

CASE REPORT

Diagnostic and Treatment Approach in the Management of Dental Anomalies Associated with Stevens–Johnson Syndrome: A Case Report

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ABSTRACT

Background: Stevens–Johnson syndrome is a rare medical condition with severe mucocutaneous lesions due to adverse drug reactions characterized by exudative multiform erythema, stomatitis, and conjunctivitis. Long-term oral consequences of such cases include xerostomia, caries, impactions, and multiple dental developmental aberrations as short root anomalies.

Aim and objective: To highlight the role of pedodontist in early diagnosis and treatment planning of dental abnormalities due to Stevens–Johnson syndrome (SJS) using a cone-beam computed tomography (CBCT).

Case description: A 16-year-old male reported a chief complaint of decayed posterior teeth. Past medical history revealed adverse reactions to an unknown drug at the age of 4 to 5 years. In addition to carious teeth, clinical examination revealed that all canines were missing along with mandibular incisors. On CBCT examination, abnormal short, plump roots with normal crown were seen in all permanent first molars and incisors along with impacted canines and mandibular incisors. This condition was diagnosed as a “Short root anomaly” (SRA) due to SJS. He was found positive to allergy tests for NSAIDs such as ibuprofen and paracetamol.

Conclusion: NSAIDs can cause a severe adverse reaction resulting in SJS. If this hypersensitivity reaction occurs early during the development of a permanent tooth it may cause dental anomalies such as short roots, root dysmorphia, agenesis, and multiple impacted teeth.

Clinical significance: This report highlights a unique case of multiple dental aberrations due to SJS and the role of a pedodontist in the early diagnosis and treatment planning of such cases with the help of CBCT. Short root anomalies can be misdiagnosed as root resorption or immature apex. Medical history, clinical and CBCT findings are essential for diagnosis and treatment in SJS patients. Careful orthodontic treatment planning is required in cases of short root anomalies.

Keywords: Cone-beam computed tomography, Differential diagnosis, Impaction, Short root anomaly, Stevens–Johnson syndrome, TAD.

International Journal of Clinical Pediatric Dentistry (2021): 10.5005/jp-journals-10005-1986

BACKGROUND

The term short root anomaly (SRA) was coined by Lind in 1972 to describe symmetrically involved central incisors mainly with short, plump roots and blunt apices resulting in crown-to-root ratios of 1:1. Short root anomaly of central incisors is also often associated with root resorption of lateral incisors and impacted canines.¹

Stevens–Johnson syndrome (SJS) is a rare and severe acute adverse reaction to certain drugs affecting the skin and mucous membrane. Long-term sequelae of SJS include dental abnormalities such as tooth agenesis, root dysmorphia with short roots.² This report presents a rare finding of dental aberrations, SRA along with multiple impacted teeth due to developmental disturbances secondary to SJS. However, the literature lacks information on the true picture of dental features of SJS using advanced 3D imaging. A thorough evaluation of such conditions is warranted with cone-beam computed tomography (CBCT), as such conditions may present with a diagnostic and treatment challenge and often require a multidisciplinary approach.

CASE DESCRIPTION

A 16-year-old adolescent male was referred to the outpatient department, specialty clinic of conservative dentistry and endodontics, with the chief complaint of multiple carious teeth in the upper and lower jaw. The patient past medical history revealed

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How to cite this article: Katyal S, Yadav V. Diagnostic and Treatment Approach in the Management of Dental Anomalies Associated with Stevens–Johnson Syndrome: A Case Report. *Int J Clin Pediatr Dent* 2021;14(4):569–574.

Source of support: Nil

Conflict of interest: None

that he had undergone eye surgery for a corneal transplant at 14 years of age. His past medical records revealed that he had an episode of a severe allergy to the unknown drug at the age of 4 to 5 years. As a result of drug reaction, he developed erythema, desquamation of skin, and painful oral ulcers. He got admitted into a private hospital where treatment was rendered for 1 month, though he was not aware of the details of the treatment given.

On obtaining past dental history, he revealed that extractions and restorations were done in few teeth in the pediatric dentistry department 2 years ago. An OPG obtained during the pediatric dentistry consultation suggested multiple retained deciduous teeth and multiple impacted permanent teeth (Fig. 1).



