

Functional Rehabilitation of Tooth in a Patient Affected with Spinocerebellar Ataxia

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Abstract:- In the current study we have tried to explain how emphasis should be put on healthcare of medically compromised children since childhood so that positive attitude can be instilled in these children and their parents towards oral health care so that dental health is not neglected and becomes an integral part of essential medical care. This article gives an insight about the spinocerebellar ataxia and the treatment modality to be given.

Keywords:- Spinocerebellar Ataxia, Functional Rehabilitation, Crown.

I. INTRODUCTION

Ataxia also known as loss of limb co-ordination affects particularly gait and causes problems in gross and fine motor control. It may be acquired or genetic in nature. The timing of onset and family history may help in differentiation between an acquired or genetic defect. Spinocerebellar ataxia are autosomal dominant progressive disorders in which degeneration of cerebellum slowly occurs, often associated with degenerative changes in brainstem, parts of the central nervous system and at times peripheral nervous system as well.¹ There are at least 27 known variants of spinocerebellar ataxia which continue to grow.

There are 3 predominant genetic categories observed in spinocerebellar ataxia: Expanded CAG/ poly Q ataxias, ataxia caused due to conventional mutations (missense, insertion, deletion, duplication) and lastly non protein coding repeat expansion ataxias.² Many SCA present with extensive cerebellar atrophy with involvement of all regions of cerebellum, including molecular, granular cell layer, purkinje cells and deep cerebellar nuclei. SCA's are often differentiated on the basis of their extracerebellar brain involvement. ²SCA's may show a varied phenotype and dynamic mutations which expand to change size and thus larger expansions may show an earlier onset of disease wherein small expansion may show a later onset of disease.

This expansion may increase when transmitted from one generation to next generation.²

Gene testing may be a powerful tool for diagnosis and prediction of spinocerebellar ataxias.³ The clinical features of spinocerebellar ataxia may include significant central nervous system involvement that extends beyond the cerebellum to the brainstem (medulla and pons) and spinal cord. Symptoms of brainstem motor neuron loss may include temporalis muscle atrophy, tongue atrophy, facial weakness, and fasciculations. Spasticity and hyperreflexia may be seen in upper motor neuron involvement. Sensory and motor problem may occur in peripheral nerve involvement. Dystonia or bradykinesia may be seen in basal ganglion involvement. Many of the patients may be bound to a wheelchair prone state as they progress in age. Nystagmus may occur in pure cerebellar ataxia's without extracerebellar involvement.^{4,5}

The pathogenesis of many neurodegenerative disease occurrence is poly Q encoding CAG repeats. A current prevailing view is suggestive that a toxic action may occur at protein level ^{2,6,7}. The oral aspects and management of such cases has been infrequently spoken of hence this case report aims at presenting the aspects that were kept in mind while treating the case. This case report represents the dental management by functional rehabilitation of a tooth in a case affected with spinocerebellar ataxia.

II. CASE REPORT

An 8-year-old patient reported to the Department of Dentistry, as a referral from Department of Paediatrics with a chief complaint of severe pain in upper left back tooth region in the past 2 days. The patient had reported with a medical history of unstable gait, frequent falls, requirement of assistance in carrying out activities of daily living like bathing and dressing, no delay in milestones of development and no history of seizures. The child's IQ assessment done using Binet-Kamath test revealed the child to have an IQ of 84 which falls into an IQ category of below average. No relevant family history was reported in pedigree analysis. In

drug history the child had been consuming Cap Evion 1 capsule once daily since the past 1 month. The diet history of patient revealed that the patient was mainly consuming divided proportion of fruits, pulses, cereals, milk and eggs in his diet in three proportional meals with less in between meal snacking. The child had infrequent exposure to sugar, aerated beverages, toffee and chocolates. The diet mainly comprised on non-sticky fibre rich food items and less sugar rich food items.

The physical examination of child revealed that the patient had an unstable gait, difficulty in walking requiring often parental assistance and easy fatigability. The child had difficulty in being seated on the dental chair by himself and needed assistance. The child was slow in speech and spoke with complete yet very short and slow responses and was slow in compliance of any commands. The child however was very co-operative and adaptable. No other delayed milestones of development were noted.

