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SPECIAL CHILD CARE: WEST SYNDROME MANAGEMENT DURING COVID-19 PANDEMIC

Dental Science

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ABSTRACT

West syndrome is a clinical condition characterized by the combination of an abnormal EEG showing chaotic pattern called hypsarrhythmia, infantile spasms, and delayed development or loss of acquired milestones. The oral manifestations of the conditions are wide and varied ranging from poor oral hygiene, bleeding gums, periodontitis, altered chronology and delayed eruption, high arched palate and extensive caries in multiple teeth. COVID-19 has affected the provision of dental care to all, especially special child care. Henceforth with an aim towards provision of treatment for reliving pain in needful patient, the oral health management was carried out keeping in mind the current pandemic scenario and practice restrictions.

KEYWORDS

West Syndrome, Oral Manifestations, Infantile Spasm, COVID-19, Special Care Dentistry

INTRODUCTION

West syndrome is a clinical condition characterised by the combination of infantile spasms, an abnormal EEG showing chaotic pattern called hypsarrhythmia and delayed development or loss of acquired milestones. It was named after the English surgeon Dr. William James West, who published the first description of infantile spasms in 1841, having witnessed the disease in his own son, James E West and named them "Salaam Tics".

Infantile spasms consist of a brief interruption of behaviour, with lifting and extension of the arms and bending forward at the waist. This may be accompanied by a rapid and forceful drop of the head. Individual spasms last only for a few seconds. Spasms often occur in clusters and a child may have several of them in a row, although occurrence of single spasms are also possible. After each episode the child may often cry as they resemble the waking up of child from sleep. About one-tenth of children with infantile spasms may not have an identifiable cause despite detailed evaluation.

Almost any brain abnormality or injury to the brain may potentially cause infantile spasms, such as brain malformations, brain infection, problems during birth like delayed cry leading to lack of oxygen or low blood glucose levels. These spasms are associated with Sturge weber syndrome, phenylketonuria, tuberous sclerosis, maple syrup urine disorder, incontinentia pigmenti, Otohara syndrome, biotinidase deficiency, pyridoxine deficiency and mitochondrial encephalopathy and abnormalities in ARX and CDKL5 gene [1]. The age of onset of infantile spasms ranges from 1 to 19 months [2]. The incidence is estimated at between 1 and 1.6/100,000 live births. Boys maybe more commonly affected than girls. [3]

Developmental delay and epilepsy may also present associated with west syndrome. Early diagnosis and treatment of west syndrome helps in better seizure control and improved development outcome. Electroencephalogram (EEG) may be the main modality of diagnosis for west syndrome, on which a chaotic pattern of brain waves called hypsarrhythmia may be noted. Apart from this, other investigations like Magnetic resonance imaging (MRI) of the brain may aid in finding the underlying cause.

A number of treatment modalities have been used for treating west syndrome. The most effective drug therapies include ACTH, steroids and vigabatrin. Other anti-epileptic medications are also used, though they are less effective. These include valproate, zonisamide, topiramate, clonazepam and clobazam. [4,5]

Oral manifestations of the syndrome are influenced by the severity of the condition. Patients may have delayed exfoliation of deciduous teeth and delayed eruption of permanent teeth. Dental implications include inability to maintain oral hygiene due to motor difficulties resulting in bleeding gums, gingival enlargement and periodontitis. Such patients usually have a high arched palate and multiple carious teeth. [6-9]

COVID-19 is a life-threatening pandemic which has infected over 63 million people, causing more than 1.4 million deaths worldwide as of November, 2020 [10]. Several countries entered a state of nationwide lockdowns such as United Kingdom, United states, India etc. The mode of transmission has been mainly described as close contact of over 10 minutes with respiratory aerosols of particle size less than 10 um [11]. While the recovery rate from the disease continues to improve, the impact on the regular oral healthcare facilities has been immense with patients receiving more conservative, interim and non - aerosol generating treatment modalities. Our case report aims to present the oral manifestations and their management in a patient diagnosed with west syndrome during the current COVID-19 pandemic scenario.

CASE REPORT

An 11-year-old female patient reported to the Department of oral and maxillofacial surgery with chief complaint of inability to eat properly due to generalised pain in the oral cavity and poor oral hygiene. The patient was triaged, wherein teleconsultation was done with the patient's guardian for primary care. Due to persistence of symptoms, patient was evaluated in the out-patient department following a negative COVID-19 rapid antigen test report.

