

SHORT ROOTED PREMOLARS AND STEVENS-JOHNSON SYNDROME

*RACHIT WALIA, ** ARVIND GARG, *** SARANG SHARMA

¹Department of Conservative Dentistry & Endodontics, ²Department of Community Dentistry, ³Department of Conservative Dentistry & Endodontics. Krishna Dental College and Hospital, Ghaziabad, U.P. India.
Email: drrachitwalia@rediffmail.com, docarvindgarg@gmail.com, sarang74in@yahoo.com

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ABSTRACT

Stevens Johnson Syndrome (SJS) is a rare condition, with a reported incidence of around one case per million people per year. Stevens Johnson Syndrome patients suffer from inflammation of the skin and mucous membranes involving eyes, digestive system, lungs and respiratory system. Many organs can become significantly affected during the disease process but rarely dental structures. This report presents a case with history of Stevens Johnson Syndrome and associated various dental anomalies.

Keywords: Stevens Johnson Syndrome; Dental anomalies; Inflammation

INTRODUCTION

First described in 1922¹, Stevens-Johnson syndrome (SJS) is an immune-complex-mediated hypersensitivity caused by many drugs^{2, 3, 4, 5}, viral infections, and malignancies. Patients suffer from inflammation of the skin and mucous membranes and might have an impact on dental structures leading to dental anomalies.

CASE REPORT

An 18 yr old male patient reported to the Department of Conservative Dentistry with complaint of pain and sensitivity to cold in first, second and fourth quadrant. On history taking, it was found that patient had an unknown drug allergy 10 yrs back.

Oral examination revealed: (Figs 1-4)

Carious, hypo plastic teeth: 14, 15, 17, 24, 25, 44, 45, 46**

Missing teeth: 13, 32, 33, 34, 37

Retained deciduous: 72, 73, 74

Partially erupting: 35

Carious exposed: 27 (Tender on percussion)

** FDI (Federation Dentaire Internationale) system is used for tooth numbering



Fig. 1:



Fig. 2:



Fig. 3:



Fig. 4:

Figures 1-4 show carious, hypo plastic 14,15,17, 24,25,44,45,46; missing 13,32, 33, 34,37; retained deciduous 72,73,74; partially erupting 35 and carious exposed 27.

General examination revealed :(Figs 5-9)

Blepharitis

Partial blindness in right eye

Mottled pigmentation, rashes all over the body

Malformed nails

** Physical and Mental growth were at par with his age.



Fig. 5:



Fig. 6:



Fig. 7:



Fig. 8:



Fig. 9: Figures 5-9 show blepharitis, partial blindness in right eye, mottled pigmentation, rashes all over the body and malformed nails.

Radiographic Observation in OPG: (Fig 10)

Short rooted premolars

Impacted: 13, 18, 32, 33, 37

Normal bone growth and morphology.



Fig. 10: Shows short rooted premolars, impacted: 13, 18, 32, 33, 37 with normal bone growth and morphology

Investigations

- Routine blood and urine examination.
- Father's and sibling's radiographs were taken to exclude hereditary pattern.
- On consultation with other specialties (Departments of Ophthalmology, General Medicine, Dermatology, Oral pathology) and examining his old case papers it was confirmed that patient had an episode of Stevens Johnson syndrome due to unknown drug allergy.

But correlation with dental anomalies could neither be established nor could they be grouped into any other syndrome.

Treatment Plan

1. Conservative composite restorations were done in all carious teeth.
2. Endodontic treatment was done in maxillary left second molar. (Fig.11)
3. Keratoplasty was advised to improve vision in right eye.
4. Monthly follow up.



Fig. 11: shows completed root canal treatment in maxillary left second molar

DISCUSSION

SJS technically is an immune-complex-mediated hypersensitivity (allergic) condition. A severe expression of the condition is known as Erythema Multiforme and its lesser expression known as Toxic

Epidermal Necrolysis (TEN). SJS is a serious disorder with potential for severe morbidity and in some cases, can be fatal.

