

Abstract

Background: This is a clinical case study that explores a collegiate volleyball athlete who was diagnosed with Stevens-Johnson Syndrome. Stevens Johnson Syndrome is a very rare and severe, skin reaction that predominantly involves mucous membranes causing skin tissue to die and detach. Stevens-Johnson (SJS) is both a physically and psychologically devastating disease. Athlete started demonstrating flu-like symptoms when she was given cold relief medicine to subside her symptoms. Two days following small rashes started forming on her feet and lips were swollen. Shortly following, those small rashes began turning into mucus-filled blisters resulting in the inability to walk. Athlete then started experiencing difficulty urinating and eye sensitivities. Athlete presented with reddish discoloration in the whites of the eyes, conjunctivitis, eyelid edema, and eye crust was forming. Athlete was taken to the local hospital where she was first diagnosed with Hand-foot-mouth disease and released with antibiotics. Symptoms did not decrease, the mucus-filled blisters began spreading to mucus membrane areas, lips became black and crusted, and vision was becoming impaired. She was then transported to another hospital where she was diagnosed with Stevens-Johnson Syndrome. Athlete is transported to a third hospital who was more equipped with a burn unit accessible to reduce her symptoms and pain. Athlete was given a feeding tube due to lesions in the mouth, making it painful to swallow. Lesions first appeared on the central trunk, palms, and soles then spread throughout her face, genital area, and proximal extremities. Oral mucosal sloughing and crusting was present as well as amnesia. Athlete received three skin biopsies, two in her right leg and one in her left upper arm. Athlete spent six days with the feeding tube and seven days total in the burn unit. Athlete returned to school and volleyball nine days following her release from the burn unit.

Differential Diagnosis: Hand-foot-mouth disease and hypersensitivity syndrome. **Treatment:** Rapid identification and withdrawal of the offending drug and transfer to a burn unit with aggressive support care were the most critical steps in management and treatment of this disease. In this case, determining which medication caused the reaction was difficult so all suspected and unnecessary medications were stopped. Fluid replacement with electrolytes was critical and was administered immediately for the individual. Athlete's wound treatment was conservative as this research based hospital had never seen a case like this so several studies were performed. Due to SJS, ulcers began forming in the athlete's eye so regular ophthalmologic care was needed to perform certain treatments on her eyes. **Uniqueness:** There is no research done on Stevens-Johnson Syndrome affecting college athletes or athletes at all. The healthcare professionals who worked with this individual still do not know what caused this abnormal reaction. But approximately 75% of SJS cases are caused by medications but can vary according to age. All cases of Steven-Johnson Syndrome are unique, case by case depending on signs and symptoms, past medical history, and skin biopsy. **Conclusion:** This case highlights the diagnosis, treatment, and management of Stevens-Johnson syndrome on 19-year-old collegiate volleyball athlete.

Introduction

Stevens-Johnson Syndrome is an extremely rare and severe drug reaction that is characterized by mucosal erosions with skin pain and detachment most commonly triggered by medications. The mucous membranes of the eyes, mouth, and genitals are commonly affected with skin detachment involving less than ten percent. Blisters form on various external and internal mucous membranes causing pain which lead to erosions and bleeding.

Purpose

The purpose of this case report was to introduce a 19 year-old Division III volleyball athlete who was diagnosed with Stevens-Johnson Syndrome with no prior medical history relevant to this disease. Even though she was out of athletics and academics for twenty days she fully returned to both with no complications following. An overview of this unique and rare disease is presented to obtain additional information and a better understanding regarding Stevens-Johnson Syndrome, from onset to return to play of a Division III volleyball player.



Etiology

Stevens-Johnson syndrome is a rare and unpredictable reaction to a medication that involves drug specific CD8+ cytotoxic lymphocytes, the Fas-Fas ligand pathway, and granule-mediated exocytosis. Although it is more common in older people and women, SJS can affect anyone with a genetic predisposition: any age, any gender, and all races. It is much more common in people who are infected with HIV, with an estimated incidence of 1/1000. Drugs are a major precipitating factor, with 50% of SJS cases. The drugs most frequently associated with SJS include several medications that are used to treat seizures; allopurinol, which is used to treat kidney stones and gout; a class of antibiotic drugs called sulfonamides; nevirapine, which is used to treat HIV infections; and a type of NSAIDs called oxicams. Researchers suspect that a combination of infections and drugs could contribute to the disease in some individuals. In many cases, no definitive trigger for an individual's SJS is discovered.

Clinical Presentation

Stevens-Johnson syndrome (SJS) will begin with a nonspecific upper respiratory tract infection, including a fever, sore throat, chills, headaches, malaise, and vomiting are often noted. The median time between initiation of medication and onset of first symptoms is between 1-10 days (Cekic 2016). Early sites of cutaneous involvement begin with small red rashes in the trunk, face, as well as palms and soles. Involvement of the buccal, genital, and ocular mucosa will also occur in the acute phase in more than 90% of patients. Within a few days of the flu-like symptoms, the skin begins to blister and peel, forming painful raw areas called erosions that resemble a severe hot-water burn. Individuals affected will have damages to their mucous membranes, including the lining of the mouth and airways which can cause trouble with swallowing and breathing. The painful blisters can also affect the urinary tract, as well as painful oral crusts and skin sloughing are present in up to 90% of all cases. Severe damages to the skin and mucous membranes makes SJS a life-threatening disease as the skin acts as a protective barrier, extensive skin damage can lead to a dangerous loss of fluids and allow infections to develop over time. Among those who survive, long-term effects of Stevens-Johnson Syndrome include pigmentation, dryness of the skin and mucous membranes, excess sweating, hair loss, and abnormal growth or loss of the fingernails and toenails.

Diagnosis

The diagnosis relies heavily on the clinical symptoms and histological features. Diagnosis is often obvious from appearance of lesions and rapid progression of symptoms. A skin biopsy, blood cultures, CBC, and glucose are all orders that should be taken for diagnostic investigations for Stevens-Johnson Syndrome.

Management

Identification and early withdrawal of the offending medication(s) if the most important action as it is associated with a better prognosis. The patient should first be transferred to the appropriate level of care which is the burn intensive care unit. In the burn unit, there is a focus on assessment and management of airway, renal function, fluid and electrolyte balance, skin and ocular surfaces, pain control, and prevention of infection. Maintenance of a proper body temperature, proper fluid-electrolyte balance, and maintenance of a strict aseptic environment is crucial. Wound care is the third aspect of management of SJS, some will utilize surgical debridement and whirlpool therapy as others will leave detached skin in place to function as a biologic dressing. Ophthalmologic consultation is also essential as good eye care with early referral prevents complications such as scarring. In the Indian Journal of Dermatology, they used banana leaves for the care of SJS patients as it reduced pain, increased comfort, and lead to early wound healing. Drug treatment for SJS is a controversial and continuously expanding but oral steroids, instituted within 24-48 hours of onset of disease and tapering over the next 7-10 days gives the best results. Dexamethasone and cyclosporine are two drugs that are recommended in the treatment of Stevens-Johnson Syndrome.

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