Case report

Dental considerations in a 4-year-old girl with Lennox-Gastaut Syndrome. Case report and literature review.

Abstract

We present the developmental, oral, clinical, radiographic findings and oral treatment of a 4-year-old girl Lennox-Gastaut syndrome (LGS), which is a severe disabling childhood epilepsy diseases that is treated with one or multiple anti-epileptic drugs (AEDs). The child was wheel-chair bound, developmentally delayed, G-tube fed, and suffered from multiple seizures and infantile spasms, The child's medical history included an under-developed pituitary gland, gastro esophageal reflux disease, vision and hearing impairment, history of chronic aspiration pneumonia, and allergies. The oral findings included no carious lesions, heavy calculus accumulation, spontaneous bleeding from the gingiva, generalized gingival hyperplasia (GH) and abnormal increased mobility in several primary teeth. The comprehensive radiographic and clinical examination and the treatment under general anesthesia are described. The etiologies of the calculus accumulation and GH are reviewed.

Key words: calculus, gingival overgrowth, anti-seizure medication

Introduction

Lennox-Gastaut syndrome (LGS) is a severe and disabling childhood epilepsy that is characterized by a triad of symptoms: 1) generalized treatment resistant to multiple type seizures; 2) slowness of intellectual growth and cognitive impairment; 3) a specific electroencephalogram (EEG) disturbance called a slow spike-and-wave pattern that is present when the child is awake.[1-5] LGS patients may have multiple daily seizures that may cause sudden and

unpredictable stiffening followed by a drop to the ground; this being a key diagnostic feature.[5-7] The pharmacologic treatment may include one or multiple antiepileptic drugs (AEDs),[4] some of which have the potential to induce gingival hyperplasia (GH).

A review of the literature identified only one report of the oral findings in a LGS patient, of a 26-year-old female who had macroglossia, supragingival as well as subgingival calculus, red, swollen and friable gingiva with generalized bleeding and localized suppuration, and gingival recession.[4] The present manuscript includes an additional , comprehensive case report of a 4-year-old girl with LGS, and presents a review of the literature on LGS and related anti-seizure medication that may induced gingival overgrowth.

Case presentation

A 4.5-year-old Caucasian female with LGS was referred to a University Clinic for dental treatment. The medical history indicated that *she* was born at 32 weeks of gestation, along with her healthy twin. *The patient* had infantile seizures and spasms 15-16 times per day and was diagnosed with LGS. *Her* medical history was significant for developmental delay, wheelchair-bound, had a gastrotomy tube (G-tube), under-developed pituitary gland, gastro esophageal reflux disease, vision and hearing impairments, history of chronic aspiration pneumonia, allergies to Depakote and Amoxicillin and leukodystrophy (degeneration of the white matter in the brain[8]). Her medications included; Vigabatrin, Clobazam, Topiramate, Fycompa, Diazepam and Rufinamide reducing the daily seizures to 3-6, and Albuterol/atropine via nebulizer. Recent hospitalizations resulting from seizures, chronic pneumonia, and adrenocorticotropic hormone therapy. The surgical history included adenoid and tonsils removal, Nissen fundoplication with hernia repair, and G-tube placement. The chief complaint as expressed by her mother was risk of

aspirating exfoliating primary teeth: the previous night the patient had a seizure, after which she was "choking and was missing a lower tooth that was swallowed or aspirated".

On examination, she had no apparent respiratory difficulties, was non-verbal, had a small "hypoplastic" face, inability to cooperate, extensive drooling, short stature and slight overweight.[9] A limited oral examination revealed sialorrhea, primary dentition with missing mandibular primary central incisors, heavy calculus on the majority of teeth surfaces, abnormal mobility (2-3 mm) in both mandibular primary lateral incisors (teeth #N and #Q), as well as generalized moderate GH. Tongue size appeared normal. A chest radiograph did not reveal tooth aspiration.