Fig. 1: A 2-year-old orthopantomogram shows short root in all permanent first molars and incisors with retained deciduous incisor and multiple impacted permanent teeth

The patient's height (180 cm), weight (65 kg), BMI (20.1), and intelligent quotient were within the normal range. Extraoral clinical examination revealed scars, mottled pigmentation on the skin, and bilateral mild marginal keratinization of eyelids (Fig. 2A). Malformation of nails such as pterygium and longitudinal ridges formation was also observed (Fig. 2B). On intraoral examination, dryness of oral mucosa due to hyposalivation was evident. All teeth were hypoplastic. Advanced dentinal carious lesions were noticed on all the erupted second permanent molars except mandibular right second molar which was impacted (tooth number 47). Multiple teeth were missing clinically includes all four permanent canines and mandibular incisors. All other permanent teeth were erupted with retained deciduous teeth. The tooth-colored restoration was also present in few teeth (Figs 2C to E). Oral hygiene was satisfactory with the absence of any periodontal pocket.

Cone-beam computed tomography was advised to evaluate the status of clinically missing teeth. A full FOV CBCT scan (16 × 8 cm FOV, 0.3 mm voxel size, I CAT™ USA) was performed to derive multiple X-ray views required for orthodontic diagnosis purposes. Cone-beam computed tomography-derived lateral cephalogram revealed class III skeletal pattern due to hypoplastic maxilla, prognathic mandible with decreased lower anterior facial height (Fig. 3). The dental pattern was class I. A detailed study on



Figs 2A to E: Extraoral and intraoral photographs: (A) Mottled pigmentation on the face and bilateral keratitis, symblepharon; (B) Malformed nails with pterygium and longitudinal ridge formation in upper limbs; (C) Intraoral labial view; (D) Maxillary occlusal view; (E) Mandibular occlusal view



Fig. 3: Lateral cephalogram obtained from CBCT suggesting class III skeletal pattern

the crown and root morphology was done using the multiplanar reconstructed (MPR) slices of CBCT using Carestream 3D software (Carestream Health Inc.) on an HP 21" LED monitor with a resolution of 1,280 × 1,024 under ambient light conditions. The CBCT scan was evaluated in coronal, sagittal, and transverse planes using oblique slicing mode by a single operator (VY). The cemento enamel junction was taken as a reference line to calculate root to crown ratio for all the teeth as recommended by Choi et al. (Fig. 4).³ Short roots with blunt apex and noticeable plump were observed with respect to all permanent first molars and maxillary incisors. The root to crown ratio for these teeth was found to be <1:1. The remaining teeth had normal crown root ratio with a normal trabecular bone in periapical area. The complete description of crown and root length is given in Table 1.

Thus, CBCT images revealed the presence of a normal crown with abnormally short roots with a characteristic onion-like appearance (tooth # 16, 12, 11, 21, 22, 26, 36, 46) molars lateral central early transitional phase, though they were completely erupted (Figs 5A to C). All the teeth which were missing on clinical examination were found to be impacted on radiographic examination. All four impacted canines were impacted with mesial angulation toward midline with normal crown and root formation (Fig. 5C). Whereas, impacted mandibular incisors (tooth no. # 23, 24, 25, 26) had normal crown but no root development (Fig. 5D). Right mandibular primary canine was retained with resorbed root. Among third molars, only tooth # 38 was present.

Even though short roots were present with respect to all permanent first molars and maxillary incisors, still they responded positively on vitality testing with EPT and cold testing. Among the carious teeth # 16, 26 was non-responsive to vitality tests, whereas 37 was vital (Figs 5E and F).

Other investigations such as routine blood examination were normal. In the allergy screening test, the patient was found to be allergic to ibuprofen and paracetamol (Fig. 6). On consultation with other departments such as ophthalmology, dermatology, and oral pathology, the present case was diagnosed as an SRA secondary to SJS. Such cases with dental abnormalities secondary to systemic allergic response are first contacted by a pediatric dentist, whose precise diagnosis and interceptive and corrective treatment plan which includes a multidisciplinary approach, involving endodontics, oral surgery, and orthodontics can provide a comprehensive layout

