Rehabilitation Medicine Approach to the Treatment of Patients with Stevens-Johnson Syndrome

Abedallh Hamad MD[1,3], Frida Shemesh MD[1,3], Avi Ohry MD[1,2,3], Yekaterina Slutzky MD[1,3], Valeria Kaplan RN MA[1,3], Svetlana Kartoon MD[1,3], and Raphael Joseph Heruti MD[1,3]

[1]Department of Rehabilitation Medicine, Reuth Medical and Rehabilitation Center, Tel Aviv, Israel
[2]Department of Rehabilitation Medicine, Faculty of Medicine, Tel Aviv University, Tel Aviv, Israel
[3]Faculty of Medicine, Tel Aviv University, Tel Aviv, Israel

KEY WORDS: dermatology, integrative approach, Stevens-Johnson Syndrome (SJS), rehabilitation medicine

IMAJ 2023; 25: 235–236

Stevens-Johnson Syndrome (SJS), or toxic epidermal necrolysis, is a rare syndrome that develops after an allergic reaction to a medication [1,2]. It affects the skin and the mucocutaneous tissue. Individuals diagnosed with SJS are rarely referred to a rehabilitation medicine (RM) facility.

The annual prevalence of SJS is about one in one million. The skin is covered with blisters. Usually, it affects about 10 % of body surface area. The patients are treated usually by ophthalmologists, dermatologists, allergologists, and immunologists. When severe complications occur, plastic surgeons and intensive care physicians may also be involved. Few publications were found that linked SJS with comprehensive rehabilitation treatment [3-5].

SJS usually begins as a serious side effect of a drug, which is sometimes accompanied by secondary infections. The list of documented families of drugs that can cause SJS includes anti-gout medications, anticonvulsants, anti-psychotics, analgesics, and antibiotics. Some viruses can induce SJS such as herpes virus, human immunodeficiency virus and hepatitis A. Other background problems that can induce this syndrome include a weakened immune system, history of previous SJS, a family history of SJS, and *HLA-B*1502* gene.

The clinical syndrome starts with flu-like symptoms followed by a painful rash that causes severe blisters. This reaction is accompanied by fever, sore mouth, fatigue, and annoying eyes pain. Later, a burning and painful sensation is felt over the body. Complications include secondary skin infection, sepsis, ophthalmic inflammation, respiratory failure, permanent skin and hair changes, and new bone formation (heterotopic ossification).

We assessed the rehabilitation outcome of patients with SJS after rehabilitation. The records of three patients who presented with a severe form of SJS and were hospitalized in a RM ward were assessed and reviewed. All patients benefitted from the RM comprehensive process. RM can successfully offer an integrative-comprehensive approach to patients with severe forms of SJS.

PATIENT DESCRIPTION

PATIENT 1

A 59-year-old male with known history of diabetes, gout, postural hypotension, and atrial fibrillation was referred to a general hospital because of SJS syndrome due to the use of allopurinol. Approximately 60% of his skin was affected during his hospitalization. He was treated surgically by skin grafts. Altered liver function tests were observed. He needed mechanical ventilation due to respiratory failure from which he was weaned. He developed a cerebral stroke with right spastic hemiplegia, aphasia, and facial nerve palsy. After stabilization, he was transferred to our hospital for further treatment. On admission to our department, he was stable hemodynamically with poor endurance for any effort. Multiple skin lesions at various degrees and skin graft were noted. He needed assistance in all basic activities of daily living (ADL) and was confined to a wheelchair. Functional independence measure (FIM) score at admission was 80/126. During his hospitalization, he received a multi-disciplinary rehabilitation treatment, which included attention to his chronic medical conditions, pain control, treatment of his skin lesions, prevention of formation of new lesions, and nutritional support. The RM process also focused on physical and occupational therapy to improve his range of motion, muscle strength, and coordination as well as improving endurance of effort. After 3 months, the patient was discharged home with intact and healthy skin. Only small scars remained. He was independent in all basic activities of daily living, yet due to lower exercise endurance he needed mild assistance in ADL functions. FIM at discharge was 102/126.

PATIENT 2

A 78-year-old woman who worked in a family factory had been diagnosed with diabetes, hyperlipidemia, osteoporosis, and keratosis. She was hospitalized in a

general hospital due to the appearance of the allopurinol induced SJS syndrome in its severe form. The dermatologic complications spread all over her body, including her face. She was treated by corticosteroids, intravenous immunoglobulin, and mechanical ventilation in an intensive care unit. After weaning from ventilation, she was referred to our department. Her main problems included severe weakness, weight loss, depression, and extensive skin lesions at the level of upper and lower limbs bilaterally. She needed mild to moderate assistance in basic ADL functions and was mobilized with a wheelchair. FIM score at admission was 92/126. During her stay at our department, she continued treatment with corticosteroids (prednisone and dexamethasone) in additional to local wound therapies with ointments, dermaGran B cream, hycomycin, and polydine solution according to the type of the skin lesion. The rehabilitation treatments included physiotherapy for muscle strengthening, improvement of joint range of motion, occupational therapy to practice and improve ADLs, dietitian consultation, protein rich nourishment, and respiratory physiotherapy to improve breathing and endurance. After 2 months, the patient was discharged home with minimal small scars. She was independent in all basic daily activities, but she needed mild assistance and walked with support of a walker. FIM at discharge 112/126.