On intraoral examination of child, it was observed that the child had no delay in eruption pattern, development of dentition or shedding pattern of teeth. The patient had an overall good oral hygiene with good gingival health. The soft tissue findings of the child including tongue, hard palate, soft palate, floor of mouth, anterior and posterior faucial pillars were essentially normal and healthy in appearance. The intraoral hard tissue findings revealed that the child had mixed dentition with the eruption pattern of permanent teeth falling in line with the age of the patient. Only one carious tooth, left deciduous upper second molar (65) was reported. Other teeth were free of caries. The tooth had deep occlusal decay with symptoms of an irreversible pulpal involvement, as the child had severe pain, sudden in onset lasting for longer duration, sharp shooting in nature which was exacerbated with hot and cold fluids and food and relieved by medication, ibuprofen(200mg).

Treatment plan was made and explained to the parent. Physician consent for carrying out necessary dental intervention was taken. Parental consent was as well sought. The child was made comfortable in the dental setup, chair was adjusted so that the child could easily settle in, and was explained about the procedure planned to be carried out. Tell show-do technique and euphemisms were used to explain the armamentarium and procedure to make child comfortable with the clinical environment. Child showed a very co-operative behaviour towards beginning of the procedure and to the commands that were being given to him. An intraoral radiograph was taken which revealed proximal caries involving enamel, dentin and progression towards pulp chamber (Figure 1). Emergency access opening was planned in the first visit keeping in mind that the child was compliant enough for undergoing such procedure in his first visit and the intensity of pain that he had reported with. Emergency access opening was carried out in the first visit, pulp chamber and canals were debrided with copious irrigation with sodium hypochlorite (5.25%), chlorhexidine, EDTA (17%) and saline keeping good suction to prevent the child from swallowing the irrigant solutions. The child was given a closed dressing with

interim restorative material (IRM). In his next visit, the patient was relatively free of symptoms, so working length was taken with the help of Propex Pixi apex locator (dentsply Sirona and Maillefer), and canal patency was established with size 10 K file and then 15 K file. Rotary preparation of canal space was done with Kedo-S rotary file system (Reeganz dental care Pvt Ltd. India) up to size D1 for mesiobuccal and distobuccal canal due to its narrow anatomy, and size U1 for palatal canal at a rotational speed of 300 rpm per minute and torque of 2.4 Ncm. After adequate preparation of canals, obturation was done with Metapex (Meta Biomed)(Figure 2). The chamber was then given a closed dressing with Cavit- G(3M ESPE) temporary filling. In the third visit the tooth was asymptomatic hence a permanent post endodontic restoration was done with packable posterior composite (Ivoclar Vivadent Tetric N Ceram)(Figure 3). In the next visit, crown cutting was performed for the patient and stainless steel crown (3M ESPE) was luted on the tooth 65 with luting GIC(3M ESPE) for its functional rehabilitation(Figure 4). The patient came for follow up after 1 week and was relatively free of symptoms and was able to chew from the involved side without any discomfort. Fones technique of brushing was demonstrated to the child and parents and possibly the use of newer battery-operated tooth brush was instructed so that the child can continue to maintain a good oral hygiene despite his decreased manual dexterity. However, a long follow up could not be scheduled as the parents refused for any follow up visits as they wanted to focus on child's medical treatment due to financial burden of treatment and the pressure of parenting a special child.

III. DISCUSSION

The present case represents dental management of a child affected with Spinocerebellar ataxia and presents an overview of systemic manifestations seen in a child affected with it and how they might affect treatment strategies.

Children are less often affected with spinocerebellar ataxia as its onset occurs usually after 18 years of age mostly except for in certain cases. The symptoms gradually worsen over the period of years.⁸

In the present case when the child made his first dental visit it was observed that the child had difficulty in maintaining balance while walking, needed assistance while walking, was easily fatigued and had difficulty in being seated on the dental chair. He needed support in the form of towel wrappings around legs and arms to keep him upright while undergoing the procedure. Children with Spinocerebellar ataxia often are affected by progressive in-coordination of walking, poor- coordination of hand movements, speech and eye movements.⁸