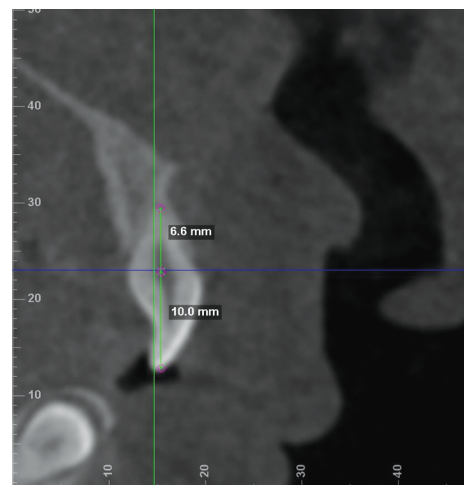
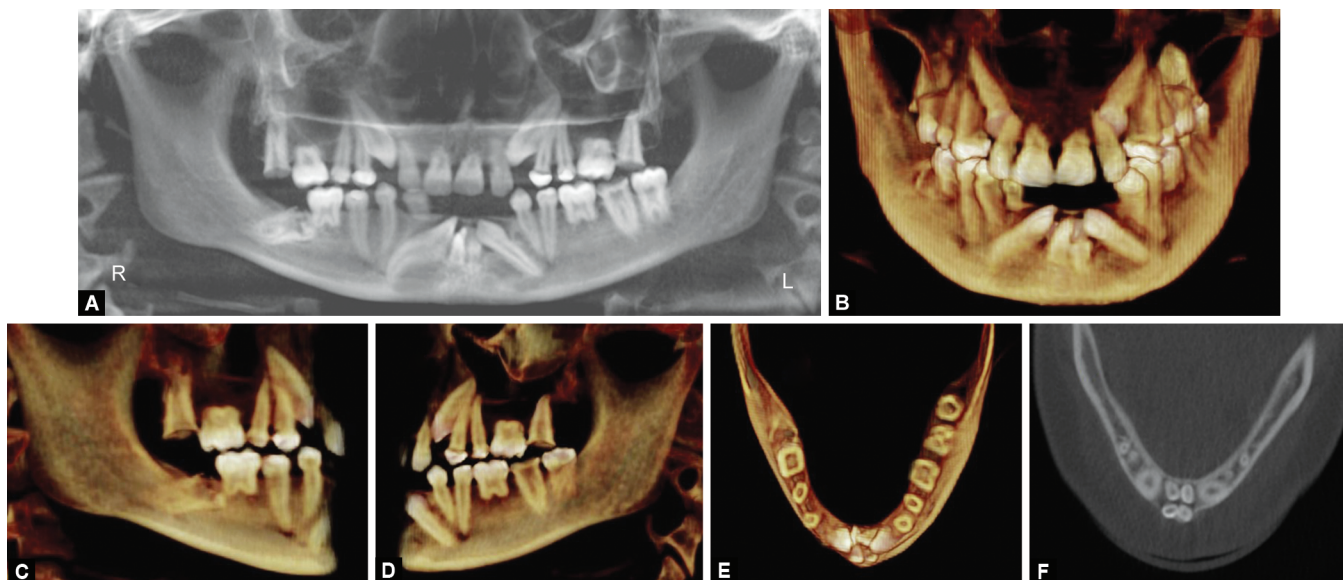


Fig. 4: Measurement of root and crown length, line joining cemento enamel junction is taken a reference line. Distance from cusp tip and root apex to the reference line is considered as crown and root length, respectively

Table 1: Linear measurements of root length, crown length, tooth length, root-to-crown ratio, and apical root morphology of teeth seen on CBCT scan

Tooth no.	Root-to-crown ratio determination		
	Root length (mm)	Crown length (mm)	Root-to-crown ratio
17	15.6	2.4 (carious)	–
16*	5.2	6.5	0.8*
15	15.9	5.4	2.9
14	14.5	5.6	2.5
13	16	10	1.6
12*	7.8	9.2	0.84*
11*	6.6	10.0	0.66*
21*	6.4	10.3	0.62*
22*	8.7	9.6	0.90*
23	15.9	10.6	1.5
24	16.4	6.0	2.7
25	14.5	6.1	2.37
26*	5.5	6.2	0.89*
27	15.2	3.4 (carious)	–
38	10.6	5.6	1.89
37	14.8	4.3 (carious)	–
36*	5.3	6.7	0.79*
35	16.6	6.2	2.68
34	14.7	6.8	2.16
33	15.2	9.6	1.58
32*	2.5	9.4	0.2*
31*	0.0	9.2	No root formed *
41*	0.0	9.4	No root formed *
42*	0.0	10.4	No root formed *
43	14.8	9.2	1.6
44	15.9	6.3	2.5
45	15.3	6.0	2.55
46*	5.3	6.9	0.76*
47	12.8	4.5	2.8

(*) Denotes teeth with short root anomaly



Figs 5A to F: (A) OPG obtained from CBCT scans; (B) Frontal view; (C and D) Right and left lateral view; (E and F) Crowding present in impacted incompletely developed mandibular incisors

Ref. No: 99945

ALLERGY SCREENING TESTS (BY IMMUNO - EIA)

TEST	RESULT	NORMALS
IgE	192 IU/mL	0.5 – 158 IU/mL
Normal For All Allergens is		< than 0.35 IU/mL

