Toxic Epidermal Necrolysis. A case report

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We present an extreme case of Toxic Epidermal Necrolysis, which corresponds to a severe form of Stevens Johnson Syndrome. This is a potentially fatal immune reaction that affects skin and mucosa, producing blisters and sloughing of the epithelium. Severe sequelae, including blindness, hearing loss, tooth malformation and esophageal destruction are seen in this case.

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INTRODUCTION

Toxic Epidermal Necrolysis (TEN) is a dermatologic painful, life-threatening problem, a severe form of Stevens Johnson Syndrome, (SJS)[1] which was believed to be part of erythema multiforme.

Bastuji,[2] proposed that erythema multiforme and TEN/SJS constitute totally different entities and this approach has been accepted for the medical community.

Stevens Johnson syndrome overlaps with toxic epidermal necrolysis. TEN/SJS are potentially fatal immune reactions that affect skin and mucosa, producing blisters and sloughing of the epithelium. SJS is less severe than TEN with the skin sloughing limited to less than 10% of the body.

The TEN/SJS etiology may be infectious or a reaction to some medications, which produce antibodies that are able to injure the epithelial cells.[3] The medications commonly associated with TEN/SJS are anticonvulsants, sulfas, non steroidal anti-inflammatory drugs[4] (NSAIDs) like ibuprofen and some antibiotics. The reaction usually develops within the first week of the medication. When infection is the cause, it is bacterial or viral.

Prodromal symptoms resemble a severe cold or flu. Then, a skin rash, with mucosal involvement, characterized by target lesions, blisters or erythematous spots appears. The lesions are Nikolsky positive. The blisters leave red, oozing and painful areas. There may be loss of nails or failure to grow. Typical ocular complications are conjunctivitis, iritis, infections, corneal perforation and blindness. Other organs, such as, liver, joints, kidneys and lung may be affected and leave sequelae.

The patients should be admitted to the burn unit or intensive care unit. The first line of treatment is immediate withdrawal of the medication. Then, intravenous immunoglobulin, nutritional and fluid replacement, pain and fever therapy, skin and mucosa care and regular assessment for infection should follow. Death may occur due to dehydration, infection, shock and thromboembolism.[5]

CASE REPORT

A 12 year old female was seen at the dental pediatric clinic at Nova Southeastern University. Her parents requested complete dentures for their daughter. Medical history revealed vaginal burning sensation at the age of 4. At that time, she was diagnosed with Kawasaki's disease and was treated with NSAIDs. After 12 hours of treatment, numerous skin and mucosal blisters appeared. The patient was admitted to the ICU, where her lungs collapsed and 40% of her skin was lost. A diagnosis of Stevens Johnson Syndrome was then established and a coma was induced. After 48 days the patient waked up with severe corneal damage, blindness and hearing loss. Esophageal lesions prevented her from eating. A feeding tube was placed but after 2 years the esophagus was repaired with transplantation of colon tissue.

Eight years later, upon extraoral examination at the dental clinic, there was identification of severe skin damage and pigmentation, lip vertical fissures, corneal damage and blindness, (Figure 1) dryness of skin and lips, and dysplastic nails. (Figure 2) Intraoral exam showed dry mouth, atrophy of tongue papillae, (Figure 3) missing teeth, enamel hypoplasia and dental cavities.

Panoramic radiograph disclosed incomplete dentition,

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Figure 1. Facial features, blindness, lip fissures, skin pigmentation

Figure 2. Dysplastic nails

Figure 3. Atrophy of tongue papillae

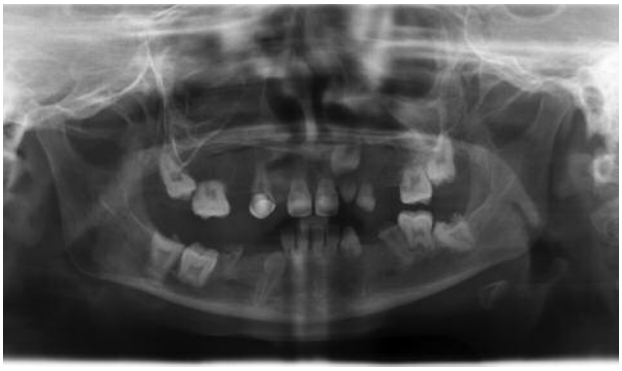

Figure 4. Panoramic radiography: incomplete dentition, short roots, impacted permanent teeth and over-retained primary teeth

extremely short roots, impacted teeth and retained primary teeth. (Figure 4)

The decision of a conservative treatment was made. After hygiene instructions and caries removal, the patient is sent for consultation to the departments of surgery, orthodontics and prostodontics in order to provide the best possible treatment with a multi-approach team effort.

CONCLUSION

Toxic Epidermal Necrolysis is a potentially fatal immune reaction that affects skin and mucosa. It is caused by infections or medications that are able to produce a reaction with formation of antibodies, which injure the epithelial cells. The initial symptoms mimic an upper respiratory tract infection; but then the skin and mucosas such as the mouth, eye and vagina, develop blisters and sloughing of the epidermis. Almost every organ of the body can have permanent damage.

Pediatric patients that manifest fever and rash while being treated with antibiotics, NSAIDS and anticonvulsants should be monitored closely in order to prevent as much as possible the extreme morbidity of this condition. The earlier the diagnosis, the less severe the sequelae.

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