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Steven Johnsons Syndrome Induced by Carbamezapene

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Abstract: Stevens – Johnson syndrome (SJS) is characterized by rapidly spreading blisters which occur in the form of macules or target lesions which has clinical features which may be potentially fatal. It is usually associated with drugs such as anticonvulsants including carbamazepine, lamotarigine, phenobarbotal, phenytoin and valproic acid.

This is a rare case report in which we describe a case of Steven Johnsons Syndrome induced by Carbamezapene that was prescribed to treat trigeminal neuralgia affecting the oro-facial region.

Keywords:- *Carbamazepine, drug hypersensitivity reaction, Steven- Johnson syndrome, trigeminal neuralgia*

I. INTRODUCTION

Undesirable or unwanted effects in the body due to the administration of drugs are called as the adverse effects of the drug. The WHO- suggested definition of adverse drug reactions are as- Any response that is noxious, unintended and which occurs at doses normally used in humans for prophylaxis, diagnosis or therapy of disease, or for modification of physiological function. (WHO)

Carbamazepine, is an anticonvulsant medication used importantly in the treatment of epilepsy and neuropathic pain. It is a drug of choice in the treatment of trigeminal and other neuralgias. It is also used in the treatment of acute mania and bipolar disorder. The common adverse effects of carbamezapenesincludes sedation, fatigue, vertigo, ataxia, diplopia, blurred vision, nausea, vomiting and confusion. Hypersensitivity reactions are the skin rashes, eosinophilia, lymphadenopathy and hepatitis. Rarely it may cause bone marrow depression with neutropenia, aplastic anaemia and agranulocytosis. On chronic therapy, it may cause water retension due to release of antidiuretic hormone. SJS is a rare disorder which may be induced due to carbamezapenes.

Stevens – Johnson syndrome (SJS) is characterized by rapidly spreading blisters which occur in the form of macules or target lesions which has clinical features which may be potentially fatal[1].SJS is usually associated with some types of anticonvulsants, including carbamazepine, lamotarigine, phenobarbotal, phenytoin and valproic acid[2]. Clinically, the above diseases present as erythema, necrosis, and extensive sloughing of the epidermis with or without mucous involvement and systemic symptoms.

The frequency of carbamazepine hypersensitivity reaction is between 1/1,000 and 1/10,000 new exposures to the drug as reported[3].

The purpose of this article is to report a case of SJS secondary to carbamazepine in a patient with trigeminal neuralgia. We would like to discuss a case report of Steven Johnson syndrome induced by carbamezapene in a 34year old male patient with trigeminal where we will be discussing about the findings and extensive management of the particular case.

II. CASE REPORT

A 34year old male patient reported to Department of Oral Medicine and Radiology with chief complaint of multiple erosive lesions on the lips and cheeks since 2 dayswhich was associated with pain and burning sensation. Patient complained of difficulty in having food, body pain and joint pain with associated fever. History of discharge from right and left corner of the eyes, history of breathlessness and weakness.

He was on medication for trigeminal neuralgia since 2 days **(TAB TEGRITAL)** after which he had these following symptoms.

On extra oral examination, upper and lower lips were edematous and swollen. Swelling was present in the upper and lower eyelids of right and left eyes. Yellowish discharge was present from the corner of the eyes. On intraoral examination Multiple ulcers were noted in the buccal mucosa, tongue, palate, oropharynx and soft palate, labial mucosa which was oval in shape, covered with yellowish slough and surrounded by erythematous area and irregular margins. Similar lesions were present in the genitals.

Based on the clinical findings and the history the diagnosis was Steven Johnsons Syndrome secondary to Carbamazepine.

III. DISCUSSION

An adverse drug reaction (ADR) can be defined as ‘an appreciably harmful or unpleasant reaction resulting from an intervention related to the use of a medicinal product; adverse effects usually predict hazard from future administration and warrant prevention, or specific treatment, or alteration of the dosage regimen, or withdrawal of the product[4].

The drugs that are associated with Steven- Johnson syndrome are a group of anticonvusants like Lamotrigine, carbamazapenes, phenytoin in doses more than 100mg/day. Antibiotics such as penicillins, cephalosporins, paracetamol, Nonsteroidal anti-inflammatory drugs, oxycamtype mainly.