DRUGS					
TEST	RESULT	TEST	RESULT	TEST	RESULT
AMOXYCILLIN	0.21	AMPICILLIN	0.21	ASPIRIN	0.11
BRUFEN	** 1.60	CEFUROXIME	0.23	CEPHALOSPORIN	0.12
CIPROFLOXACIN	0.12	CLOXACILLIN	0.19	DICLOFENAC	0.12
DOXYCYCLIN	0.29	ERYTHROMYCIN	0.12	METAMIZOL	0.10
NIMESULIDE	0.27	NORFLOXACIN	0.23	OFLOXACIN	0.18
OXACILLIN	0.28	PARACETAMOL	* 1.00	PENICILLIN	0.17
SULPHA	0.27	TETRACYCLINE	0.20	TINIDAZOLE	0.26
XYLOCAIN	0.09				

Comments : * Mild Allergy ** Midhigh Allergy *** High Allergy

Please note that drug testing has some limitations due to sensitivity of allergens. Hence Clinical correlations should be checked

Fig. 6: Report of allergy screening tests suggests that patient is allergic to ibuprofen and paracetamol

and direction to the patient. Hence, it becomes important for pediatric dentist to have extensive knowledge of syndromes and underlying pathology.

Written and informed consent for the publication was taken from the patient’s father.

DISCUSSION

Stevens–Johnson syndrome is a rare but serious, bullous, mucocutaneous disease. Though the etiology of SJS is idiopathic, the most common causative agent includes adverse reactions to

certain drugs such as sulfonamides, anticonvulsants, and NSAIDs.⁴ On the allergic screening test, the present case was found to be allergic to ibuprofen and paracetamol. In the present case, cutaneous pigmentation and scarring along with malformed nails (pterygium formation and longitudinal ridges) were observed. Ocular changes such as dryness of the eyes lead to a partial loss of vision. Also, bilateral keratitis of eyelids with symblepharon was noted. The oral complication such as dryness of mouth was present secondary to xerostomia resulting, thereby increasing the caries activity. On correlating with the patient’s history, it can be concluded that these long-term dental complications were due

to SJS attack secondary to adverse drug reaction of NSAIDs at an age when root formation of permanent incisors and first molars was in progress causing their stunting. Eruption of few teeth was affected as these systemic allergies affect tooth germ layers such as Hertwig’s epithelial root sheath.

The CBCT scan has the advantage of distortion-free and true size image reconstruction and visualization in three planes when compared with IOPA and OPG. Linear measurements performed on CBCT scans have been used to calculate the anatomic R/C ratio as the localization of cemento-enamel junction used as a reference point is more accurate on CBCT images. The method proposed by Choi et al. was used in the present case to measure the R/C ratio and any teeth with an R/C ratio value <1.1 were considered to have SRA. Thus, in the present case, SRA was diagnosed in all permanent first molars and incisors, as they have reduced the R/C ratio with normal surrounding tissues.³

Various physiological and pathological conditions can lead to short roots. There are several diagnostic possibilities for the tooth exhibiting a normal crown with a short root. Though over the years several publications reported reasons for teeth with SRA, true SRA was attributed to genetic reasons only. Incomplete root formation, external apical root resorption, dentin dysplasia type I, and post-trauma root hypoplasia in the deciduous predecessor are some of the differential diagnoses of SRA (Table 2).

In the present case, decreased R/C ratio was found due to short roots with bilateral involvement in incisors and molars. Clinically, patient was asymptomatic with respect to short-rooted teeth, and radiographic findings were seen incidentally. The normal trabecular pattern in the periapical area suggests a healthy periodontium with no external root resorption. Also, all short-rooted teeth in the present case have a different degree of root completion. So, the reason could be sought into some developmental disturbances during the primary and early mixed dentition period as the mesially angulated impaction of all four canines was observed with complete root formation. This type of impaction is usually seen when there is resorption of root of lateral incisor as explained by Miller’s canine guidance theory.⁵

The study of the development stage of the teeth is an established method for age estimation.⁶ The detailed analysis of CBCT images suggests that the growth disturbance must have occurred around the age of 4.5–5 years, which coincides with the patient’s history of severe illness, resulting in cessation of root development in permanent first molars and incisors. All the teeth for which root formation initiates after the age of 6 years were fully formed. Since the tooth bud of the third molar also forms around the age of 5 years, its agenesis could be attributed to developmental disturbance due to the systemic condition.

