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Case Report

Pediatric Dental considerations in Pachygyria-Polymicrogyria: A rare case report

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ABSTRACT:
Children with neuro developmental delay are at an increased risk for dental caries, missing teeth, orthodontic and periodontal problemsthroughout their lifetime. This group of patients not only presents challenges when they face dental treatments but also encounter more difficulties obtaining specialist dental care than other segments of the population. This paper attempts to highlight a case of Polymicrogyria and Pachygyria, a cortical malformation disorder, in a young boy reporting at an exclusive pediatric dental practice presenting with gingival bleeding and mobile teeth. Clinical examination suggested the presence of severe generalized gingival hypertrophy along with exfoliating teeth. Limited oral hygiene practices due to special needs challenge leading to poor biofilm control was observed. The parents were educated about adequate home oral hygiene protocol and were brought under dental home and anticipatory guidance. Palliative care with topical anaesthetics was advised for marginal gingivitis of the primary canines. Since this is one of the rare disorders, this case report also emphasizes on dental considerations and management of children with Polymicrogyria and Pachygyria.
Keywords: Pachygyria, Polymicrogyria, neuro developmental disorder, dental home

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INTRODUCTION

Malformations of cortical development (MCD) comprise disorders of disrupted cerebral cortex malformation caused by various genetic, infectious, vascular, or metabolic aetiologies (1). They are common causes of epilepsy and neuro developmental delay (2). Polymicrogyria (PMG), one of the most common MCD disorders, is characterized by abnormal cortical lamination and an unusual folding pattern of the cerebral cortex such that all or part of the brain surface is taken up by an excessive number of small gyri (folds). Although PMG is usually an isolated finding, it can sometimes occur in concomitance with other brain abnormalities. Pachygyria is a distinct brain malformation in which reduced or absent cortical folding is combined with a thick cortex(3). Pachygyria and PMG-like cortical malformations may result from mutations of genes that encode tubulin or microtubule-associated-protein (MAP) genes, which are critical for the migration of neurons (4).

The affected patients have a wide array of clinical presentations ranging from epilepsy, developmental delay, intellectual disability, hemiparesis, cerebral palsy, poor muscle control and feeding/swallowing difficulties. Severity depends on the location and extent of cortical involvement, but most patients have developmental delay or behavioural problems in childhood and around 70% develop seizures during the first 5 years of life (5). Diagnosis is mainly post-natal and is based on neuro imaging studies such as brain MRI.

Oral health is often neglected in patients affected by MCD due to a spectrum of neurological issues which are demanding to deal with. Majority of patients are on anticonvulsants which directly affect their oral

health and render them at an increased risk of Drug-induced Gingival enlargement (DIGE) and gingival bleeding. Preventive and palliative oral health care is significantly pertinent yet overlooked in these individuals. Despite being one of the most prevalent cortical malformations, there is scarce literature discussing its dental considerations. This report attempts to highlight a case of PMG and pachygyria in a 5-year-oldboy with emphasis on dental considerations and management.

CASE REPORT

A 5-year-old boy was referred to an exclusive Pediatric Dental clinic by pediatrician with the chief complaint of bleeding gums and mobile upper teeth for last 3-4 months. Medical history revealed that the child had PMG and Pachygyria which were detected on an MRI of the brain at 3-4 months of age after the first seizure. It was localized to front oparietal, temporal and occipital regions sparing the anterior frontal and anterior temporal regions. The child's medical history further revealed developmental delay and gross motor delay leading to delayed milestones, first evident at 2.5 years of age. There was speech and language delay as well. He had a history of seizures and was on multiple anti-epileptic drugs for the last 3.5 months. He had 3-4 episodes of poorly controlled seizures in a day lasting less than a minute. He had been recently hospitalized due to West syndrome with pneumonia and had a nasogastric tube inserted.

Intraoral examination was done under physical restraint due to the child's lack of ability to cooperate and revealed the presence of severe generalized gingival hypertrophy(Figure 1 - 4). There was gingival bleeding even on mild palpation. Limited oral hygiene practices due to special needs challenges leading to poor biofilm control was observed. Marginal gingivitis with respect to all deciduous canines was noted. There was physiologic mobility in the primary maxillary and mandibular central incisors which presented a potential aspiration risk. There was no history of pain or swelling in the mouth. History of oral deleterious habits of bruxism and mouth breathing was also elicited from the child's parents which indicated poor muscle coordination, control and involuntary oral activity. A preliminary dental diagnosis of DIGE secondary to medications-Midazolam, Sodium Valproate, Clonazepam, Brivaracetam, Perampanel was made.

