Atypical Steven Johnson syndrome secondary to Mycoplasma Pneumoniae infection in the adult population. A diagnostic dilemma

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Abstract

Stevens-Johnson syndrome (SJS) is a severe mucocutaneous reaction, most commonly triggered by medications, characterized by extensive necrosis and detachment of the epidermis. Mucous membranes are affected in over 90 percent of patients, usually at two or more distinct sites (ocular, oral, and genital).The incidence of SJS secondary to mycoplasma pneumoniae is rare, with only 1–5% of total reported cases. Furthermore, this has been mainly reported in children and young adults. Here, we present to you a rare case of mycoplasma pneumoniae induced steven johnson syndrome in a 28 year old male

Case report

A 28 year old male was brought unconscious to our casualty department. History from his roommates revealed that the patient had been complaining of fever and headache for 1 week. He did not seek any medical attention, only took over the counter medications(T. Paracetamol, Lozenges, Neodeca eye drops). Prior to his admission, his roommates last saw the patient 4 days ago, where they only noticed conjuctival redness, with no skin lesions.

Upon arrival to our casualty, his GCS was E1V1M1, lethargic, dry and was intubated for airway protection. On examination, noted his eyes had conjunctival injection bilaterally, pupil 3mm reactive. On his face noted sloughed erosion over the bilateral eyelids and around the cheek area. Dusky erythematous macular rash noted all over the facial area sparing the scalp. Oral mucosa revealed red crusted lips with minimal bleeding upon contact. His body showed dusky erythematous macular rash noted all over the trunk, bilateral upper limbs, bilateral thighs with multiple areas of sloughed skin over the central chest area and sacral area.Nikolsky sign was positive. Genital region showed denuded dermis all over the scrotal area, with minimal bleeding upon contact. SCORTEN - 2

His blood work showed multiorgan involement with transminitis (ALT: 120, AST: 336, total bilirubin 45) and non-oligouric acute kidney injury KDIGO stage 3 (Urea: 52 , Na 144 , K 3.0 , Creatinine 550). Viral screening was negative for HIV, HEP B, HEP C, HSV 1, HSV 2. Mycoplasma pneumoniae IgM titer was positive on enzyme immunoassay. Titre was 1: 1280.Blood cultures were positive for E.coli. He was promptly treated with IV hydrocortisone for 7 days before being tapered to oral prednisolone. IV meropenam

was initiated for the bacteremia and IV azithromycin for mycoplasma pneumonia coverage. He was nursed in ICU for a total of 8 days



Fig 1: Skin rash over face with extensive blistering lesions with mucosal involvement.



Fig 2: Extensive skin rash with sloughing seen over anterior chest region



Fig 3: painful, erythematous edema over soles of foot

Discussion

Mycoplasma pneumoniae is one of the commonest causes of atypical pneumonia. 25–33% of infected patients may show some cutaneous manifestations most notably exanthems, urticaria, and SJS. Infection with *M. pneumoniae* may produce a variety of extrapulmonary manifestations, including neurological, hepatic, cardiac, and dermatological diseases before, during, or after pulmonary involvement. SJS is a critical disease, characterized by high fever, extensive blistering lesions, mucosal involvement, and atypical target-like skin lesions which often progress to toxic epidermal necrolysis (TEN). According to a widely accepted classification, SJS and TEN are considered a disease continuum and are distinguished chiefly by severity, based upon the percentage of body surface affected by blisters and erosions. SJS is the less severe condition, in which skin detachment is <10 percent of the body surface. TEN involves

detachment of >30 percent of the body surface area. SJS/TEN overlap describes patients with skin detachment of 10 to 30 percent. In addition to SJS, *M. pneumoniae* has also been noted to cause erythema multiforme (EM) and “atypical SJS” manifesting as severe mucositis without skin lesions. The distinction between SJS and EM major is controversial; however mucous membrane involvement generally favors a diagnosis of SJS. SJS usually evolves as an idiosyncratic reaction to medication (typically anticonvulsants or antibiotics), but *M. pneumoniae* has been cited as the most common infectious cause.

