Norwegian Scabies and a Toxic Shock Syndrome Toxin 1–Producing Strain of Staphylococcus aureus Endocarditis in a Patient with Trisomy 21

Norwegian scabies is a rare ectoparasitic infestation by large numbers of the mite Sarcoptes scabiei var. hominis [1]. The condition is characterized by hyperkeratotic lesions of the hands, feet, and scalp; large warty crusts of skin are also present that are irregularly thickened and deeply fissured. Because this ectoparasitic variant is most commonly associated with immunocompromised hosts (patients with AIDS, leukemia, diabetes, and transplants), it is believed that Norwegian scabies represents an opportunistic process [2]. Patients with neuropathy, spinal-cord injury, and skin anesthesia are also at risk. Norwegian scabies is often difficult to recognize clinically and can be mistaken for erythrodermic psoriasis, hyperkeratotic eczema, Darier’s disease, and severe contact dermatitis [1].

Skin infectious due to Staphylococcus aureus or Streptococcus pyogenes have been associated with mite infestations [1]. We describe a case of Norwegian scabies that was complicated by the development of acute endocarditis due to a toxic shock syndrome toxin-1 (TSST-1)–producing strain of S. aureus in a patient with trisomy 21. To our knowledge, this is the first report of an association between Norwegian scabies and acute endovascular infection by a TSST-1-producing strain of S. aureus.

See editorial response by Mathisen on pages 646–8.

A 31-year-old man with Down syndrome (trisomy 21) and long-standing diffuse, desquamative, hyperkeratotic skin lesions presented for evaluation of fever, weakness, gingival bleeding, diarrhea, abdominal pain, and rectal bleeding of 4 days’ duration. Twelve hours before evaluation a diffuse, hemorrhagic, petechial rash erupted. Upon initial examination, the oral temperature was 103°F; the pulse was 100 beats/min, and the systolic blood pressure was <90 mm Hg. Physical examination revealed a lethargic man with characteristic Down facies who was in extreme distress. Intense erythroderma, diffuse hemorrhagic petechiae, mucosal erythema, conjunctival hyperemia, epistaxis, gingival bleeding, and rectal bleeding were observed. The cardiac examination revealed a normal S1 and a loud S2; no murmur was evident. The lungs were clear to auscultation. An 18-cm liver was palpated, and the spleen tip was felt 5 cm below the left costal margin. No lymphadenopathy was noted. The neurological examination was remarkable for a depressed sensorium, nonfocal reflexes, and the absence of Kernig’s and Brudzinski’s signs.

A urine analysis revealed 4+ proteinuria, bacteruria, and hematuria. Laboratory evaluation revealed the following values: hematocrit, 19.9%; WBC count, 28,000 × 10^3/L (48% segmented forms, 47% band forms, 2% lymphocytes, 1% monocytes, and 2% myelocytes); platelet count, 19,000/mm^3; prothrombin time, 34 seconds (normal, 10–12 seconds); direct bilirubin level, 2.0 mg/dL; lactate dehydrogenase level, 692 U/L; aspartate aminotransferase (AST) level, 83 U/L; blood urea nitrogen level, 60 mg/dL; creatinine level, 4.0 mg/dL; and an anion gap of 28. Arterial blood gas determinations while he was receiving 2 L of oxygen by nasal cannula revealed the following values: pH, 7.26; PCO2, 21 mm Hg; HCO3−, 9.1 mmol/L; and PO2, 108 mm Hg. A chest radiograph demonstrated bilateral patchy pulmonary infiltrates. A neutrophil with an intracytoplasmic bacteria was seen on review of the peripheral blood smear. Despite aggressive supportive therapy and treatment with antibiotics, the patient died 12 hours after admission. Blood cultures were positive for S. aureus.

Global scabetic infestation of the skin with focal microabscesses was noted on postmortem examination. Acute endocarditis of the tricuspid, aortic, and mitral valves; septic coronary emboli; and acute vegetations on the foramen ovale were evident. Brain abscesses in the distribution of the middle cerebral artery and cerebellar hemispheres were noted, as were infected pulmonary emboli. The renal cortex displayed diffuse petechial hemorrhages, consistent with disseminated intravascular coagulation. In addition, focal acute pyelonephritis and mesangial and capillary proliferative glomerulonephritis were present. Electron microscopic examination of a kidney demonstrated abundant mesangial and subendothelial electron-dense immune deposits. Hepatic steatosis and portal triaditis as well as lymphatic and splenic hemophagocytosis were also found, consistent with toxic shock syndrome (TSS). Review of bone marrow sections did not reveal acute leukemia. Adrenal hemorrhage was not found. The S. aureus strain was shown to produce TSST-1.

