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RUSSELL-SILVER SYNDROME – A RARE CASE REPORT

Dental Science

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ABSTRACT

Russell-Silver Syndrome (RSS) is a hetergenous syndrome which is characterized by severe intrauterine and postnatal growth retardation and typical dysmorphic features. The clinical picture is extremely diverse due to numerous diagnostic features reflecting a heterogeneous genetic disorder. It is a syndromic disorder present at birth that involves poor growth, low birth weight, short stature and differences in the size of the two sides of the body. This case report presents a dental management of an eleven year old female patient with Russell-Silver Syndrome

KEYWORDS

Russell-silver Syndrome, Short Stature, Intrauterine Growth Retardation.

INTRODUCTION

The Russell-Silver Syndrome (RSS) is a single entity characterized by prenatal growth deficiency and postnatal dwarfism. It is a pattern of malformation first described by silver et al in 1953 based on two cases and then by Russell in 1954 based on seven cases. The incidence various from 1:3000 to 1:100000 live births and the chromosomes consistently implicated are the 7 and the 17. Parents with this syndrome do not have any mental problems, therefore this specific considerations may not be required during oral management and preventive treatment.[1]

Criteria for diagnosis include intrauterine and postnatal growth retardation, limb asymmetry and triangular shaped face. Other features occasionally associated include downtuned corners of the mouth, fifth finger clinodactyly, low set ears, weak muscle tone, blue sclera, café-au-lait spots and achromic patches.[2]

Babies with this condition have a low birth weight and often fail to grow and gain weight at the expected rate. The syndrome is associated with an increased risk of delayed development and learning disorder. Though the disorder is not uncommon, it is commonly not diagnosed, due to lack of awareness among practicing pediatricians and the subtle clinical features. This article describes the dental management of a female patient with Russell-Silver Syndrome.[3]

CASE REPORT

An eleven year old female patient reported to the Department of pedodontics with the chief complaint of intermittent and pricking type of pain in lower front teeth for the past 2 months. Medical history and records revealed the presence of Russell-Silver Syndrome. According to her medical history, growth hormone was never administered to the patient during her childhood.

During the clinical examination the patient presented a short stature 140cm, leg asymmetry of 4cm and a small size of the fifth finger in both hands. No mental retardation was noticed.

Examination revealed a triangular face with asymmetrical ears, the mandible was small with pointed chin and the corners of the mouth were turned down. The lateral facial view revealed a convex profile. Panoramic radiographs showed clinically missing 16, grossly destructed 31 and 41 and root stump 36 and 42. Intraoral radiograph revealed well defined periapical radiolucency in relation to 31, 41 and 42. However patient presented delayed dental eruption and diagnosis of Russell-Silver Syndrome was done on the basis of clinical and radiographic features.

As treatment part, oral prophylaxis, root canal treatment in 31, 41 and 42, extraction of Rs 36 and 42 followed by removable partial denture in relation to 16, 36 and 42 were done. Oral hygiene instruction was given and patient reviewed periodically every month.

Fig – 1 Facial profile

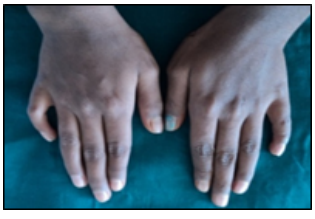

Fig – 2 Shortened fifth finger

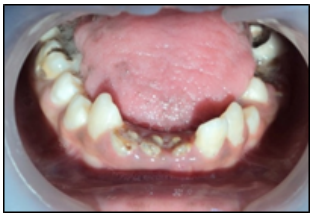

Fig – 3 Intraoral view

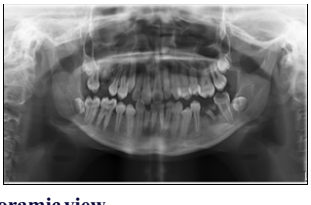

Fig – 4 Panoramic view

DISCUSSION

The occurrence of Russell-Silver Syndrome appears to be sporadic in most instances has been noted to occur in all racial groups. Maternal uniparental disomy of chromosome 7, in which a child has inherited both copies of a region of the chromosome from the mother, has been shown to play a role. It is important to note that no single explanation to date can account for the hetergenecity of the phenotypic findings. Russell hypothesized an intrauterine challenge or stress at 6 to 7 weeks.[4]

Intrauterine growth retardation results in reduction in total body cell mass and after birth growth proceeds normally with the child always remaining small in comparison with their peers. Insufficient growth hormone secretion has been suggested as contributory factor in some studies. The craniofacial morphology involves frontal bossing, a reduced length of the total cranial base, small linear facial dimensions, decreased posterior facial height, mandibular retrognathia and reduced mandibular measurements.[5]

The dental manifestations reported as microdontia, congenital absence of lateral incisors and second premolars, presence of primary double molar tooth and more consistently relevant, crowding of the teeth, especially in the mandible. Major intraoral features of this syndrome that have been reported are a high arch palate, delayed tooth eruption, hypodontia and crowding.[6]

Episode of hypoglycemia is also an important feature in the diagnosis of RSS. In our patient, the episode of hypoglycemia is evidenced. Diagnosis of RSS remains clinical as no definitive etiology or specific tests has been established. The five core clinical diagnostic criteria are, intrauterine retardation, poor postnatal growth, preservation of occipito frontal circumference, classic facial phenotype, asymmetry (especially of the extremities).[7]

Due to the clinical and genetic heterogeneities of this syndrome, patients whose features fulfill four of these five criteria could be diagnosed with Russell-Silver Syndrome. General treatment includes growth hormone therapy, high caloric diet, and limb lengthening. Growth hormone therapy improves short term growth acceleration in patients with RSS despite adequate endogenous levels of the hormone.[8/]

Our patient had low birth weight, poor postnatal growth and development delay. The girl also had dysmorphic face, skeletal deformities and delayed bone age and also had clinodactyly, low muscle tone, hypoglycemia and feeding problems. Genetic analysis of our patient could not be due to lack of logistic support.[9]

Abnormal appearance and hypodontia, which are major clinical symptoms of RSS, may have adverse effects on the social behavior of the patient. Therefore, oral rehabilitation is necessary for the patient to have similar appearance and function of her/his peers.[10]

Hypodontia is attributed to craniofacial dysplasia of the RSS population, which results in the differing cranial base measurements and mandibular length. Decision making and interdisciplinary management of the RSS patient with orthodontics or prosthodontics is recommended to improve both the sagittal and vertical skeletal relationship during craniofacial growth and development as well as to provide improvements in esthetics, speech and masticatory efficiency.[11]

CONCLUSION

Russell-Silver Syndrome is a growth and skeletal disorder associated with significant morbidity. Meticulous history, examination and radiographic investigation can confirm the diagnosis. Early diagnosis and initiation of treatment is very much important to prevent complication.

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