CASE 3

A 75-year-old woman with mild cognitive impairment and chronic essential hypertension with no known allergies to drugs was admitted to our ward after total hip replacement due to sub-capital fracture of the femur. She needed mild help in basic ADL and was confined to a wheelchair due to hip weakness and pain. FIM at admission 89/126. During her stay she received resprim (co-trimoxazole) as treatment for a urinary tract infection. Two days later, she developed skin erosions over her lower lip, followed by diffuse skin lesion over her left hip and upper chest. She was diagnosed with SJS secondary to resprim. Resprim was stopped and wound treatment was initiated with oral antihistamine drug (antihiston 2 mg × 3/day) as well as local dermal cream tevacutan, which was later replaced with silverol to improve the skin lesions. After 3 weeks of treatment, full recovery of all skin lesions was noted. Two months of extensive rehabilitation therapy included physiotherapy for muscle strengthening, improvement in range of motion, occupational therapy to practice and improve daily activities, dietitian consultation, and protein rich nourishment. She was discharged home independent in ADLs with need of slight assistance in dressing and bathing. She walked independently with the support of walker. FIM at discharge 102/126

COMMENT

We described our experience with the rehabilitation treatments of three patients who developed a severe form of SJS. SJS is a minor form of Lyell syndrome, which is characterized by toxic epidermal necrolysis in which epidermal detachment affects more than 30% of the body surface area. SJS usually affects less than 10% of the body surface area. It is a rare mucocutaneous disease that affects one in one million healthy subjects each year and one in 100,000 in people with a compromised immune system. Our interdisciplinary comprehensive rehabilitation team approach included careful attention to the electrolyte and fluid balance, a continuous daily careful care of the extensive skin lesions, which is similar to partial thickness burn injury, general medicine attention to the previous background diseases, physiotherapy, skilled nursing, occupational therapy, and psychosocial support. Nutritional consultation was a crucial ingredient of the treatment. The RM approach with close attention to the skin lesions care in patients with SJS can contribute to full recovery of this rare syndrome.

The rehabilitation team was also confronted with acute problems as painful sensations, mucocutaneous rash and blister formation, weakness, and fatigue. Sometimes the dermatologic lesion spreads into the mouth, thus starting parenteral feeding. Other goals of treatment were preventing respiratory complication and providing, if necessary, mechanical ventilation, preventing muscular atrophy and pressure sores due to prolonged confinement to the bed. In Israel, patients with severe dermatological problems (burns, severe psoriasis, or SJS) are rarely referred to our facilities. We believe that patients with acute severe dermatological problems, can benefit from our comprehensive RM approach.

Correspondence

Dr. A. Ohry
Dept. of Rehabilitation Medicine, Faculty of Medicine, Tel Aviv University, Tel Aviv 69978, Israel
Fax: (972-3) 638-3649
Email: aohry@tauex.tau.ac.il; aohry@hotmail.com

References

1. Stevens-Johnson Syndrome Foundation. What is SJS? [Available from http://sjsupport.org/?page_id=451]. [Accessed 15 December 2020].
2. National Organization for Rare Disorders Stevens-Johnson syndrome and toxic epidermal necroloysis. [Available from https://rarediseases.org/rare-diseases/stevens-johnson-syndrome-and-toxic-epidermal-necrolysis/]. [Accessed 15 December 2020].
3. McDonald K, Johnson B, Prasad JK, Thomson PD. Rehabilitative considerations for patients with severe Stevens-Johnson syndrome or toxic epidermal necrolysis. A case report. *J Burn Care Rehabil* 1989;10 (2):167-71.
4. Gibson CJ, Poduri KR. Heterotopic ossification as a complication of toxic epidermal necrolysis. *Arch Phys Med Rehabil* 1997; 78 (7): 774-6.
5. Samanci N, Balci N, Alpsoy E. Heterotopic ossification related to toxicepidermal necrolysis in a patient with Behçet's disease. *J Dermatol* 2005; 32 (6): 469-73.

The soul would have no rainbow had the eyes no tears.
John Vance Cheney (1848–1922), American poet, essayist and librarian