Detailed medical history revealed, patient was the first offspring born out of a full-term normal vaginal delivery from a non-consanguineous marriage. She had delayed cry at the time of birth. Intake of antenatal iron and folic acid supplements was present. The parents had no associated co-morbidities. The patient had evolving cerebral palsy with spastic quadriplegia, global developmental delay and convergent squint. Infantile seizures began within 7 days of birth and her last reported episode was at the age of 9 months. EEG records at 6.5 and 8 months of age were consistent with modified hypsarrhythmia. Repeat EEG at 3.5 years of age reported normal sleep record with no epileptiform abnormalities. MRI brain at 4 months revealed ill-defined signal abnormality in high fronto-parietal region bilaterally, predominantly involving the white matter with diffuse thinning of corpus callosum suggestive of hypoxic insult at the time of birth rather than leukodystrophy. Repeat MRI at 11 years showed findings consistent with chronic hypoxic ischaemic insult. Gliotic changes

were seen in bilateral temporo-parietal regions and white matter along bilateral central and external capsule causing ex-vivo dilatation and undulating margins of lateral ventricles. Pre-operative electrocardiography (ECHO) revealed situs solitus laevocardia. The patient had been immunised as per schedule for her age and was not currently under anti-epileptic medications.

On general physical examination, the patient had inability to form coherent speech and did not respond to verbal commands but moved her head in the direction of sound. She had inability to sit, stand and hold her head position. She had hyperextended spine and pelvic deformity resulting in crossing of the right leg over the other. (Figure 1) Haematological investigations were within normal limits.

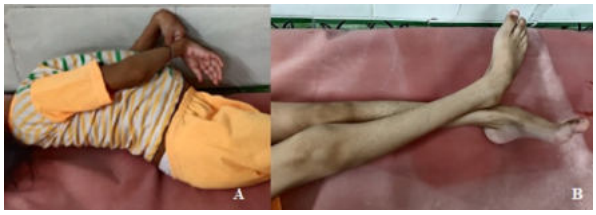

Figure 1 – (A) Hyperextensibility of spine and (B) Pelvic deformity causing crossing of the lower limbs.

Oral hygiene care for the patient was performed by the patient's mother with a small sized tooth brush once daily. Diet history of the patient revealed that she was on liquid to semi-solid soft diet primarily comprising of carbohydrate rich food. In between meal snacking was infrequent in the child.

Extra-oral examination revealed scarring over the midline of lower lip caused by chronic lip biting. She also had habit of bruxism. Intra-orally, the patient had poor oral hygiene with generalised gingival inflammation and calculus deposits. She had missing 12 and 22, palatally erupted 53, multiple carious teeth – 11, 16, 21, 26, 31, 41 along with multiple root stumps 64, 65, 73, 74, 75, 84, 85, 26, 46. Multiple hypoplastic white lesions were observed on the teeth owing to the lack of proper oral hygiene maintenance. She had a high palatal vault with decreased transverse width of the maxilla. An oro-nasal palatal fistula was present with respect to 15, about 1 cm from the cervical margin, caused by a previous extraction at a different centre 2 years back. (Figure 2) At present, there was no complain nasal regurgitation. Faucial pillars, buccal mucosa, tongue and floor of the mouth were apparently normal.

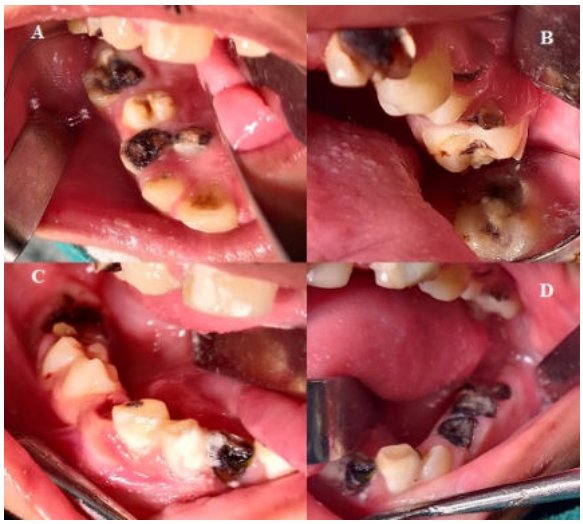

Figure 2 – Pre-operative intra-oral views of (A) Maxillary right quadrant, (B) Maxillary left quadrant, (C) Mandibular right quadrant and (D) Mandibular left quadrant.

After a negative Rt-PCR COVID-19 report, physician and pre-anaesthetic clearance, the procedure was carried out under general anaesthesia. Informed written parental consent was sought. Premedication included syrup promethazine, 2 hours prior to surgery. Following sedation, airway was secured with a flexo-metallic tube via the oro-endotracheal route. Cuffed tube was secured with a tube stich using 1-0 silk, to prevent extrusion or dislodgement. Throat packing was done to prevent the aspiration of fluid or debris.