In the present case, root completion of lateral incisors was halted around Nolla’s 7th stage. Lower mandibular incisors were also impacted with tooth development halted at Nolla’s stage 6. As the permanent mandibular incisors lost eruption potential, the deciduous teeth remain retained until extracted at the age of 14 years. From the above discussion, abnormal or lack of root formation of lateral incisors appear as a potential reason for palatal impaction of all canines based on Miller’s canine guidance theory.⁵ In the SJS, necrosis of the upper dermis and the lower epidermis occurs based on severity. In mild lesions, only a few epidermal cells, whereas in severe lesions the whole epidermis gets necrosed. Enamel follicles also originate from the basal layer of the oral epithelia, thus it is quite possible that during the acute phase of

Table 2: Differential diagnosis of short root conditions reported in the literature

Conditions	Findings	Etiology
Short root anomaly	Short, plump root in incisors and premolars	Genetic
		Dentin dysplasia and dentinogenesis imperfecta
	Hypoparathyroidism	Dental developmental disturbance
		Calcium metabolic disorder (systemic disturbance)
	Thalassemia	Hematological disorder
	Long-term phenytoin therapy in epilepsy	Calcium metabolic disorder
Short roots due to radiotherapy and chemotherapy before age of 12	Environmental insult due to childhood malignancy	
Short root associated with short stature and syndrome	Short root associated with other syndrome	Down, Aarskog, Seckel (bird-headed dwarfism), Rothmund–Thomson, skeletal dysplasia, “familial otodentodysplasia”
		Stevens–Johnson syndrome
Immature apex	Localized short root	Scleroderma
		Laurence–Moon–Bardet–Biedl syndrome
External root resorption	Generalized/localized short root	Trauma
Root hypoplasia in predecessor’s tooth	The short root of deciduous predecessor tooth	Orthodontic forces/trauma, periapical lesion, cyst
		Alveolodental trauma

SJS damage to keratinocytes of Hertwig’s epithelial root sheath or enamel follicle occurs. As a result of which further elongation of the root in an apical direction could be arrested. As normal width of the pulp chamber and root canals can be observed in the present case, damage to odontoblast due to SJS is not suggested in the present case.^{7–9}

The multidisciplinary approach, in this case, would involve a team consisting of a pedodontist, endodontist, orthodontist, oral surgeon, and prosthodontist.

Pedodontist is the first specialist from the dental fraternity to come across any case of SJS at an early age. Hence, extensive knowledge of clinical presentation and dental developmental challenges can reduce the misery of syndromic patients. Prevention of caries and space maintenance can make the job of other dental specialties easier as well. Also, parents can be asked for genetic consultation before they plan another child.

The endodontist and conservative dentist should make sure to correctly diagnose and differentiate inflammatory root resorption from developmental SRA as in SJS. Caries should be treated at the earliest and conservatively as teeth in this syndrome might have dentinal dysplasia and are already compromised in morphology.

These patients are usually referred to an orthodontist for impacted teeth. The orthodontic force in teeth with short roots predisposes the teeth to a higher risk of root resorption. At times, the orthodontic movement is contraindicated for teeth with severe forms of SRA. In most cases, clinical and radiographic monitoring can be used to control root resorption during the course of orthodontic tooth movement.¹⁰ An accurate diagnosis is important to plan the treatment and biomechanics. The risk and benefits should be evaluated before treating a complex case such as canine impaction.¹¹ Only light forces should be used with prolonged time in-between activation of appliances along with sequential monitoring of root resorption clinically and radiographically. The tooth movements requiring heavy forces like bodily retraction, intrusion, and torquing of incisors should be avoided. It is also recommended to bond only a few teeth and evaluate the progression of root resorption followed by complete arch bonding if there is no adverse effect. Extrusive mechanics to the teeth affected by SRA is to be very carefully avoided. Anchorage planning is critical as first molars show compromised root length and have poor anchorage value, thereby skeletal anchorage from TADs can be considered. Segmental mechanics with Miniscrew supported cantilever spring for canine eruption can help avoid forces on the already compromised dentition.

Finally, the prosthetic rehabilitation of missing teeth can be planned along with the improvement of reduced vertical dimension.

CONCLUSION

Differential diagnosis of SRA and its possible cause ruling out trauma-induced inflammatory root resorption from developmental rhizomicry is important for correct treatment planning. Teeth affected in SJS depends on the age at which acute hypersensitive reaction happened. Bilateral inheritance pattern was evidenced in this case supporting the literature already available. A multidisciplinary approach is crucial for the effective management of a case with dental anomalies due to SJS.

CLINICAL SIGNIFICANCE

The multidisciplinary approach is crucial for effective management of cases with dental anomalies due to SJS which includes accurate diagnosis, caries prevention due to xerostomia, facilitation of eruption of impacted teeth, and occlusal improvement.

ACKNOWLEDGMENT

This case was reported at the Department of Endodontics and Conservative Dentistry, All India Institute of Medical Sciences, New Delhi, India.

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