

Adult Staphylococcal Scalded Skin Syndrome (SSSS)

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Investigations

Complete blood picture, renal and liver function tests were normal. C-reactive protein was raised to 54.6 mg/l (normal value <9.9 mg/l). Skin swab from umbilicus and left groin grew *Serratia marcescens*. Exfoliative toxin assay was not performed, as it was not currently available in Hong Kong.

CASE SUMMARY

History

Miss Wong, a 20-year-old student, was referred to us for evaluation of suspected toxic epidermal necrolysis (TEN). She was well all along until two months ago when an itchy rash developed over her trunk and flexures. The rash improved with topical steroid and emollient prescribed by the referring doctor. One week prior to the present consultation, the patient noticed a painful sensation over her trunk and flexures followed by peeling of her flexural skin. This was not preceded by any symptom of upper respiratory tract infection and the mucosal areas were uninvolved. Otherwise the past medical history, drug history and family history were unremarkable.

Physical examination

The general condition of the patient was good except for a low-grade fever. Generalized erythematous patches were noted over the trunk, waist, bilateral groins, elbows, neck and popliteal fossae symmetrically (Figure 1). Erosion was found over the para-umbilical area, upper abdomen and submammary areas. Peeling of skin was noted over bilateral axillae and right groin (Figure 2). However no bulla was detected. Oral mucosa was spared. Nikolsky's sign was negative.

Differential diagnosis

The differential diagnoses included staphylococcal scalded skin syndrome, toxic epidermal necrolysis, erythema multiforme, viral exanthema and other autoimmune bullous diseases.



Figure 1: Generalized erythema over the body as if being 'scalded' by hot water



Figure 2: Peeling of the skin is evident on both groins especially on the right side

Skin biopsy was performed at the abdominal skin, which showed epidermal parakeratosis, subcorneal polymorphs and focal superficial denudation suggestive of acantholysis. Epidermal spongiosis and superficial perivascular predominant lymphocytic infiltrate was apparent. Gram stain and PAS stain did not demonstrate any microorganism. Immunofluorescence study showed only trace dermoepidermal junction IgM staining. The biopsy was consistent with a diagnosis of staphylococcal scalded skin syndrome with underlying eczema. TEN was not made as a diagnosis in this case as there was no history of prior oral medication, the area of skin detachment was less than 5% and the patient had a prompt and uneventful recovery. Besides, the histological findings were incompatible with TEN as only the subcorneal layer was involved.

Diagnosis

A diagnosis of staphylococcal scalded skin syndrome was made based on both the clinical and histopathological findings, although the culture did not grow staphylococcus aureus.

Treatment

Patient was given a course of antibiotics with ampicillin and cloxacillin. In view of the generalized erythema and erosions, potassium permanganate bath and fucidin cream was prescribed.

Progress

The erosions healed without scarring in a few days and the patient was discharged.

REVIEW ON ADULT STAPHYLOCOCCAL SCALDED SKIN SYNDROME

SSSS was first described in children by Von Rittershan in 1878. In 1972, Levine and Nordon described the first case in an adult.¹ Up to the year 2000, approximate 40 cases of adult SSSS had been reported in the literature.

SSSS is not uncommon in infants and young children below five years of age,² but it is rare in adult. Its rare occurrence is probably related to (i) protective

antibodies to exfoliative toxin (ET), (ii) better ability to metabolize the toxin and (iii) rapid toxin excretion.

Among the reported cases, there was a male predominance of 2:1. The age of onset ranged from 10 to 89 with 50% occurring before the age of 50. While some patients had no underlying diseases, predisposing factors included kidney failure,³ underlying malignancy,⁴ immunosuppression (such as HIV infection,⁵ chemotherapy and long term steroid), drug addiction and alcoholism.

Clinically, patients with SSSS may have fever and malaise developing a few days after localized staphylococcal infection. They may develop tender diffuse erythematous eruption with flaccid bullae. Nikolsky's sign is usually positive. Rarely large sheets of skin can be easily peeled off revealing moist erythema (scalded skin). The face, axillae, groin and neck are commonly involved. With appropriate treatment, the erosion rapidly dries and desquamation occurs in a few days. Prognosis is usually good in children, but is guarded in adult cases.

The isolation of the causative organism is not always successful. In the reported cases, exfoliative toxin (ET) producing staphylococcus aureus had been isolated from 45% of mucocutaneous lesions, which included vesicles, bullae, pustule, erosions, abscess, discharging sinus and conjunctivae. Bacteraemia occurred in more than 50% of cases, this was usually associated with a poor prognosis (a mortality rate of >50%). Those without underlying disease usually recovered uneventfully. The portal of entry was related to indwelling catheter, abscess, septic arthritis, infected A-V shunt and parenteral injection, but in many cases, no primary infective site was identified.

The causative organism, staphylococcus aureus, has three phage groups (Types I, II and III). All phage groups have the ability to produce both or either one of the two types of ET (ETA and ETB).⁶ ET can be detected by the following methods:

1. Newborn mouse bioassay:⁷ when exfoliative toxin-producing staphylococcus aureus culture supernatant is injected subcutaneously into a newborn mouse, Nikolsky's sign can be observed a few hours later. This method is non-specific and cannot distinguish between ETA and ETB.
2. Immunologic methods, for example double immunodiffusion, slide latex hemagglutination, radio-

immunologic assay, ELISA, and electrosyneresis. These methods are toxin specific.

3. Molecular biology. Two oligo-nucleotide probes are synthesized that are specific for a part of the genes coding for ETA and ETB. This method allows detection of strains that produce low levels of toxin in vitro.

The actual mechanism of ET causing SSSS is unknown. ET could have a lipase or protease activity causing intraepithelial clefting among keratinocytes.

The diagnosis of SSSS is usually based on (i) clinical features, (ii) culture of causative organism (iii) identification of ET and (iv) biopsy finding. Differential diagnosis includes TEN, erythema multiforme, autoimmune blistering disorders and viral exanthem.

Treatment consists of penicillinase-resistant, semi-synthetic penicillin. Supportive care especially careful handling of patient is essential as the patient's skin may easily come off.

Learning Points

Dermatologists should consider SSSS in suspected case of TEN and early differentiation of SSSS from TEN is essential to avoid mismanagement.

References

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