M. pneumoniae-associated SJS most commonly affects children and young adults (mean age 15.3 years to 19 years) compared with drug associated SJS (mean age 31.4 years to 46.6 years). In younger patients, SJS may be preceded by symptoms of upper respiratory tract infection from 2 days to 2 weeks before the appearance of the rash. Most adults generally develop symptoms and rash almost simultaneously, perhaps due to an immune reaction to previous exposure. Oral lesions are present in 100% of cases, genital lesions in 75%, and ocular lesions in 66%. *M. pneumonia*-induced SJS is associated with less severe complications and less internal organ involvement than those resulting from other causes

The laboratory diagnosis of *Mycoplasma* infections can be obtained by culture, serology utilizing enzyme immunoassay (EIA), or complement fixation (CF) methods and polymerase chain reaction (PCR) based testing. Isolation of *M. pneumoniae* by culture is considered to be the gold standard of diagnosis, but requires 10–14 days and is not readable available. Serology is most frequently used, and the most widely available methods are the CF and EIA tests.EIA-based-tests that detect both IgM and IgG are more sensitive and specific than CF; however, the production of IgM antibodies may be inconsistent during the acute phase of infection, particularly in adults.

Management of patients with Stevens-Johnson syndrome usually is provided in intensive care units. No specific treatment of Stevens-Johnson syndrome is noted; therefore, most patients are treated symptomatically. Initial management includes supportive care with special attention to fluid and electrolyte balance.
Withdrawal of the suspected offending agent is critically important. Timing of withdrawal has been linked to outcome. Underlying diseases and secondary infections must be identified and treated. Supportive systemic therapy includes parenteral nutrition to replace the protein loss and to promote healing of cutaneous lesions. Intravenous insulin therapy may be required because of impaired glycoregulation. Environmental temperature raised to 30-32°C reduces caloric loss through the skin. Heat shields and infrared lamps are used to help reduce heat loss.Anticoagulation is also necessary during inpatient stay. Because of the association between Stevens-Johnson syndrome

and sulfonamides, avoid the use of silver sulfadiazine.0.5% silver nitrate or 0.05% chlorhexidine is recommended to paint and bathe the affected skin areas. Several skin care approaches have been described.Unlike burn guidelines, which recommend surgical or high-velocity saline debridement of detached epidermis, the dermatologic SDH expert panel favors a conservative approach to preserve detached epidermis as a biologic dressing.Biologic dressings may include the porcine cutaneous xenografts, cryopreserved cutaneous allografts,amnion or collagen-based skin substitutes. with variably successful results. Antishear strategies, such as limiting dressing changes, using an air-fluidized bed, and selecting nonadherent dressings, are recommended. Application of an emollient, such as petrolatum jelly, to the skin enhances barrier function, reduces transcutaneous water loss, and encourages re-epithelialization. Several therapeutic modalities have been advocated for the treatment of Stevens-Johnson syndrome including plasmapheresis, immunosuppressive therapy, and intravenous immunoglobulin (IVIG). In view prolonged treatment with systemic steroids has been associated with an increased prevalence of complications, the currently advocated approach is short-term (4-7 days), high-dose intravenous corticosteroids. There are a few ophthalmology literature which advocate systemic and topical steroids to minimize ocular morbidity. IVIG has usually been reserved for more severe cases, where encouraging results were reported when IVIG was used in high doses very early in the course of the disease and for a short period

Conclusion

The diagnosis of SJS is a challenging one, with a myriad of causes leading to this deadly syndrome. A thorough clinical examination with a detailed history is paramount in determining the causative etiology. The diagnosis of mycoplasma pneumoniae should always be in our differential as it tends to involve extrapulmonary organs, leading to an atypical presentation, such as in this case. More research and data with a multidisciplinary approach involving dermatology, ophthalmology, infectious disease, oral maxillofacial, nephrology, gastroenterology is needed to help improve awareness of condition and patient’s outcome

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