Staphylococcal scalded skin syndrome (SSSS) is a generalized exfoliative dermatitis complicating infection from exfoliative toxin-producing strains of *Staphylococcus aureus*. The disease is most commonly seen in infants and children, and may result in large outbreaks in neonatal nurseries. It was first described in children by von Rittershain in 1878.\(^1\) In 1972, the first case of adult SSSS was reported.\(^5\) The syndrome usually occurs in adult patients who have renal disease or immunosuppression, and the risk of death from it is less than 5% among children, but as high as 60% in adults.\(^3,4\)

SSSS is usually related to phage group II *Staphylococcus aureus* that produces epidermolytic exotoxin,\(^6,7\) which is a heat-stable, chromosomally mediated product with both species and tissue specificities.\(^1,7\) Two serologic forms of exfoliative toxin have been independently isolated by several laboratories and are now referred to as exfoliatin A (ETA) and exfoliatin B (ETB).\(^8,9,10,12\) ETA is the most common toxin responsible for SSSS and is generally produced by *S. aureus* belonging to phage group II. In contrast, ETB alone has been isolated mainly from nonphage group II *S. aureus*.\(^11\) In patients with bullous impetigo, the toxin produces blisters locally at the site of infection, whereas in cases of scalded-skin syndrome, it circulates throughout the body, causing blisters at sites distant from the infection.\(^3\)

We describe a case of staphylococcal scalded skin syndrome in an immunosuppressed adult with renal insufficiency.
Case report

A 17-year-old Thai woman was admitted to hospital for 2 days because of high grade fever and a chill. She was diagnosed as having had end stage renal disease from lupus nephritis class IV for 6 months. Two cycles of intravenous cyclophosphamide and pulse methylprednisolone were unsuccessful. Current medications included prednisolone at 10 mg/d, furosemide, amlodipine, hydralazine, omeprazole, simvastatin, sodium bicarbonate, and erythropoietin. She had been on hemodialysis twice a week for 4 months. Physical examination revealed a temperature of 39.8°C, blood pressure of 130/70 mmHg, and inflammation and foul-smelling pus on the skin around the permanent catheter at the right subclavicular region. One day after admission, she developed multiple blisters on the nasal bridge, chest, and abdomen, and multiple erythematous patches on the perioral area, trunk, and extremities. The lesions began on the face and then appeared on the trunk and extremities. The blisters ruptured quickly and the erythematous patches desquamated into erosive patches (Fig. 1). Laboratory findings showed a hemoglobin level of 9.9 g/dL, hematocrit of 30%, white blood cell count of 30,700 /mm³ with 84% neutrophils, platelet count of 90,100/mm³ and blood creatinine of 12.1 mg/dL, total complement activity (CH50) of 96.1 CAEUnit, C3 level of 932.0 μg/mL, C4 level of 312.0 μg/mL, and ANA titer of 1:80. A chest x-ray revealed nothing remarkable. Gram’s staining of the pus was positive for gram-positive cocci in cluster. Five specimens of blood culture were taken at 1-hour intervals; 2 specimens were from the permanent catheter and 3 from the peripheral veins, and a of them were positive for methicillin-sensitive S. aureus. An echocardiogram was negative for infective endocarditis. The patient diagnosed with staphylococcal scalded skin syndrome. She was treated with intravenous cloxacillin at 4 gm/d and the catheter was removed. One week after the treatment, the patient recovered (Fig. 2) and a repeated hemoculture was negative for bacteria.

Discussion

Staphylococcal scalded skin syndrome (SSSS) is a common disorder that is usually seen in infants and children, but rarely in adults. Renal failure and immunosuppression are the most important risk factors among adult patients, and these were found in our patient. Other asso-
associated diseases included malignancy, diabetes mellitus, and/or HIV infection. SSSS in a healthy adult has also been reported.