The child could comprehend all commands being given to him but was slow pertaining to his response levels. His IQ assessment done using Binet-Kamath test revealed that he had an IQ level of 84 which falls into an IQ category of below average. Dysarthria or speech impairment may present as a finding in cases affected with spinocerebellar ataxia.⁸

However, rubber dam could not be used in the present case as the child had easy fatiguability and difficulty with keeping the mouth open for a prolonged period. Children with Spino-cerebellar ataxia may also present with symptoms of brainstem motor neuron loss including temporalis muscle atrophy, tongue atrophy, facial weakness, and fasciculations.^{4,5}

After emergency access opening, instrumentation of canal was done with Kedo- S rotary file system. This newly introduced paediatric rotary file system is the first file system introduced for primary teeth. It comprises of three Ni-Ti rotary files with a total length of 16 mm and working length of 12mm. the three files are D1, E1 and U1. D1 has a tip diameter of 0.25 mm and can be used in narrow canals of molars while E1 has a wider tip diameter of 0.30 mm and can be used in wider molar canals. Rotary instrumentation was preferred in this case to decrease instrumentation time due to easy fatiguability of the child and improve the quality of obturation. Jeevanandan and Govindaraju evaluated instrumentation time and quality of obturation between paediatric rotary instrumentation with Kedo -S files and manual instrumentation technique in children of age 4-7 years in primary molars and concluded a positive reduction of instrumentation time and better quality of obturation when rotary Kedo-S files were used.⁹

Metapex (composition :30.3% calcium hydroxide, 40.4% iodoform,22.4% silicone oil and 6.9% others) was preferred as an obturating material as calcium hydroxide and iodoform mixture has been considered to be nearly ideal pulp canal filling material for primary teeth. Studies have reported that the material can be easily applied, resorbs slightly faster than the rate of root resorption of primary teeth, is radiopaque and has no toxic effect on permanent successors.¹⁰

Posterior packable composite restoration was preferred in this case as a strong evidence was drawn from a meta-analysis of 59 randomized controlled trials of both Class I and II composite showing overall success rate of about 90% after 10 years for the material.¹¹ Also the material is more preferred for a smooth finish line formation while crown cutting is being performed.

Stainless steel crown was also given to allow the teeth to be functionally loaded as the tooth had already lost a lot of coronal tooth structure and dentin due to caries and canal preparation. Also, the use of stainless-steel crowns prevents the teeth from future decay as they provide full coverage, increased durability and longevity and are also cost effective.¹²

Child and his parents were instructed for using Fones technique of brushing as the child had impaired manual dexterity and henceforth may be able to practice this technique effectively as self-help skills. The parents were also advised for using newly introduced battery-operated brushes (Oral- B Cross Action battery powered toothbrush) as they may aid in increasing the efficacy of cleaning and reduce the manual dexterity required while utilizing

traditional tooth brushes. Also, they are cost effective as they are not as expensive as electric tooth brushes.

The child however, did not report for his subsequent follow up visits as the parents were already going through tumultuous pressure of handling a special child, his medical visits and slow progress in school due to intellectual deficit. Their refusal to report for subsequent visits also puts an eminent light on the amount of pressure these parents go through while caring for a special child.

The dental management of children with special health care needs is often ignored due to such reasons and thereby should be given a priority so that these children can maintain good oral health and pain free life. A caries free mouth and well-maintained oral hygiene may aid them in achieving their nutritional requirements thus developmental milestones in a better manner. Also, the help of general anaesthesia may be taken in highly uncooperative cases.

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FIGURES



Figure 1: Pretreatment radiograph of tooth 65 showing carious progression involving enamel, dentin and pulp.

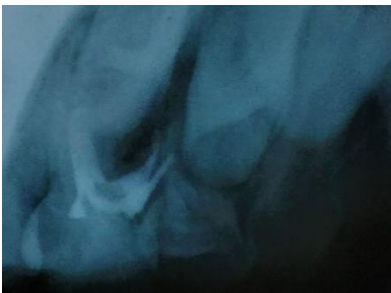


Figure 2: Post treatment radiograph post completion of pulpectomy in tooth 65.



Figure 3: Intra-oral photograph after completion of post endodontic restoration.



Figure 4: functional rehabilitation of tooth 65 with stainless steel crown.