Restorations were carried out first, followed by extractions. 11, 16, 21, 26, 31, 41 were restored using type IX glass ionomer cement (GIC). Extractions of deciduous root stumps - 73, 74, 75, 84, 85 and grossly decayed permanent teeth – 26, 46 was carried out. (Figure 3). Extraction sockets were secured with figure of eight suture using 3-0 polyglactin 910. Oral prophylaxis was done. Haemostasis was achieved. Oral cavity was cleared of secretions using high volume suction. Tongue stitch was made through the dorsal surface of the tongue and secured extra-orally to the cheek to prevent fall back and clear airway in the immediate post-operative period. Extubation was uneventful. There was mild ooze from extraction socket in the immediate post-operative period which was effectively managed with intra-oral pressure packing. No other treatment emergent adverse events were noted. Patient was discharged the next day after adequate observation. Further follow-up was done for a period of 2 months. Subsequently patient was kept on telecommunication as she moved to a different state.

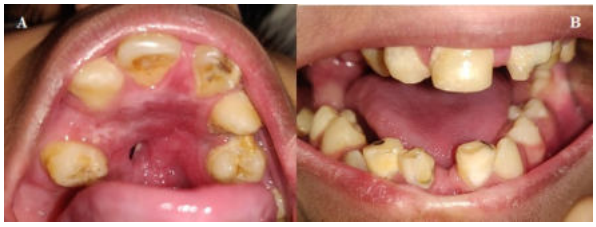

Figure 3 – Post-operative intra-oral views of (A) Maxillary arch and (B) Mandibular arch.

DISCUSSION

Oral hygiene care in syndromic cases always poses a challenge to the oral healthcare professional. Intellectual deficit, decreased manual dexterity, limb abnormalities and lack of special care prelude adequate oral hygiene maintenance in such patients. Carbohydrate rich semisolid diet further accelerate the caries activity. Therefore, primary prevention and timely intervention form the pillars of management of children with special care needs. Optimal function is facilitated by interdisciplinary action by paediatric dentist, oral surgeon, endodontist, anaesthetist, physiotherapist and speech therapist. The emergence of the COVID19 pandemic has raised new challenges in the field of special care dentistry, with primary focus on the minimisation of aerosol generation and appointment duration to reduce the exposure to the patient and the oral healthcare professional.

Patients with West syndrome may have prolonged retention, delayed or ectopic eruption, congenitally missing or impacted teeth, multiple decayed teeth, poor oral hygiene with periodontal problems, high arched palate and crowding of teeth [6-9]. Anti-epileptic medications administered have a propensity to cause gingival enlargement, further increasing plaque formation and calculus deposits. Patient compliance is proportional with the severity of the condition. Focussed dental care in compressed appointments is the key.

The patient presented to us during the national COVID19 lockdown period. Therefore, our goal was prompt definitive treatment with minimum exposure. In our case, lack of patient cooperation and risk of inadvertent aspiration during the procedure prompted us to choose general anaesthesia over conscious sedation or local anaesthesia.

Pre-operative RT-PCR was done to confirm the patient's COVID19 status. A COVID19 negative report was achieved hence, procedural intervention was scheduled. Deciduous root stumps of 73, 74, 75, 84, 85 were extracted in view of poor prognosis and succedaneous tooth eruption. 11, 16, 21, 26, 31, 41 were restored. Restorations were done with type IX GIC as Interim Therapeutic Restorations (ITR) in lieu of ease of working, chemical bonding with the tooth structure, biocompatibility and rapid setting time. It also acts as a fluoride reserve preventing the progression of active carious lesion, thereby conditioning them to receive a definitive permanent therapy [12,13]. The American Academy of Paediatric dentistry, in its revised policy of 2017, has also recognised ITR as a beneficial provisional technique in contemporary paediatric restorative dentistry [14]. Hence in our case, it was used as an interim restorative material, as the patient had multiple active carious lesions which benefitted from the fluoride releasing effect of GIC.

Extractions were performed for 73, 74, 75, 84, 85, 26 and 46 due to their poor prognosis and pain and discomfort being profound in the child. Fluoride containing toothpaste was recommended with a powered tooth brush to improve oral hygiene status. Controlled non-cariogenic diet was advised to reduce caries activity and oral hygiene maintenance. Parental counselling played a pivotal role in improvement of oral hygiene.