MG was admitted to the hospital the day before the dental treatment under GA, maintained with intravenous fluid to avoid the conflict between being *nil per os* and her need for frequent G-tube feeding. *The* mother reported that the patient was apparently having pain while grinding her teeth. Under GA, a radiographic and clinical examinations revealed no caries, no evidence of dental pulp pathology (Figure 1), all maxillary primary incisors (Teeth D, E, F, G), and both mandibular lateral primary incisors (Teeth N and Q) had abnormal mobility (about 3 mm), nearly all teeth were covered with heavy calculus (Figure 2), generalized moderate GH, and a band of gingiva over the occlusal surface of the mandibular right first primary molar (Tooth S, Figure 3A), and gingiva over the occlusal surface of the maxillary right first primary molar (Tooth B, Figure 4a). The GH was non-hemorrhagic, soft, slightly fluctuant and pink (Figures 2, 3a&b, 4a&b). Calculus removal was accomplished with an ultrasonic and hand instrumentation, followed by an application of a fluoride varnish. The gingival tissue over teeth B and S were removed with a surgical blade (Figures 3b & 4b). Teeth # D, E, F, G, N and Q were extracted. The pot-operative recovery was uneventful.

Discussion and literature review

Dr. William Lennox, first described LGS in 1930s, Lennox and Davis later reported its triad, which was further expanded by Gastaut.[11-12] The median onset age of LGS is about 4 years (range: 0.6-28.9 years) with a peak onset of 5 years.[13-14] LGS is uncommon (3-10% of childhood epilepsy) and has a mortality rate ranging from 3% to 7%.[2, 3, 12] The tonic seizures are characterized by an EEG diffuse high voltage slow wave followed by generalized low voltage fast activity, reflecting sustained fast neurological firing over a wide cortical area.[5, 15] 80% of LGS patients will continue to have seizures into adulthood.[2, 16]

Based on our literature review, this is the second case in which the oral characteristics of LGS are described, and the first one in a child. In our case the dental consideration included behavioral and management issues, gingival hyperplasia as a result of side effects caused by anti seizure medication, poor oral hygiene and a risk of aspiration from loose teeth and difficulties in swallowing. Comparison of both cases is restricted by the patients different age groups; the previous report was in a 26-year-old female.[4] Both cases received AEDs and had GH and severe calculus accumulation, the previous case had periodontitis and macroglossia that encumber proper OH while in the present case the tongue size was normal and increased abnormal tooth mobility with no radiographic evidence of alveolar bone loss. Oral pain was reported in the previous case associated with gingival swelling, gingival recession and periodontitis while in the present case, pain was assumed to be related to biting on the gingival tissue over the occlusal surfaces.

GH commonly starts with the eruption of the permanent dentition and may be influenced by genetic predisposition.[17] However, in the present case there was no history of GH in the family,,[17] indicating that the GH may have been caused by one or more AEDs most likely

vigabatrin. The aim of AEDs is to control or decrease seizures without producing unacceptable adverse effects that impair quality of life; however, AEDs have been most frequently associated with adverse drug reactions.[17] The pharmacologic treatment of LGS includes AEDs such as vigabatrin, valproates, felmabate, and benzodiazepines which may potentiate each other side effects, as in cases in which GH is potentiated by the combination of phenytoin and calcium channel blockers, or cyclosporine and calcium channel blockers.[18-20]

Interestingly, multiple AEDs have an additive effect on GH, that might explain the additive effect of multiple anticonvulsant therapy to GH.[25]

GH might include an abundance of dense connective tissue or acellular collagen that can be an impediment to tooth eruption.[36, 37] Delayed eruption has also been associated with severe bruxism in children with cerebral palsy.[38, 39] In the present case, the primary dentition was normal.[40] However, the clinical crowns of the primary teeth appeared shorter than normal and there was gingival tissue at the occlusal surfaces of teeth B and S, suggesting a combination of GH and delayed eruption that could be related to the GH and bruxism (Figures ,3a, 3b).