Comprehensive pediatric dental counselling and oral health education was done with audio-visual aids. Detailed discussion about the treatment plan and the importance of establishing a Dental Home and preventive care for the child was done with the parents. A 3-step care plan was advised. Palliative care with topical anaesthetics for marginal gingivitis of the primary canines along with an oral hygiene protocol customised for special needsin form of pea sized 1000 ppm fluoride toothpaste, gum paint/gel along with warm saline rinses was advised. Extraction of the mobile maxillary central incisors was planned after clearance from his Pediatric neurologist to reduce aspiration risk. A nightguard/occlusal splint would be given to the patient for bruxism in the subsequent visits once primary anterior teeth exfoliate and will be re-fabricated semi-annually based on jaw's growth and development.

Figure 1: Intra-oral view of the left mandibular region: Gingival hypertrophy embedding the primary molars

Figure 2: Intra-oral view of the left maxillary region: Gingival hypertrophy embedding the primary molars

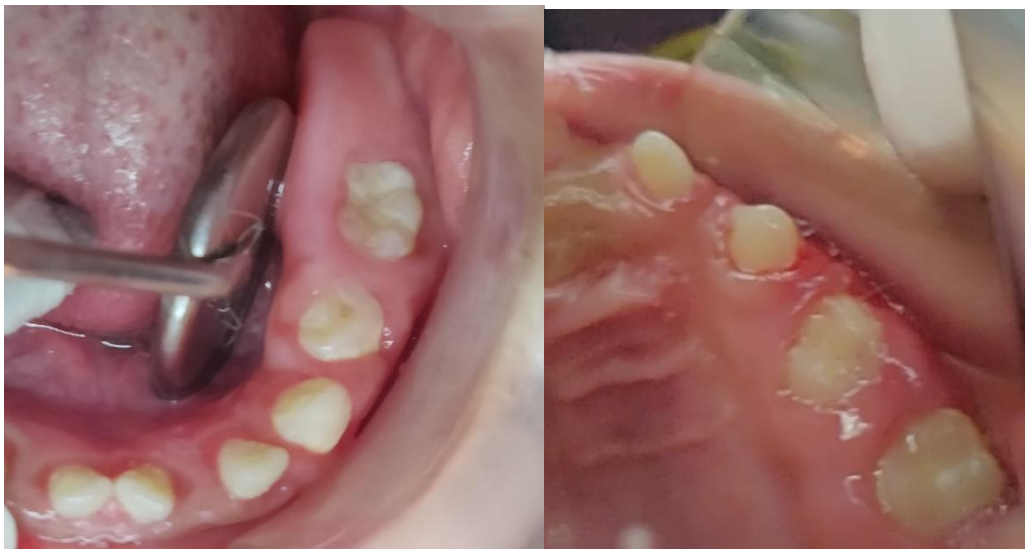


Figure 3: Intra-oral view of the right mandibular region: Gingival hypertrophy up to the occlusal surface of the primary molars