Trigeminal neuralgia involves acute, lancinating pain, triggered by non-nociceptive stimuli, in the distribution of one or more divisions of the trigeminal nerve. The 1st line of medications used to treat the painful symptoms of trigeminal neuralgia are anticonvulsants which involves carbamazepine[5].

Carbamazepine has been strongly associated with SJS. Although SJS has multiple etiologies, it is commonly triggered by viral infections (herpes simplex virus is the infectious agent more commonly involved) and neoplasias. However, the most common cause is the use of medications. Among the drugs implicated more often are allopurinol, antibiotics, anticonvulsants, and non-steroid anti-inflammatories.

Recently, in a seven-year study, Devi et al. concluded that anticonvulsants were the cause implicated most in SJS especially in the first eight weeks of treatment, and the main drug responsible (more than 80%) was carbamazepine[7].

Pathogenesis is non- specific.Dysregulation of the immunologic reaction is thought to be one of the most important causes. The death of keratinocytes due to apoptosis is currently thought to be the major mechanism[8]. SJS can be characterized as a hypersensitivity syndrome because of the preexistence of pharmacogenetic and immunologic abnormalities to the administered drug. It is difficult to prevent SJS be- cause drug hypersensitivity reactions occur in an un- predictable manner. Carbamazepine, which is widely used to treat seizure disorder, bipolar disorder, trigeminal neuralgia, and chronic pain, is one of most common causes of drug hypersensitivity reactions

The initial symptoms include fever, myalgia and weakness for 1-3 days before the development of lesions. The lesions then spread at a rapid rate involving the face and upper trunk areas within 4 days or within hours.The initial skin lesions are usually poorly defined macules with darker purpuric centers that coalesce[9]. In the present case the initial presentation of the lesions occurred after 2 days after the ingestion of carbamezapene for trigeminal neuralgia.

Diagnosis is clinical. However, skin biopsy helps confirm the diagnosis, usually excluding bullous diseases not related to drug therapy. At the early stages there is full-thickness epidermal necrosis and detachment, with an only slightly altered underlying dermis. Immunofluorescence studies only help exclude other bullous disease.

The treatment includes early recognition of the condition, cessation of suspected drugs if any, supportive therapy, referral if required, initiation of specific therapy, prevent complications and future episodes.

In the present case, the patient was admitted and isolated. A regimen of 4mg corticosteroid was given intravenously twice daily, IV fluids with antibiotics, anelgesics and antihistamines were administered.Tab Rebagen to treat ulcers, Momate- cream- for the lesions and crustations on the lips, Lexanox Ointment to treat Oral lesions,Refereshliquigel eye drops and Moxigram eye ointment to treat swelling in the eyes and discharge, Liquid paraffin oil for back and palm lesions. Vital signs were monitered every hour and a heated environment was provided.

The patient came for a follow up appointment after 2 months where all the symptoms were noticeably regressed.

The main action in SJS is early recognition of the drug reaction and withdrawal of the drug, since any delay can be seriously deleterious to the patient. There is definitive treatment for SJS other than supportive care. Certain retrospective studies have suggested that intravenous immunoglobulin may be effective in stopping the progression of SJS, other studies showed limited benefit on the mortality rate or progression of the disease[6].

IV. CONCLUSION

Clinicians have be aware of the adverse reactions of drugs and it should be administered carefully. Carbamazapenere administration should be avoided in the patients with a previous history of Steven Johnson Syndrome.

