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Case Report

Dental findings in patient with West Syndrome: A case report

Dr. Tariq Akbar, Dr. Sobia Hassan, Dr. Anum Zahid Kiani, Dr. Suman Arooj, Dr. Abdul Haq*, Dr. Areeba Shaheen Elahi

Department of Periodontology, Riphah International University

***Corresponding Author's Email: drabdul.haq15@gmail.com 00923149096181**

Abstract

We report a case of 2.5 years old male patient who presented with atrophy of upper and lower limbs, jerky head, developmental delay, severe redness in upper and lower dentition and hypsarrythmia on EEG. Later diagnosed as a case of West Syndrome. In order to increase awareness and preventive dental management regarding the dental problems in patients with West syndrome, I present a case of West syndrome having certain oral characteristics. Although this case didn't require extensive treatment but if neglected would have led to a more invasive dental treatment.

Keywords: Congenital abnormalities, Electroencephalogram, Infantile spasm, West syndrome

INTRODUCTION

West W. J first described the West Syndrome in 1841 (Wong, 2001). West Syndrome is characterized by a triad of infantile spasm, arrest of psychomotor development and profound paroxysmal electroencephalogram (EEG) abnormalities (hypsarrhythmia) (Proposal for revised classification of epilepsies and epileptic syndromes, 1989). The peak age of onset of disease is between 3 and 7 months commencing usually before 2 years of age (Hrachovy and Frost, 2008). Consensus statement of West Delphi Group (2004) categorized West syndrome based on etiology of infantile spasm into three subgroups: Symptomatic, idiopathic and cryptogenic. Symptomatic infantile spasm is where the origin of infantile spasm is identifiable such as pre/peri/post-natal cerebral ischemia; brain congenital abnormalities; chromosomal abnormalities; tuberous sclerosis; endocrine/metabolic etc. The idiopathic infantile spasms are with no identifiable underlying cause or neurological sign or symptoms. Cryptogenic is when there is suspicion of being symptomatic due to preceding developmental delay or neurological symptoms, but with no identifiable underlying structural or biochemical cause (Lux and Osborne, 2004).

In a study done by Regis et al, the major clinical features of West Syndrome identified were generalized tooth wear, gingival enlargement and multiple white spot lesions (Regis et al., 2009).

In another study done by Rômulo Rocha the major clinical features found were generalized tooth wear and gingival enlargement, altered chronology and sequence of dental eruption, primary canine cusp-to-cusp relationship, ectopic dental eruption, and mildly arched palate. Multiple white spot lesions were also observed, possibly associated with poor oral hygiene, due to a fermentable carbohydrate-rich diet, and continuous use of sugar-containing medications (Lúthvígsson et al., 1994).

Another study by Fedora Della Vella reported that West Syndrome patients present with multiple oral abnormalities, including altered eruption timing, teeth agenesis, teeth shape and position abnormalities, plaque and calculus accumulation, malocclusions and bad oral habits (mouth breathing, nails biting) (Della Vella et al., 2019).

A literature search (PubMed/Medline/Medknow/ Google scholar) retrieved only a few paper addressing

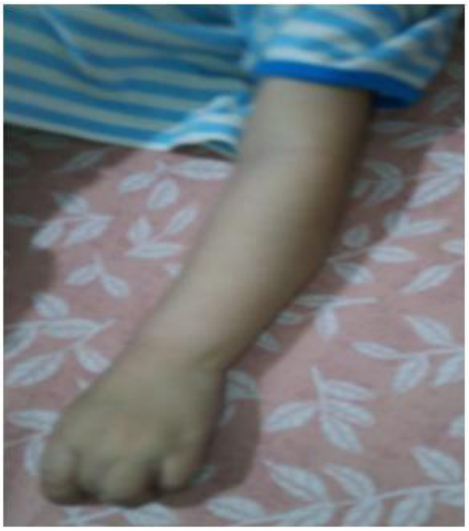

Figure 1a. Shows atrophy of upper limb.

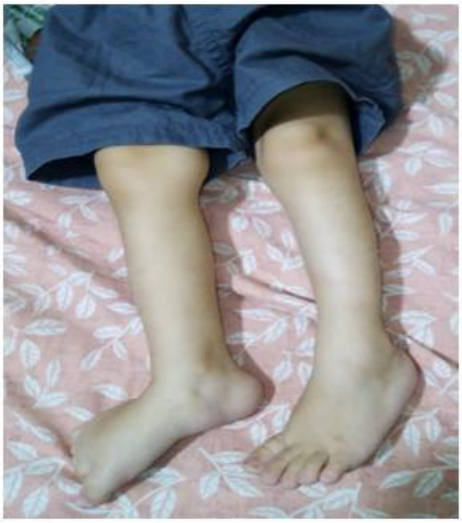

Figure 1b. Shows atrophy of lower limb.