Despite the positive correlation between plaque scores, gingival inflammation, and severity of GH in children, the role of OH as an etiologic factor for GH has not yet fully clarified since most of the studies have been cross-sectional.[19, 25] However, the relevance of OH is emphasized in the previously reported LGS case in which non-surgical periodontal therapy was effective in controlling periodontal disease, and prevention of oral diseases is preferable for high-risk patients.[4] In the present case however, OH performance is complicated by the child's inability to perform the most simple measures and to cooperate with her parents.

A full mouth gingivectomy in the primary dentition was reported by Breen et al. (2009) in a case of a 28 month old with hereditary gingival fibromatosis in which only 4 mandibular teeth

were partially erupted.[17] In the present case, we included the removal of the gingival tissue from the occlusal surfaces of the primary molars that most likely were the origin of oral pain (Figures 3b & 4b); in retrospective, a gingivectomy could have been adequate for the maxillary right primary cuspid and lateral incisor that had minimal clinical crowns (Figure 4a); the patient will continue to be under follow-up and will be scheduled for gingivectomy if required.

Children and adolescents who are unable to meet their nutritional needs orally and depend on G-tube feeding at a significantly increased risk of poor oral health, specially tartar accumulation an subsequent gingivitis.[10, 41, 46] In the present case, the possibilities of recurrence of calculus accumulation are high. Based on our search of the literature, it appears that this is the youngest case reported with severe generalized calculus accumulation.

Aspiration of exfoliating primary teeth is apparently most uncommon or non-reported since our review of the literature disclosed only one case of aspiration of a maxillary primary cuspid by a 9 year 11 month old child with cerebral palsy, emphasizing the fact that the possibility of aspiration of primary teeth is exacerbated in debilitated patients.[47] also, avulsion of primary teeth due to trauma and their aspiration is possible.[48] This emphasizes the need to consider the need to refer children who "lost" a primary tooth that cannot be found to a chest radiograph, especially in children with developmental disturbances, and a history of aspiration pneumonia which involves the entry of infectious pharyngeal contents into the lower airway.[41] Relevant is the fact that low salivary flow associated with GT feeding may predispose the growth of salivary bacteria that, when mixed with food or liquid, provide a substantial inoculum to the lungs if aspirated.[41]

In conclusion , LGS in young child presents a significant challenge to the dental professional, both the neurologist and the pediatric dentist should be aware of the potential complication and work as team on behalf of the patient and the family of the LGS patient.

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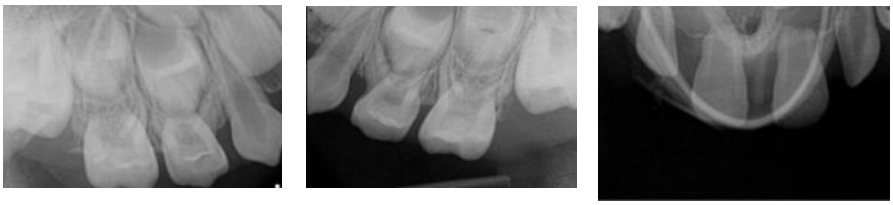

