Stevens-Johnson Syndrome

Save to myBoK

by Sue Prophet, RRA, CCS

The term "erythema multiforme" was first used by von Hebra in 1866 to describe a disease characterized by symmetrically distributed, pleomorphic, evolving cutaneous lesions, located primarily on the extremities, and by a proclivity for recurrence. In 1922, Stevens and Johnson described two boys with febrile illnesses associated with cutaneous lesions similar to those of erythema multiforme. They also had stomatitis and severe conjunctivitis that resulted in visual impairment. This severe form of erythema multiforme became known as Stevens-Johnson syndrome (SJS). Stevens-Johnson syndrome is also known as erythema multiforme major; the mild cutaneous form described by Hebra is called erythema multiforme minor. Toxic epidermal necrolysis (TEN), or Lyell's syndrome, is also considered to be in the erythema multiforme category. In Lyell's syndrome, the epidermis is damaged over large confluent areas of the skin surface. Both Lyell's syndrome and SJS are life-threatening disorders with an insidious onset. It is still not known whether these three syndromes represent a continuum or distinct etiopathologic entities.

Stevens-Johnson syndrome is a systemic disease involving the skin and mucous membranes, which may involve other organs as well. It is believed to be a cell-mediated hypersensitivity reaction to distinct immunologic stimuli, including infectious agents and drugs. The cutaneous lesions are those of erythema multiforme, which may be erythematous papules, vesicles, bullae, or iris lesions. Mucosal lesions develop and include conjunctivitis and oral and genital ulcers. Gastrointestinal bleeding, pneumonia, arthritis, arthralgia, fever, and myalgia can also develop.

Antibiotics, especially sulfa drugs and anti-convulsants (especially phenytoin), are the most common medications known to precipitate development of SJS. Barbiturates have also been implicated in SJS. Drug-induced SJS generally appears one to two weeks after initiation of drug therapy. Since SJS is a very serious illness, if a patient develops a skin rash along with fever, malaise, and/or conjunctivitis, and the patient is taking a medication (especially a sulfonamide or anticonvulsant), he/she should be referred to a hospital right away.

Infections commonly associated with SJS development are herpes simplex and *Mycoplasma*. Usually when herpes simplex is the precipitating factor, patients develop erythema multiforme minor rather than SJS. Erythema multiforme typically develops seven to 20 days after the appearance of a herpetic lesion. Herpes-associated erythema multiforme generally occurs in young adults. Both type 1 and type 2 herpetic infections have been associated with erythema multiforme.

The treatment of SJS depends on the suspected precipitating cause. In the case of drug-induced SJS, the drug should be discontinued. The responsible drug will be one that the patient has been receiving for one to three weeks. If the patient had a previous sensitizing exposure, the onset could be shorter. If an infection is suspected as the cause, it is important to identify the infectious agent and institute appropriate therapy. Antiviral therapy reduces the severity of SJS in patients in whom herpes simplex infection is the precipitating factor. Chronic antiviral therapy has been shown to be effective in some cases in suppressing attacks of recurrent erythema multiforme minor in patients with herpes-simplex induced disease. In patients with *Mycoplasma*-induced SJS, anti-microbial therapy is initiated.

In addition to identifying the precipitating factors and eliminating them, corticosteroids are often prescribed. Corticosteroids act to dampen cell-mediated immunity, which is responsible for the symptoms of SJS, as well as for other cell-mediated immunologic phenomena such as graft-versus-host disease and contact dermatitis. In general, the initial corticosteroid dose should be at least the equivalent of 1 mg/kg of prednisone. Once the disease is controlled, tapering of the dose should be gradual, generally over several weeks. If there is a recurrence of symptoms, the corticosteroid dose should be increased and tapering should be more gradual.

The involvement of the skin and mucous membrane generally reflects the condition of other organ involvement. Thus, if the skin and mucosal lesions are improving but the chest film or liver functions are worsening, it is important to seek alternative

explanations for these complications. If the inciting immunologic stimulus cannot be suppressed, or the dosage of corticosteroid is insufficient to suppress the immune system, SJS may progress.

Dermatologic and mucosal lesions should be evaluated, as well as the potential involvement of other organs, including the lung, liver, and kidneys. Consultations from specialists in ophthalmology, infectious disease, dermatology, or plastic surgery may be required, depending on the pattern and severity of organ involvement. Fluid, electrolyte, and nutritional status must be monitored and maintained. In severe disease, pain may need to be controlled. Since SJS is difficult to diagnose early, it is not unusual for a secondary infection or associated complication such as elevated liver functions or gastrointestinal bleeding to have begun prior to proper diagnosis and treatment.

Code Assignment

Stevens-Johnson syndrome, erythema multiforme minor, and Lyell's syndrome are all classified to code 695.1. If drug-induced, assign the appropriate E code from the "Therapeutic Use" column of the Table of Drugs and Chemicals as an additional diagnosis to identify the drug. If drug-induced erythema multiforme is due to a poisoning (i.e., overdose or wrong substance given or taken) rather than an adverse effect, assign the appropriate poisoning code for the responsible drug, code 695.1 to describe the specific manifestation, and an E code to indicate the circumstance of the poisoning. In the case of a poisoning, a "Therapeutic Use" E code would not be assigned. If the poisoning is the reason for admission or encounter, the poisoning code should be sequenced as the principal diagnosis, with code 695.1 as a secondary diagnosis code.

Reference

Grammer, Leslie C. "Stevens-Johnson Syndrome: A Rash that Affects More than Just Skin." *Medscape Respiratory Care* 1(8), 1997; http://www.medscape.com.

Sue Prophet *is AHIMA's director of classification and coding.*

Article Citation:
Prophet, Sue. "Stevens-Johnson Syndrome." *Journal of AHIMA* 69, no. 3 (1998): 67-68.

Driving the Power of Knowledge

Copyright 2022 by The American Health Information Management Association. All Rights Reserved.