous lesions first noticed over lower jaw and neck for 4 days and then noticed over the groin and umbilicus. On day 5 these erythematous lesions were noticed on the right eye and the child had complaints of pain and irritability upon touching the lesions. Child also complained of itching of palm and soles. She also had associated wet cough for 4 days and a fever spike on 3rd day. On day 5 she was brought to the hospital and clinical and laboratory investigations were done. Upon head to foot examination she was found to have erythematous macule below right eye and around lower jaw. These macules showed peeling upon minimal pressure. This was an indication of positive Nikolsky's sign, in view of which an initial diagnosis of Staphylococcal Scalded Skin Syndrome (SSSS, Ritter's disease) was done. The patient was then immediately initiated on oral Ampiclox. On day 6 the child complained of perioral reddness and reddness all over the body. The patient was switched to i.v. Ampiclox (275mg, 6 hourly). Syp. paracetamol (150mg, s.o.s.) was initiated in view of fever. The laboratory investigations showed leucocytosis (11,100 cells/cu.mm.), mild neutropenia (21%), eosinophilia (17%) and borderline ESR levels.CRP was negative and blood culture showed no growth. The rest of the investigations were normal. Syp Atarax (5mg B.D.) was initiated in view of eosinophilia. By the 4th day of admission (day 8 of disease) child improved showing healing of the lesions, no new lesions were observed and pain and itching sensations were reduced. On day 5 the child was discharged with advice to continue syp. Amoxicillin-clavulaunate (285mg, B.D.) for 3 days and saline compress for 3 days.

DISCUSSION

The first description of SSSS was done by a German physician who observed the condition in young children in a Czechoslovakian foundling asylum in 1878[7]. The exfoliative toxins involved in this condition are responsible for causing a spectrum of disease ranging from localised blisters to extensive exfoliation, which has previously been called dermatitis exfoliativa, pemphigus neonatorum, Lyell's disease and Ritter's disease. SSSS is rarely fatal in children and in adults it carries a mortality of over 50% who usually have serious underlying medical problems [8]. It can be fatal in children when diagnosis and appropriate treatment are delayed.

A diagnosis of staphylococcal scalded skin syndrome is based upon identification of characteristic symptoms, a thorough clinical evaluation, and a detailed patient history. Cultures can be taken from areas that harbor the bacterial germ including the conjunctiva (corners of the eyes), nasal passages, umbilicus and nasopharynx area. The confirmation usually requires biopsy. A biopsy can reveal non-inflammatory superficial splitting of the epidermis, which is indicative of the disorder and can differentiate it from similar disorders [9]. The results of biopsy may take time, which can delay the treatment and can be fatal for the patient. In this case diagnosis was purely based on clinical examination and that avoided the chances of mortality. SSSS presents as a macular erythema followed by diffuse epidermal exfoliation. General malaise, fever, irritability, skin tenderness may be noted.Other signs which may also be present, are facial edema, conjunctivitis, and perioral crusting. Mucous membranes are spared, but dehydration may be present and significant. Nikolsky's sign is one of the major signs to be noted [1]. An elevated WBC count, ESR levels and nikolsky's signs are the immediate diagnosis parameters which were clearly shown by this patient.

Penicillinase-resistant anti staphylococcal antibiotics are the treatment of choice in this disease. In this patient ampicillin-cloxacillin was the antibiotic started. The stratum corneum was quickly replaced, and healing occurred within 4 days after start of treatment. The conditions which could be considered as a differential diagnosis was toxic epidermal necrolysis (TEN). In TEN however mucus membrane involvement is seen in this patient it was not observed. In SSSS, blistering affects only the superficial skin and not the mucosa or deeper skin layers. This phenomenon is due selectivity of the desmoglein cleavage by the ETs and the differential expression of particular desmogleins in different layers of the skin and mucosa. The ETs selectively hydrolyze Dsg-1, whereas Dsg-3 remains unaffected. Dsg-1 is expressing all strata of the skin, whereas Dsg-3 is only expressed in deeper strata [10]. Therefore, in the deep layers of the skin, the disruption of Dsg-1 by ETs is compensated by Dsg-3 and exfoliation only occurs in the stratum granulosum, where Dsg-3 is not present [11]. This explains the non-involvement of the mucous membranes. An elevated ESR levels were seen in this patient, which can prompt many physicians to initiate a corticosteroid therapy also this being the treatment option for TEN. The use of corticosteroid therapy in SSSS can complicate the condition and predispose the child to infections like sepsis and prolong the prognosis of the disease [12].

CONCLUSION

Ritter's disease occurs almost exclusively in infants and children under the age of 6. It is a potentially fatal condition but can respond well to conventional treatment if prompt diagnosis is made. Robust hygiene measures are also imperative for its successful management .Due to high prevalence of fever and exanthema in children there is high chance of misdiagnosis that can occur for this disease, especially as Staphylococcal scalded skin syndrome is mainly a clinical diagnosis. This case raises awareness on the importance of a multidisciplinary approach to be done in patients presenting with cutaneous symptoms of unknown etiology.

ABBREVIATIONS

SSSS: staphylococcal scaled skin syndrome
ET: Epidermolytic Toxin
ETA: Epidermolytic Toxin A
ETB: Epidermolytic toxin B
Dsg-1: Desmoglein 1
Dsg-3: Desmoglein 3
TEN: Toxic Epidermal Necrolysis
ESR: Erythrocyte Sedimentation Rate
WBC: White Blood Cells
CRP: C-Reactive Protein

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