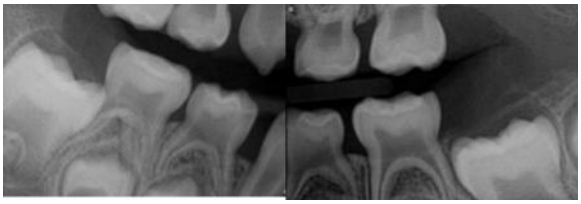

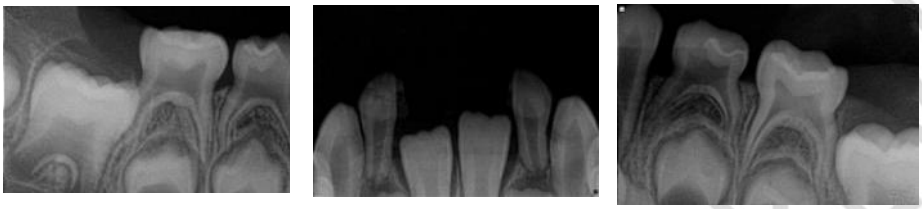

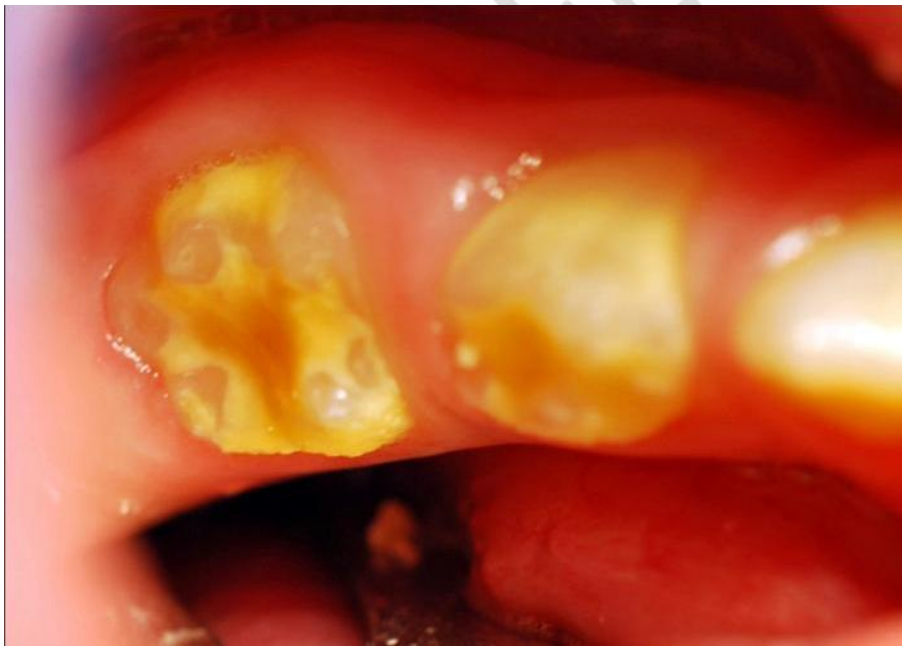