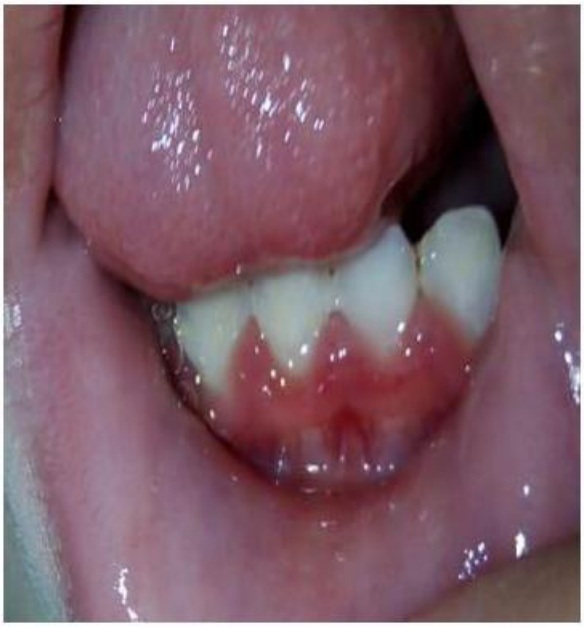

Figure 2. Shows generalized gingival inflammation.

dental findings and dental care management in West syndrome patient (Riikonen, 2001).

CASE REPORT

A 2.5 years old male child with a chief complaint of severe redness in upper and lower dentition presented to Periodontology Department, Islamic International Dental Hospital Islamabad. He was born to healthy consanguineous parents. The child was born by an uncomplicated caesarean delivery at full term and had a birth weight of 2.4 kg. The family history contained no report of similar cases. Patient had difficulty in breast feeding initially, at the age of 6 month the child had recurrent episode of a convulsive seizure. A diagnosis of West syndrome was based on EEG finding of hypsarrhythmia and infantile spasms (brief synchronous flexor or extensor spasm of head, trunk and limb), which occurs in cluster. Among the typical manifestation of west syndrome, neuropsychomotor delay is the most evident characteristic, especially atrophy of upper and lower limbs (Figure 1a and 1b). Review of the child medical history revealed that he had a delay in neuropsychomotor

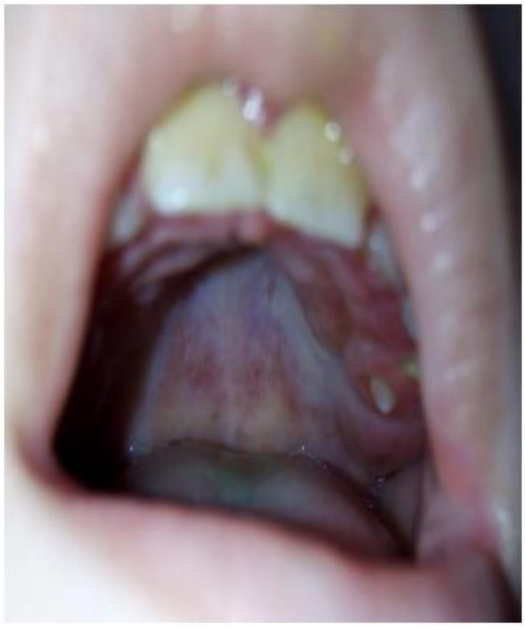

Figure 3. Shows palatal vault

development, which started at age of 8-9 month, even at age of 2.5 year he could not hold his neck or sit. Furthermore, the patient had poor concentration, not able to communicate and obey commands and displayed difficulties in understanding, focusing, restlessness and poor motor skills.

Intraoral examination revealed generalized gingival inflammation with bleeding (figure 2) mildly high palatal vault. (Figure 3) Generalized plague deposits were seen mainly due to the lack of oral hygiene maintenance (Figure 3). Tongue and mucosa were normal. Patient was prescribed topical anesthetic gel (lignocaine) to be applied on sore gums thrice daily. Mother was instructed about caries and periodontal disease prevention gave her diet counseling to reduce child's sugar intake and trained her to maintain proper oral hygiene. Emphasis was given for a recall visit after every 3 months.

DISCUSSION

Dental management of patients with special health care needs present unique challenges due to limited communication skill, severe intellectual deficit and inability to obey commands and follow oral hygiene instructions. Physical and behavioral characteristic of children with West Syndrome, lack of motor coordination, neurological developmental delay presents an additional challenge to the clinician.

This patient had the entire characteristic feature and was diagnosed as cryptogenic form of west syndrome due to unknown etiology, normal pregnancy and birth and the child had normal development prior to onset of infantile spasm. (Della Vella et al., 2019).

In a study done by Regis et al.[8]found in his study that major clinical feature of West Syndrome were generalized tooth wear, gingival enlargement and multiple white spot lesions. Another study also showed altered chronology and sequence of dental eruption, primary canine cusp-to-cusp relationship, and ectopic dental eruption (Regis et al., 2009). This patient presented to the hospital early enough that only gingival signs and mildly high arched palate were seen otherwise white spot lesions and dental caries can also be expected in such a patent if oral hygiene and other measures remained neglected. A delay in the dental eruption can only be judged when the eruption time is over by 6 months, which is not possible in my case since the patient is just 2.5 years old.