We believe that this patient developed fatal staphylococcal sepsis and endocarditis as a complication of infestation by S. scabiei. Further, the S. aureus strain produced TSST-1. TSS is an acute illness characterized by fever, erythroderma, diarrhea, conjunctival hyperemia, hypotension, multiorgan dysfunction, and desquamation, and can present as a life-threatening illness associated with innocuous or inapparent infection. In this case, the break in integument was caused by the ectoparasite, S. scabiei. The production of TSST-1 by the S. aureus strain may have contributed to the fatal outcome [3].

It has been shown by Shelley et al. [4] that Staphylococcus species heavily colonize the scabetic burrows in patients with erythrodermic Norwegian scabies. This intense colonization may predispose patients with scabies to bacteremia. The portal of entry of the staphylococcal infection is almost certainly the skin furrows created by the generalized scabies. Blood cultures are usually negative in cases of TSS.

The strain of Staphylococcus recovered from our patient was shown to produce TSST-1 [5]. As a superantigen, αTSST-1 is known to be a potent stimulator of IL-1, IL-6, and TNF from human monocytes [6]. Superantigens bind directly to major histocompatibility class II molecules to form a complex that attaches to Vβ, the variable region, of the T cell receptor. This attachment stimulates T cell proliferation, cytokine production, and T cell lysis. TSST-1 can also inhibit myocardial contractility.

It is tempting to speculate that an unknown immune system dysfunction believed to be responsible for the association between Norwegian scabies and Down syndrome may have contributed to our patient’s staphylococcal sepsis, endocarditis, and death.
Clinicians should be alerted to this unusual complication of Norwegian scabies in patients at risk.

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References

Editorial Response: Of Mites and Men—Lessons in Scabies for the Infectious Diseases Clinician

The article by Bonomo et al. [1] is an important reminder that scabies, although generally considered a nuisance condition, may occasionally have the potential for real harm. The article also reminds us of the changing epidemiology and sometimes misleading clinical presentation of this still common infection. Although the scabies mite was first identified during the early 1600s, it was not until the early 1700s that the organism was linked to the pruritic skin condition that now bears its name. For infectious diseases specialists, the scabies mite is particularly significant, given that it was one of the first microorganisms actually linked to human disease—in a small way presaging the ‘‘germ theory’’ of disease. Work by Mellanby [2] during World War II, in which conscientious objectors volunteered to be infected with the scabies mite, firmly established much of what we know about the transmission of scabies and the clinical course of the infection. More recently, by use of a clever in vitro model of human infection that employs synthetic skin, biologists have begun to unravel elements of the host immune response to scabies [3]. The increasing number of immunocompromised patients has led to surprising changes in both the epidemiology and clinical presentation of this once ubiquitous infection. Practicing infectious diseases clinicians should be particularly aware of the following trends in both clinical presentation and therapy for the disease.

See article by Bonomo et al. on pages 645–6.

The changing epidemiology of scabies. Although classically a disease of poverty and overcrowding, scabies continues to be a periodic problem in modern, developed societies. Scabies epidemics appear to occur in cycles—perhaps as new immunologically naive populations become available—and current reports suggest that we are experiencing yet another cycle. In addition, scabies is increasingly recognized among immunocompromised patients including patients with underlying cancer and AIDS [4]. Nosocomial outbreaks in hospitals and nursing homes, which often go unrecognized for long periods, can represent a costly problem that may be difficult to control [5].

Atypical features may delay the diagnosis of scabies. In the case reported by Bonomo et al. [1], a patient with trisomy 21 presented with a longstanding history of diffuse, eczematous, hyperkeratotic skin lesions. Crusted (Norwegian) scabies was diagnosed only in hindsight, after the patient died. Although immunologic studies were not performed in the case of Bonomo’s patient, individuals with trisomy 21 have underlying immunologic defects that increase the risk of recurrent infections [6]. Crusted scabies is a variant of scabies seen in immunocompromised patients with underlying immunologic defects caused by cancer, diabetes, immunosuppressive therapies, and HIV infection [4]. The typical case of scabies in an immunocompetent patient is generally associated with a relatively low mite load; an average of 5–10 mites are detected on the body surface. The rash, and itch, represent an immunologic response to the burden of mites, eggs, and fecal pellets (scybala). Immunocompromised patients with crusted scabies are less able to control the infection immunologically and, consequently, develop huge mite burdens—on the order of thousands to millions!

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