Figure-2

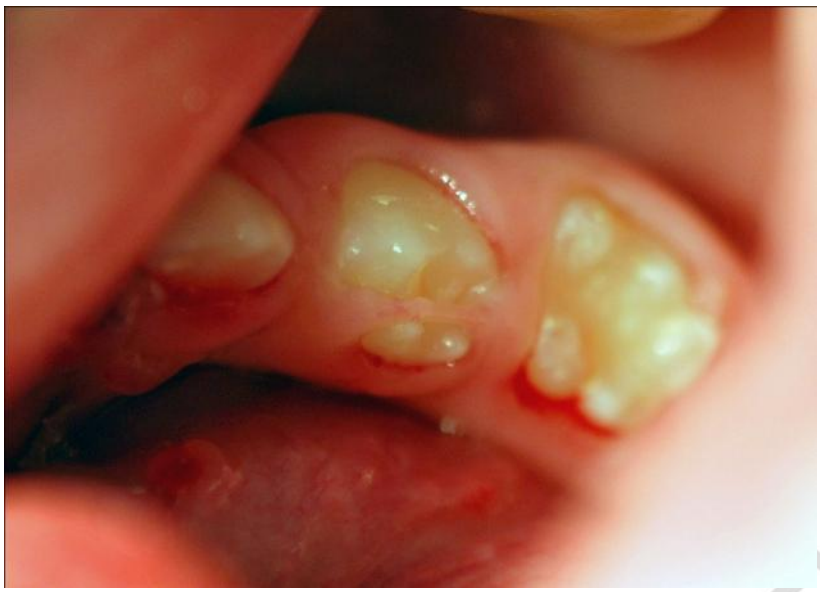

Figure-3a

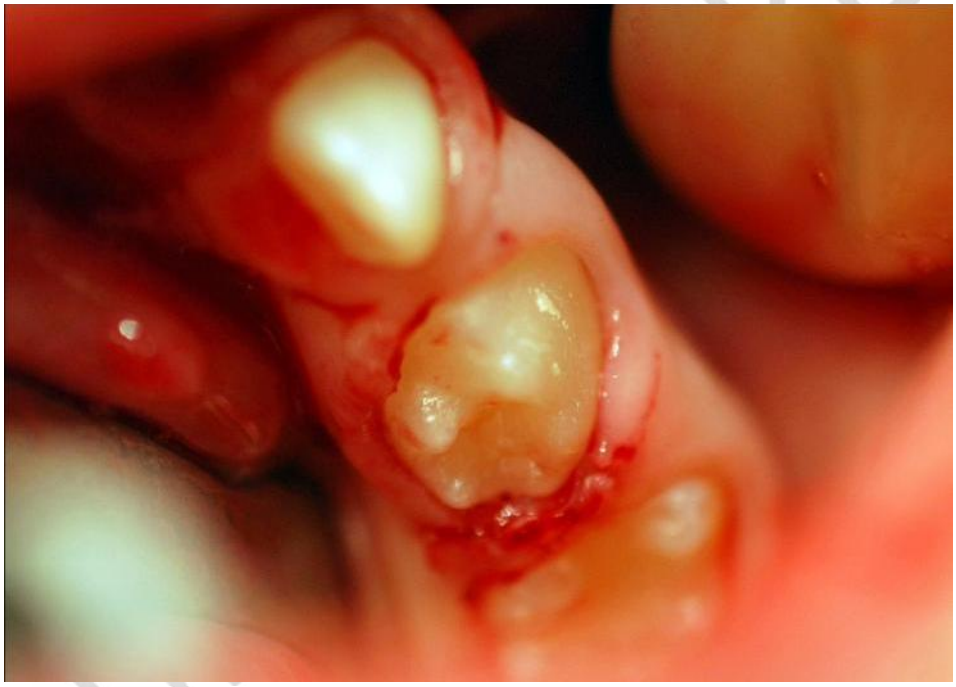