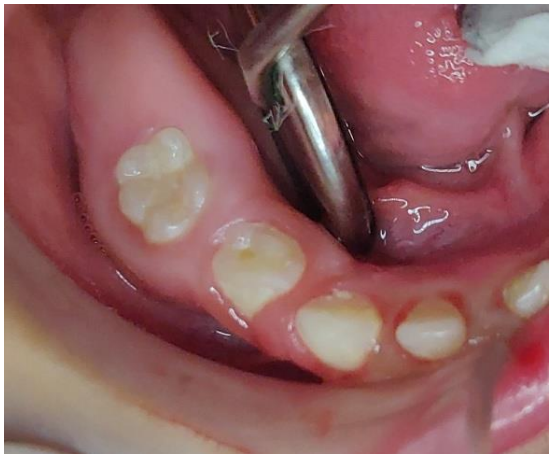
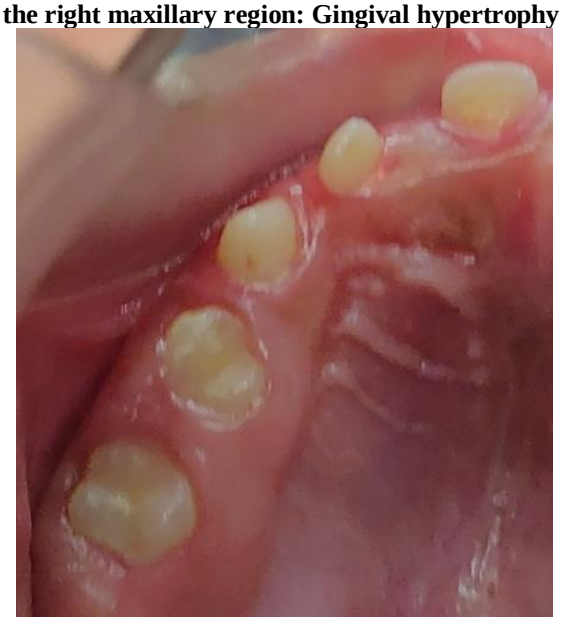


Figure 4: Intra-oral view of the right maxillary region: Gingival hypertrophy with bleeding on probing



DISCUSSION

As discussed previously, PMG, although rare, accounts for about 20% of all MCD. Depending on the severity of the condition and the parts of the brain affected, it may exhibit spectrum of neurological problems like epilepsy, delayed development, variable motor signs and cognition(6). The resulting lack of dexterity in patients and burden of care for the caregivers impede oral hygiene maintenance predisposing them to an increased risk of dental caries and periodontal problems. Additionally, individuals with PMG and Pachygyria are chronically on medications such as anti-epileptics and benzodiazepines which cause not only gingival changes but also alter their saliva leading to poor plaque control and an increased risk of periodontal problems, caries, and halitosis(7). The present case had developmental delay which was a significant contributory factor to his poor oral hygiene and excessive plaque accumulation. The use of drugs like Sodium Valproate for seizure control caused gingival hyperplasia by causing critical changes in fibroblast function, which results in an increase in the extracellular matrix of the gingival connective tissue(8).Planning dental treatment for patients with MCD can be challenging due to various barriers, such as behavioural challenges presented by the child, the unwillingness and unpreparedness of the dental professional due to the lack of knowledge of the condition, and the lack of parents' awareness and motivation. A multidisciplinary approach should be adopted for management which includes the Paediatric Neurologist, Paediatric dentist, Paediatric Occupational therapist and the child's parents working together in this quintet along with the child himself

Emphasis should be given to the prevention of dental diseases in consideration of the challenges the treatment imposes and the high burden of care on the caregivers. Parental counselling regarding preventive approaches and the dental home concept must be done, as was in this case. Oral hygiene and dietary

counselling, optimal fluoride supplementation can vastly help to decrease the incidence of new caries lesions. Flossing is recommended to remove interproximal plaque and a floss holder can be used in children with limited manual dexterity. For maximum protection against dental caries, professional topical fluoride applications can be combined with home-based fluoride paste (9). Most of these children ingest soft diet which contains refined carbohydrates. The parents/caregivers should be counselled regarding the non-cariogenic diet and strategies to alter the frequency if necessary (10). As per the guidelines on Caries Risk Assessment, children with special health care needs have High Caries risk and so a follow-up protocol becomes essential to meet their preventive needs (11). The family, in this case, was kept on a strict preventive protocol of 3 months where a preventive fluoride varnish application shall be planned.

CONCLUSION

Management of dental diseases in children with PMG-Pachygyria should be aimed at both short term and palliative care as well as long-term treatment options. The treatment modalities provided for the current patient aimed to control the progression of the disease including gingivitis and poor oral hygiene. However, more focus has to be given to long-term maintenance that impacts not only the oral health of these children but also their quality of life. Awareness about this disease among the fellow paediatric dental colleagues through this literature will help in serving children with this rare disorder in best possible way.

AUTHORS CONTRIBUTION

The work was carried out in collaboration among all the authors. All authors have read and approved the final manuscript.

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CONFLICT OF INTEREST

The authors have no conflicts of interest to disclose.

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