Reports over the years have provided dental care customised to cater the patient's individual needs comprising of preventive schedule, oral prophylaxis, restorations, pulpotomy/pulpectomy of salvageable teeth and extractions for teeth with hopeless prognosis [6-9]. Both preventive and interventional treatment measures help in improving the quality of life and oral health lead by children affected by it.

CONCLUSION

The management of West syndrome requires a multidisciplinary approach involving a paediatrician, neurologist, dental professional, speech therapist and physiotherapist. The therapy is directed towards control of seizures and management of oral manifestations according to their severity. It also garners significant responsibility on the part of the treating specialised oral health practitioners to provide prompt treatment to such individuals to lead a pain free life.

REFERENCES

1. National organization for rare disorders (NORD). (2020). West syndrome, https://rarediseases.org/rare-diseases/west-syndrome, (accessed 30 November 2020).
2. Singhi, P., & Ray, M. (2005). Profile of West syndrome in North Indian children. Brain & development, 27(2), 135–140. https://doi.org/10.1016/j.braindev.2003.10.007
3. Orphanet. (2020). Rare diseases - West syndrome, https://www.orpha.net/consor/cgi-bin/OC_Exp.php?Lng=GB&Expert=3451, (accessed 30 November 2020).
4. Hancock, E. C., Osborne, J. P., & Edwards, S. W. (2013). Treatment of infantile spasms. The Cochrane database of systematic reviews, (6), CD001770. https://doi.org/10.1002/14651858.CD001770.pub3
5. Wilmshurst, J. M., Gaillard, W. D., Vinayan, K. P., Tsuchida, T. N., Plouin, P., Van Bogaert, P., Carrizosa, J., Elia, M., Craiu, D., Jovic, N. J., Nordli, D., Hirtz, D., Wong, V., Glauser, T., Mizrahi, E. M., & Cross, J. H. (2015). Summary of recommendations for the management of infantile seizures: Task Force Report for the ILAE Commission of Pediatrics. Epilepsia, 56(8), 1185–1197. https://doi.org/10.1111/epi.13057
6. Regis, R. R., Rocha, C. T., Torres, C. P., Queiroz, I. F., & de Queiroz, A. M. (2009). Oral findings and dental treatment in a child with West syndrome. Special care in dentistry: official publication of the American Association of Hospital Dentists, the Academy of Dentistry for the Handicapped, and the American Society for Geriatric Dentistry, 29(6), 259–263. https://doi.org/10.1111/j.1754-4505.2009.00106.x
7. Khatri, A., Kalra, N., Tyagi, R., Baweja, M., & Khandelwal, D. (2014). Dental findings in patients with West syndrome: a report of two cases. Journal of the Indian Society of Pedodontics and Preventive Dentistry, 32(2), 168–171. https://doi.org/10.4103/0970-4388.130988
8. Dantas-Neta, N. B., Carvalho e Souza, C. H., Mendes Alencar, S. M., Prado Júnior, R. R., & Mendes, R. F. (2014). Dental findings in children with West syndrome. Special care in dentistry : official publication of the American Association of Hospital Dentists, the Academy of Dentistry for the Handicapped, and the American Society for Geriatric Dentistry, 34(6), 291–294. https://doi.org/10.1111/scd.12068
9. Della Vella, F., Contaldo, M., Fucile, R., Panza, F., Dibello, V., Kalemaj, Z., Ninivaggi, R., Petruzzi, M., & Serpico, R. (2019). ORO-Dental Manifestations in West Syndrome. Current topics in medicinal chemistry, 19(31), 2824–2828. https://doi.org/10.2174/1568026619666191114122732
10. Worldometer. (2020). COVID-19 Coronavirus pandemic, https://www.worldometers.info/coronavirus, (accessed 30 November 2020).
11. World Health Organization (WHO). (2020). Transmission of SARS-CoV-2: implications for infection prevention precautions, https://www.who.int/news-room/commentaries/detail/transmission-of-sars-cov-2-implications-for-infection-prevention-precautions, (accessed 30 November 2020).
12. Tyas M. J. (1991). Cariostatic effect of glass ionomer cement: a five-year clinical study. Australian dental journal, 36(3), 236–239. https://doi.org/10.1111/j.1834-7819.1991.tb04710.x
13. Croll, T. P., & Nicholson, J. W. (2002). Glass ionomer cements in pediatric dentistry: review of the literature. Pediatric dentistry, 24(5), 423–429.
14. American Academy of Pediatric Dentistry, (2020). Policy on interim therapeutic restorations (ITR), The Reference Manual of Pediatric Dentistry. Chicago, III.: American Academy of Pediatric Dentistry, 72-3.