The first symptoms of SSSS are usually fever and malaise followed by prodrome of sore throat or conjunctivitis. Tender, erythematous patches of skin rapidly evolve into flaccid bullae on the face and flexural areas, which are Nikolsky’s sign-positive. The bullae rupture easily, leaving superficial erythematous erosions that resemble “scalded” skin. The lesions of SSSS can be similar to other bullous diseases, particularly toxic epidermal necrolysis (TEN). In TEN, the lesions almost always involve mucous membranes, and the patients usually have a history of drug intake.

Histopathologic examination is useful for distinguishing between these two conditions at an early stage. The findings in TEN include splitting at the dermo-epidermal level, with full thickness epidermal necrosis, and basal layer vacuolar degeneration. SSSS, on the other hand, demonstrates subcorneal splitting at the granular layer without evidence of cellular necrosis or inflammatory infiltrate.

We diagnosed this case as SSSS without a skin biopsy because of typical clinical manifestation, *S. aureus* in the hemocultures, and the response to antibiotics.

Two exfoliative toxins (ETA and ETB) are the exotoxin responsible for SSSS. They act as proteases that target the protein desmoglein-1 (Dsg 1); a desmosomal cadherin protein that functions in cell-to-cell adhesion. This protein is found in the superficial granular layer of the epidermis, resulting in acantholysis and subcorneal separation of the upper layer of the skin.

The increased frequency of staphylococcal scalded skin syndrome in children may be explained by the relative quantity of Dsg 1 in the skin, which varies with age, and because immature renal function in children may impair clearance of circulating exotoxins. Another theory suggests that the exfoliative toxins may possess a superantigenic activity. The presence of antibodies specific for exotoxins may decrease the frequency of SSSS in adults. SSSS caused by methicillin-resistant *Staphylococcus aureus* was also reported.

Since the mortality rate is much higher in adults, it is important to recognize SSSS early and treat it intensively with antibiotics.

**References**

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กลุ่มอาการแผลพองลวกจากเชื้อ Staphylococcus aureus ในผู้ใหญ่ที่เป็นโรคลูกฝาหนึ่งราย: รายงานผู้ป่วยและทบทวนบทความ

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บทคัดย่อ ภาวะ Staphylococcal scalded skin syndrome พบได้บ่อยในผู้ป่วยเด็กแต่พบได้น้อยมากในผู้ใหญ่ซึ่งมักพบมาร์คเดิมหรือได้รับยาลดภูมิภูมิต้านทาน รายงานนี้เป็นรายงานในผู้ป่วยหญิงไทย อายุ 17 ปีที่สัมผัสในโรงพยาบาลต่างอ้าวการไขมันสูงนาน 6 เดือน ceiving การรักษาด้วยยาฟอกเลือดผ่านเครื่องไตเทียม 2 ครั้งต่อสัปดาห์และยาฟอร์มิโดซิโน 10 มิลลิกรัมต่อวัน ตรวจพบว่าหนังบริเวณรอบๆสายหลอดบริเวณผิวหนังชุงและมีแผลในกลุ่มต่างๆ หนังวัสดุจากจ้ากับไขมันในโรงพยาบาลผู้ป่วยผู้ป่วยมีสิ่งมีความผิดปกติทางผิวหนัง ผิวหนังของแผลเป็นแดงและเป็นแผลที่ผิวหนังของขาและหัวใจ ผลหลังจากผิวหนังเป็นชั้นเพื่อให้ผิวหนังชุ่มชื่นได้รับการวินิจฉัยเป็น Staphylococcus aureus ผู้ป่วยตอบสนองดีจากการรักษา รายงานนี้มีประโยชน์ในการรักษาผู้ป่วยผู้ป่วยที่มีภาวะมาจากโรคกลุ่มต่างๆ ที่ผ่านมาและมีการตอบสนองจากการรักษาด้วยยาฟอร์มิโดซิโนและยาฟอกเลือดผ่านเครื่องไตเทียม เขียนในวารสาร 2553;49(3):117-121.

คำสำคัญ: กลุ่มอาการแผลพองลวกจากเชื้อ Staphylococcus aureus ผิวหนัง ไขมัน ได้รับการวินิจฉัยจากโรคกลุ่มต่างๆ 