Seckel Syndrome Manifestations and Dental Care

DR. HUDA SALEM BANDER AL RAKAF

Dental Department, Prince Sultan Military Medical City, Riyadh, Saudi Arabia

INTRODUCTION

This reports a case of two Saudi male patients with prenatal and post natal growth retardation, characteristic craniofacial dysmorphism, proportionate short stature and skeletal anomalies of Seckel Syndrome. Parents are first cousins, supporting autosomal recessive mode of inheritance.

A literature review reveals that Seckel Syndrome and anesthesia management have been reported as difficult mask ventilation, difficult airway management and difficult venous canulation. A variety of tracheal tube should be at hand as many as these children have a smaller trachea than would be suspected by age and physical size. These factors should always be considered when anesthetizing children with the Seckel Syndrome.

OBJECTIVE

The purpose of this poster was to present the orodigital facial manifestations of the Seckel Syndrome and to present our experience in providing dental care.

CASE REPORT 1

A 21-year-old male was born 700g, premature (delivered in 25 weeks) although mother's pregnancy was normal. Patient's height is only .85 m, shorter than younger brother and skin is roughly hyperkeratinized. Patient's IQ is 75-80 and does not go to school. Blood examination results was not good as shown in Table 1.

Patient's deciduous teeth erupted in 2 years and started to exfoliate early in 5 years which he pulled out himself. Patient presented to the clinic with chief complaint of inflammation of the gingiva and movement of teeth. Intraoral examination showed inflamed gingiva and movement of all teeth.

OPG results showed short root and loss of bone. Lateral skull radiograph revealed more to Class II skeletal relationship, both maxilla and mandible are retrognathic, backward rotation of mandible and slightly protruded upper incisor. Patient is uncooperative and refuse further treatment. Treatment plan was to have all permanent teeth extracted and replace with upper and lower denture. Implant was not an option due to the small size of his jaw.

Hard samples were sent to pathology for histologic review. Microscopic findings showed obliteration of the pulp with tertiary dentin. Only small amount of fibrous pulp with diffuse calcification is seen. Secondary dentin is observed with malformed dentinal tubules. Irregular cementum on the root surface is also present. The features are consistent with Seckel Syndrome. Molecular studies are important to subtype this lesion.

CASE REPORT 2

A 13-year-old male was born 1.5 kg, delivered at 32 weeks normal and grew-up better that older brother. Patient is 1m high and skin is better. His mental development is better with 85-90 IQ and he is now in the fifth grade. Blood examination results showed within normal limits as shown in Table 1.

First tooth erupted in 6 months till 2 years and exfoliated gradually till his 10 years. Chief complaint when patient presented to the clinic was to replace anterior teeth only. Intraoral examination revealed minimal inflammation with no movement.

OPG showed short root. Lateral skull radiograph showed skeletal more to Class II relationship, backward rotation of mandible and lower incisor retrocline. Patient is caries free and replacement of anterior teeth by band on molar was done. Patient was cooperative during the entire treatment.

Cephalometric measurements of patients in Case 1 and Case 2 were detailed in Table 2.

Treatment plan was done for both of them. There was no need for them to receive antibiotic prophylaxis. They were given oral hygiene instructions and were recalled every 3 months.

Table 1. Blood examination results of the patients.

BLOOD EXAMINATION	PATIENT 1	PATIENT 2	REFERENCE
WBC	10.3	6.8 N	6.0-17.5
RBC	3.2L	4.2N	3.7-5.3
HGB	9.5L	12.5N	10.3-13.5
HCT	0.2L	0.39N	0.33-0.39
MCH	29	30N	23-31
MCHC	32.9	33.2N	31-35
RDW	15	14N	11-14
PLT	290	335N	150-450
MPV	8.6	11.1	
LYMPH	5.8	3.9N	4.0-10.5
MONO	1.2H	0.6N	0.2-1.3
NEUTS	3.1	1.2N	1.5-8.5
EOS	0.1	0.2	0.0-0.1
CALCIUM	2.4	2.18 N	2.2-2.7
CORRECT CALC	2.4	2.13N	2.2-2.7
SODIUM	138	145N	40-220
POTASSIUM	4.9	4.7N	3.2-5
CREATININE	0.1L	17.1N	8.8-17.7
UREA	5.5	4.4N	2.3-7.5
INORGANIC PHOSPHATE	2.06H	1.2N	0.6-1.5
ALKALINE PHOSPHATASE	1143H	358N	0-390
ALANINE TRANAMINASE	377H	15N	2-40
GROWTH HORMONE	15.3H	3.9N	0-10
TSH	12.6H	3.0N	0.25-3.1

TABLE 2. Cephalometric measurements of the patients.

	PATIENT 1		PATIENT 2	
	Value	Dev Norm	Value	Dev Norm
SNA (^{0})	74.3	-2.2	71.7	-2.9
SNB (^{0})	73.0	-2.3	72.0	-2.9
ANB (^{0})	1.3	-0.2	0.90	-0.6
Occ Plane to SN (^{0})	15.7	0.5	21.3	2.8
Pog – NB (mm)	0.6	-1.4	1.0	-1.0
MP – SN (^{0})	44.2	1.9	40.8	1.3
FMA (MP-FH) (^{0})	31.9	2.0	29.7	1.4
Interincisal Angle (U1-L1) (^{0})	117.4	-2.1	122.0	-1.3
U1 – SN (^{0})	113.0	1.8	99.2	-0.7
U1 – NA (mm)	4.1	-0.1	2.3	-0.7
U1 – NA (^{0})	38.7	2.8	27.4	0.8
L1 – NB (mm)	4.4	0.2	5.4	0.8
L1 – NB (^{0})	22.7	-0.4	29.8	0.8
FMIA (L1-FH) (^{0})	62.5	-0.4	52.2	-1.5
IMPA (L1-MP) (^{0})	85.5	-1.4	98.1	0.4
Lower Lip to E-Plane (mm)	1.0	1.5	0.1	1.1
Upper Lip to E-Plane (mm)	-1.0	3.5	-1.2	2.7
Soft Tissue Convexity (^{0})	120.7	-2.3	120.6	-2.8
Wits Appraisal (mm)	1.5	2.5	-0.4	0.6

CONCLUSIONS

In conclusion, the extraoral findings are congruent to Seckel Syndrome and is autosomal recessive. As a child is born prematurely and with low birth weight, the more it is prone to develop gingival inflammation, periodontal disease, shorter root, mobile teeth, easily shedding of teeth and enamel hypoplasia. The early resorption and premature exfoliation of the primary molars exhibited to the pulpal inflammation related to the caries. Therefore, a comprehensive dental care management for Seckel Syndrome patients is important.

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LEGENDS OF FIGURES

(a) Lateral cephalogram of patient 1, (b) Panoramic radiograph of patient 1, (c) Intraoral view of teeth of patient 1, (d) Hands of patient 1 showing deformity and roughly hyperkeratinized skin, (e) Lateral view of patient 1, (f) Frontal view of patient 1, (g) Patient 1 and 2 compared to each other, (h) Frontal view of patient 2, (i) Lateral view of patient 2, (j) Hands of patient 2 showing better skin, (k) Intraoral view showing anterior teeth of patient 2, (l) Panoramic radiograph of patient 2, (m) Lateral cephalogram of patient 2.