Erythema Multiforme is of two types: minor and major. Erythema Multiforme minor is less severe of the two types and accounts for 80% of Erythema Multiforme. The rashes⁶ appear over a few days and may be associated with minor burning or itch. They are more intense over the back of the hands and feet. The rashes last for 1 to 2 weeks and then recede leaving residual brown pigmentation⁷.

The hallmark of Erythema Multiforme major or SJS is the development of large blisters in the mouth, on the skin, around the anus or genitals, in the throat or even on the eyes⁸. In addition, affected persons may develop reddish skin rashes in various shapes and sizes, joint pains, fever and itching. SJS has a high significant mortality rate⁹.

Pathologically, cell death results causing separation of the epidermis from the dermis. The death receptor, Fas, and its ligand, FasL, have been linked to the process. Some have also linked inflammatory cytokines to the pathogenesis.

A Caucasian predominance has been reported. The male-to-female ratio is 2:1. Most patients are in the second to fourth decade of their lives; however, cases have been reported in children as young as 3 months.

Stevens Johnson Syndrome is classically a clinical diagnosis, thought to be due to a hypersensitivity complex affecting the skin and mucous-membranes. Current theories indicating immunological causes, suggest a route involving CD8+ cytotoxic T-cells triggering keratinocyte apoptosis, but the current understanding is far from complete.

The 4 etiologic categories are (1) infectious, (2) drug-induced^{2, 3, 4, 5}, (3) malignancy-related, and (4) idiopathic.

Viral diseases that have been reported include herpes simplex virus (HSV), AIDS, coxsackie viral infections, influenza, hepatitis, mumps, mycoplasmal infection, lymphogranuloma venereum (LGV), rickettsial infections, and variola.

Malaria and trichomoniasis have been reported as protozoal causes. In children, Epstein-Barr virus and enteroviruses have been identified. Drug etiologies include penicillin and sulfa antibiotics. Anticonvulsants including phenytoin, carbamazepine, valproic acid, lamotrigine, and barbiturates have also been implicated. In late

2002, the US Food and Drug Administration (FDA) and the manufacturer Pharmacia noted that SJS had been reported in patients taking the cyclooxygenase-2 (COX-2) inhibitor valdecoxib. Various carcinomas and lymphomas have been associated. SJS is idiopathic in 25-50% of cases.

Few cases have been reported in the literature where patients having history of Stevens Johnson Syndrome had short rooted premolars^{10, 11, 12, 13}. As in this case, all premolars had short roots which could be attributed to affliction of the disease at 8-9 years but surprisingly roots of canine were not affected, rather root of impacted canine was also fully formed (Fig: 10) which develops at the same time as premolar does. Radiograph suggests that impact of SJS was more on lower left teeth as more teeth were retained, impacted and incompletely formed on this side but this could not be said true for general symptoms as loss of vision was more pronounced in the right eye. The cause of these variations couldn't be established but correlation between the age of the patient and the time of disease occurrence is suggested as far as dental structures are concerned.

Treatment plan was made to restore all the cavities conservatively with composites followed by regular monthly check up. In relation to maxillary left 2nd molar, endodontic therapy was done (Fig 11). All impacted, partially erupted and retained deciduous teeth were kept under observation.

CONCLUSION

It may be concluded from the typical differences in length between the roots of the erupted, retained and impacted teeth that this condition may be due to complete cessation of the growth of the teeth at the age of 9 or 10 years. Since the patient suffered a fulminant attack of Erythema Multiforme (Stevens-Johnson syndrome) at this age, and since no other possible explanation of the short roots has been found, it is concluded that this clinical condition may have been the reason for the short root anomaly found. Damage or even destruction of the epithelial root sheath during the disease may be assumed to be the direct cause of the failure of full root development.

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