Figure-3b

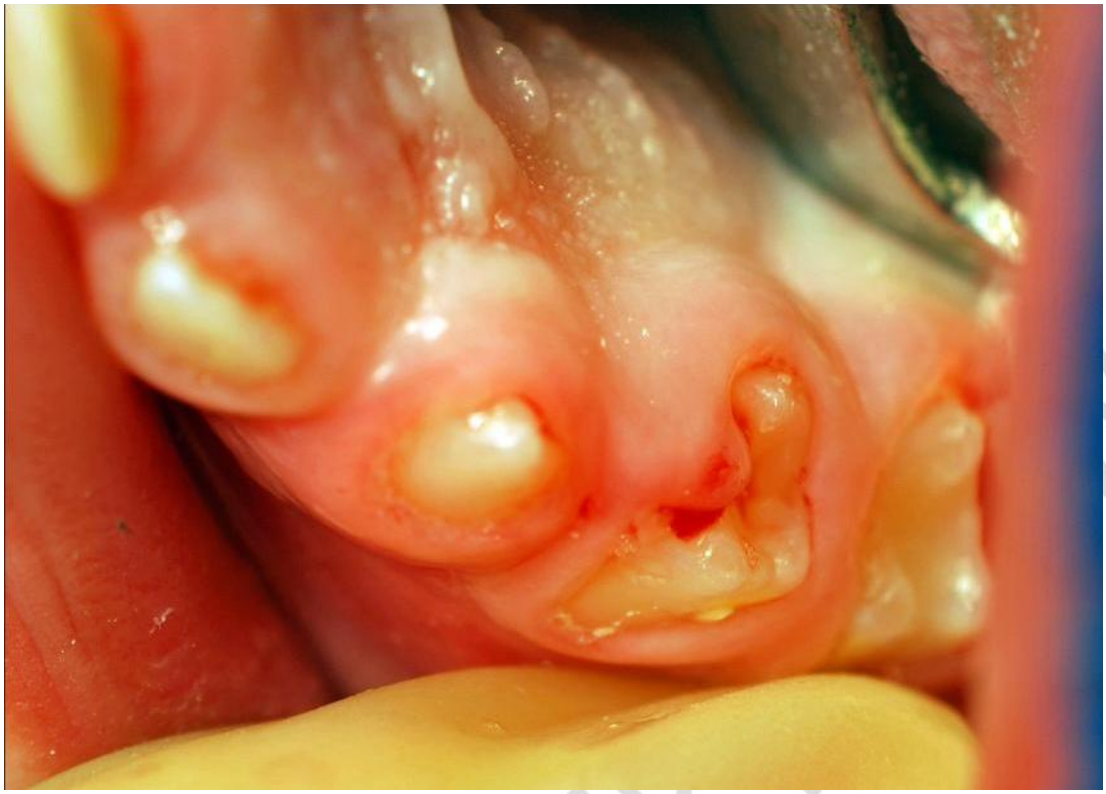

Figure-4a

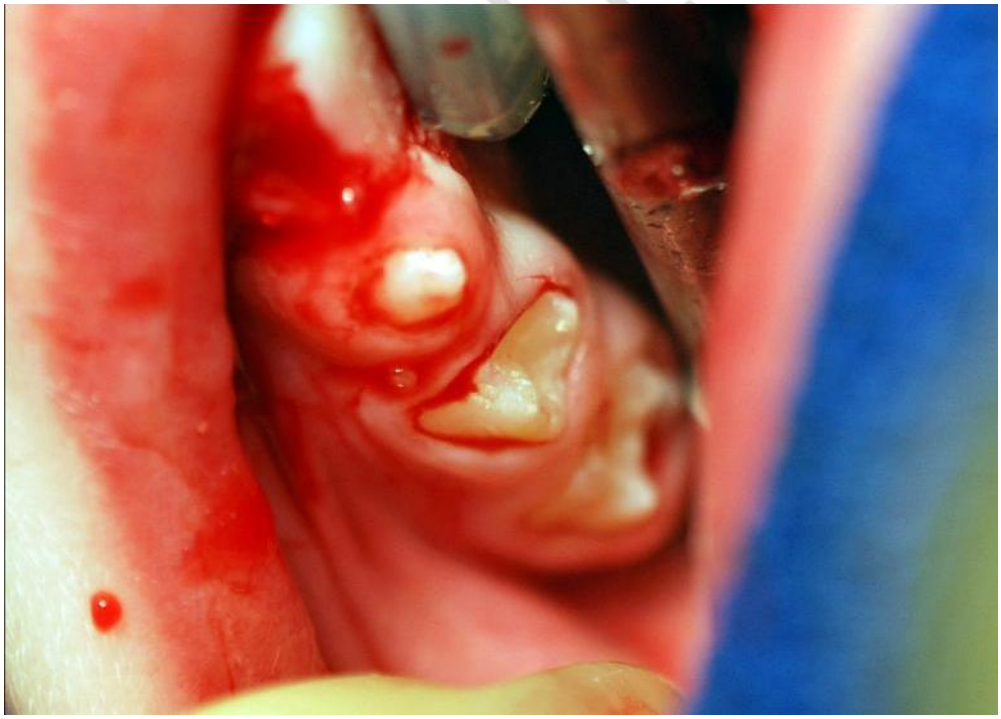

Figure-4b