Children being treated with antiepileptic drug are more susceptible to developing periodontal problem (Angelopoulos, 1975). The most common antiepileptic drug that causes gingival enlargement is phenytoin (Tan et al., 2004; Tan et al., 2004). A study reported that phenytoin caused gingival overgrowth in a significant number of children (53.6%) within 3 months. Patients on carbamazepine did not show any signs of gingival overgrowth (Tan et al., 2004; Tan et al., 2004). My patient was taking carbamazepine for epilepsy control which indicates that the cause of gingival inflammation is not due to the antiepileptic medication rather is caused by the syndrome itself.

The high caries risk of patient with west syndrome is a result of their poor oral hygiene due to lack of motor coordination and cognitive impairment (Regis et al., 2009). The patient and patient's parent should be made

aware of importance of preventive measure such as controlled and non-cariogenic diet, regular use of sugar free medication, fluoride mouth rinse and fluoride dentifrices (Regis et al., 2009). Professional care such as oral prophylaxis, pit and fissure sealant must be provided where needed. Topical application of fluoride varnish can usually be used safely to prevent and treat enamel white spot lesion in such a patient (Regis et al., 2009; Khoroushi and Kachuie, 2017).

Riikonen (2001) showed that children affected with West Syndrome do have a normal lifespan although they often have profound mental and physical deficits. Therefore, patients with West Syndrome need more dental care along with medical care to avoid dental problems with emphasis on preventive care.

CONCLUSION: Provide note

REFERENCES

Appleton RE (2001). West syndrome: Long-term prognosis and social aspects. Brain Dev;23:688-91. [PUBMED]

Angelopoulos AP (1975). A clinicopathological review. Diphenylhydantoin gingival hyperplasia: 2. Aetiology, pathogenesis, differential diagnosis and treatment. Dent J;41:275-7, 283.

Della Vella F, Contaldo M, Fucile R, et al. (2019). ORO-Dental Manifestations in West Syndrome. Curr Top Med Chem.;19(31):2824-2828. doi:10.2174/1568026619666191114122732

Della Vella F, Contaldo M, Fucile R, et al. (2019). ORO-Dental Manifestations in West Syndrome. Curr Top Med Chem.;19(31):2824-2828. doi:10.2174/1568026619666191114122732

Hrachovy RA, Frost JD (2008). Severe encephalopathic epilepsy in infants: Infantile spasms (West syndrome). In: Pollock JM, Bourgeois BF, Dodson WE, Nordli DR Jr, Sankar R, editors. Pediatric Epilepsy: Diagnosis and Therapy. New York, NY: Demos Medical Publishing;.p.249-68.

Khoroushi M, Kachuie M (2017). Prevention and Treatment of White Spot Lesions in Orthodontic Patients. Contemp Clin Dent.;8(1):11-19. doi:10.4103/ccd.c

Li LM, Russo M, O'Donoghue MF, Duncan JS, Sander JW (1996). Allergic skin rash with lamotrigine and concomitant valproate therapy: Evidence for an increased risk. Arq Neuropsiquiatr;54:47-9.

Lúthvígsson P, Olafsson E, Sigurthardóttir S, Hauser WA (1994). Epidemiologic features of infantile spasms in Iceland. Epilepsia;35:802-5.

Lúthvígsson P, Olafsson E, Sigurthardóttir S, Hauser WA (1994). Epidemiologic features of infantile spasms in Iceland. Epilepsia; 35:802-5.

Lux AL, Osborne JP (2004). A proposal for case definitions and outcome measures in studies of infantile spasms and West syndrome: Consensus statement of the West Delphi group. Epilepsia;45:1416-28.

Proposal for revised classification of epilepsies and epileptic syndromes (1989). Commission on Classification and Terminology of the International League Against Epilepsy. Epilepsia;30:389-99.

Regis RR, Rocha CT, Torres CP, Queiroz IF, de Queiroz AM (2009). Oral findings and dental treatment in a child with West syndrome. Spec Care Dentist;29:259-63.

Regis RR, Rocha CT, Torres CP, Queiroz IF, de Queiroz AM (2009). Oral findings and dental treatment in a child with West syndrome. Spec Care Dentist.;29(6):259-263. doi:10.1111/j.1754-4505.2009.00106.x

Riikonen R (2001). Epidemiological data of West syndrome in Finland. Brain Dev;23:539-41. PUBMED

Tan H, Gurbuz T, Dagsuyu IM (2004). Gingival enlargement in children treated with antiepileptics. J Child Neurol.;19(12):958–63

Tan H, Gurbuz T, Dagsuyu IM (2004). Gingival enlargement in children treated with antiepileptics. J Child Neurol.;19(12):958–63

Wong V (2001). West syndrome - The University of Hong Kong experience (1970-2000). Brain Dev;23:609-15.[PUBMED]