

MISDIAGNOSIS OF STEVENS-JOHNSON SYNDROME

Brian Camazine, M. D.

INTRODUCTION:

Erythema multiforme (EM), Stevens-Johnson Syndrome (SJS) and toxic epidermal necrolysis (TEN) are mucocutaneous disorders forming a disease spectrum. Although the etiology is unclear, these disease processes appear to be a cell-mediated cytotoxic reaction directed against the epidermis. In the severest form, there is a large loss of epidermis as well as multisystem involvement including the respiratory and gastrointestinal tracts. Thrombocytopenia and sepsis are common.

CASE PRESENTATION:

A ten year old male was brought to the emergency room following loss of consciousness while playing at school. The patient had no recollection of what occurred. His parents related several episodes over the past two months during which he appeared to "blank out for a few seconds". His emergency room workup was negative and he was diagnosed with a partial complex seizure disorder and started on Tegretol. Three weeks later the patient returned to the emergency room with fever, malaise, dehydration, sore throat, and erythematous circular skin lesions. He was diagnosed with scarlet fever, hydrated, and started on penicillin. Forty-eight hours later, he was brought back to the emergency room by paramedics. He was critically ill with fever and dehydration. On physical examination his skin was diffusely erythematous with areas of exfoliation. His oral mucus membranes were blistered with areas of erosion. He had severe bilateral conjunctivitis. Chest xray showed bilateral pneumonia. He was diagnosed with SJS/TEN and transferred to a burn center. Over the next two days, he developed progressive sepsis, and renal and pulmonary failure requiring ventilatory support and dialysis. He expired six days later.

The family initiated a lawsuit against the emergency medicine physician for failing to get a history of Tegretol use and misdiagnosing the SJS.

DISCUSSION:

EM/SJS/TEN has been linked to many drugs, especially anticonvulsants, as well as infections and malignancies. Many cases are idiopathic. While mortality is relatively low in EM, it can be as high as 60% with TEN. The initial severity of the disease may not be recognized because early symptoms may be mild. Target skin lesions are pathognomonic, especially in the presence of mucosal lesions. When the disease is

severe, there is multisystem involvement as well as mucocutaneous disease. It is critical for the initial treating physician to make the diagnosis because the disease can progress rapidly resulting in severe sequelae and death.

CONCLUSION:

The medicolegal implications of EM/SJS/TEN are obvious. The disease can be severe with a high mortality. The initial presenting signs and symptoms may be mild leading to an incorrect diagnosis. A high index of suspicion combined with a detailed history and physical should lead to early diagnosis.

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