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FIGURES

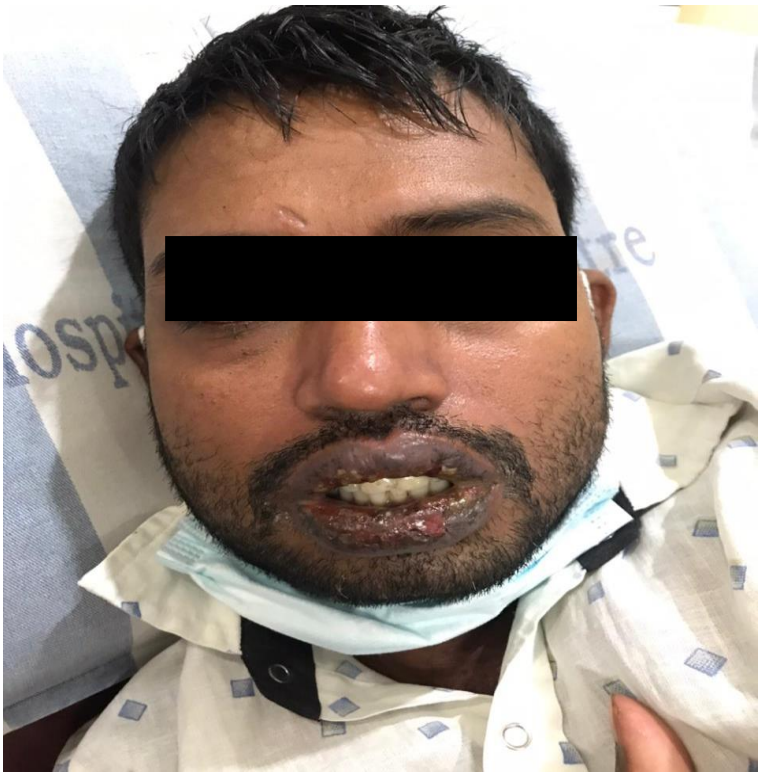

Fig. 1: Extra oral image

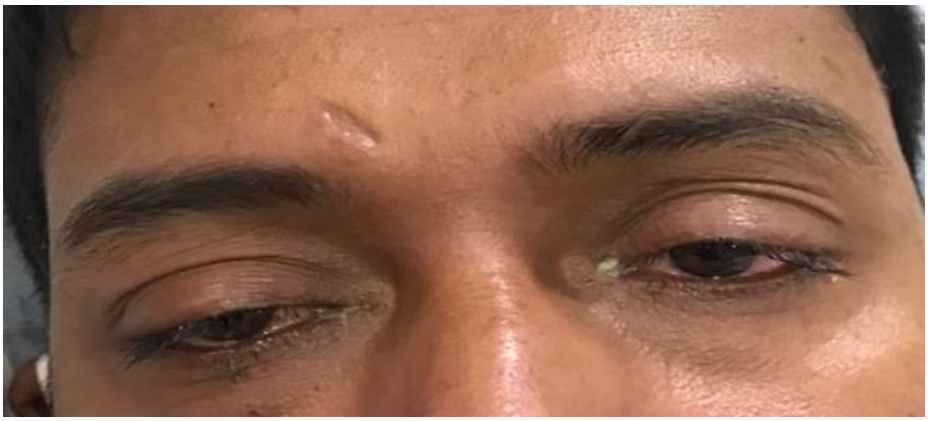

Fig. 2: Edematous right and left eye with yellowish discharge

Fig. 3: Crustations in the upper and lower lip with swelling

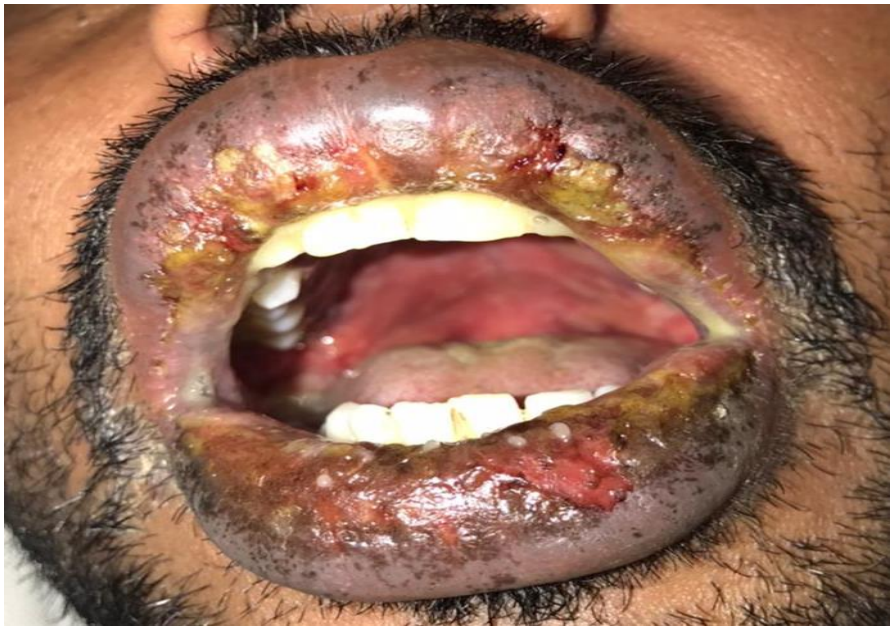

Fig. 4: Intra Oral Image

Fig. 5: Follow up picture- 2 months