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Dental management of a child with alexander's disease

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Abstract
Alexander disease is a rare autosomal recessive disease, characterized by degeneration of white matter of the central nervous system. It is a leukodystrophy that usually occurs in early childhood and is characterized by megalocephaly, demyelination and multiple Rosenthal fibres. Specific magnetic resonance imaging findings and gene mapping are essential to diagnose the disorder. The patient mostly presents with frontal predominance; megalocephaly, seizures, spasticity, psychomotor deterioration, excessive vomiting, difficulty in swallowing and speaking, poor coordination, and loss of motor control, slowly progressive paresis, bulbar signs, brisk reflexes, but often with an intact mental state and are may often be fatal.
This case report deals with the dental Management of a 10years old child brought to the out-patient department of paediatric and preventive dentistry, Guru Nanak Institute of Dental Sciences and Research, with a chief complain of pain and blackish discoloration in his lower front teeth region and frequent bleeding from gums, suffering from Alexander disease.

Keywords: alexander disease, leukodystrophy, dental management

Introduction
Alexander disease is a rare, sporadic, autosomal dominant leukoencephalopathy. It is caused by mutation in the gene for glial fibrillary acidic protein (GFAP) that maps to chromosome 17q21 [1]. According to the National Institute of Neurological Disorders and Stroke, the destruction of white matter is accompanied by the formation of Rosenthal Fibres which are abnormal clumps of protein that accumulate in astrocytes in the brain and it affects the growth and development of myelin sheath [2]. It is of 4 types based on age at clinical presentation: neonatal, infantile, juvenile, and adult [3], the most common being the infantile form that usually begins during the first 2 years of life and the least being the adult type. Infantile onset Alexander disease is characterized by frontal predominance; megalocephaly, seizures, spasticity and psychomotor deterioration. These children may have excessive vomiting, difficulty in swallowing and speaking, poor coordination, and loss of motor control, slowly progressive paresis, bulbar signs, brisk reflexes, but often with an intact mental state and are may often be fatal.The disease occurs in both males and females, and no ethnic, racial, geographic or cultural/economic differences are noted. No cure or treatment procedure is known yet and the prognosis is poor [4]. With early onset, death usually occurs within 10 years from the onset of symptoms. Here we present a case report of dental management of a 10 years old male child with juvenile Alexander disease.

Case Report
A 10 years old male child was brought to the OPD of Department of Paediatric and Preventive Dentistry, Guru Nanak Institute of Dental Sciences and Research, with a chief complain of pain and blackish discoloration in his lower front teeth region and frequent bleeding from gums for the past 1 year, as reported by the mother. Bleeding usually occurs while cleaning the gums, accompanied with profuse redness and swelling of the gums and have intensified over the past 2 months.
The patient's parents gave history of delayed developmental milestones of their son. There was no history of consanguineous marriage of the parents. They reported that this is their only son and he had standard birth weight and greater head circumference (macrocephaly) at birth. There was history of seizures 5 months after birth accompanied with recurrent chest infections. This was followed by gradual cognitive abnormalities and psychomotor deficiencies, for which they had taken paediatrician's consultation. However, he was put on phenytoin oral suspension twice daily, which controlled the symptoms for the time being. Since the past 4 years there has been increasing feeding difficulties, accompanied by vomiting and exacerbation of the symptoms for which he had undergone percutaneous endoscopic gastrostomy. Medical reports, MRI and gene mapping, confirmed genetic mutation of GFAP gene (Alexander disease).
On general physical examination, gait and postural abnormality was noted (he could not walk and was brought on wheel chair). The patient had inability to speak, understand, respond and presented with social skill deficiency. The head circumference slightly greater than the normal limits and the occipital region appeared to

be flattened (Fig 1).
On intraoral examination, 31 and 41 were found to be mobile (Grade II) and carious. 65, 75, 36, 84, 85 were also found to be carious. There was diffuse swelling and erythema of the marginal and attached gingiva with profuse bleeding on probing (Fig 2). He also presented with high arched, constricted palate, fissured tongue and dried, chapped lips with multiple bleeding spots. The patient had to undergo extraction of 31 and 41 regarding the same. Hence Local anaesthetic sensitivity testing and haematological investigations were done, where the parameters turned out to be normal. Following the same, mouth prop was placed and high-volume suction tip was held in mouth and type II GIC restoration was done wrt 65, 75, 36, 84, 85. (Fig 3) Extraction was done wrt 31 and 41 under local anaesthesia (Fig 4). Topical fluoride application was done after haemostasis was achieved. No post operative complications were reported.
The mother was also instructed to apply petroleum jelly on the child's lips frequently and topical application of choline salicylate + lignocaine (Dologel CT) over the affected gingiva 3 – 4 times daily for 1 month. Paracetamol 250mg syrup (5ml) was adviced twice daily for 3 days to control post operative pain. The patient was recalled after 1 month for follow up check-up.

Discussion
Stewart Alexander (1949) first described this leukodystrophy as progressive degeneration of fibrillary astrocytes, after which several subtypes have been identified [3]. Neonatal form is recognized usually after birth, infantile form within two years of age, juvenile form within four to fourteen years of age and adult after eighteen years of age. There is no definite treatment. Supportive therapy includes general care and nutritional supplementation, antibiotic treatment for intercurrent infection, antiepileptic drugs for seizure control, assessment of learning disabilities and cognitive impairment, and physical and occupational therapy if needed [5]. In the present case, events of seizures (status epilepticus) started 5 months after birth which was accompanied by progressive generalized weakness, psychomotor deficiencies, seizures, and developmental delays. The head circumference was slightly greater than normal and the occipital region appeared to be flat.
The differential diagnosis of AD are disorders that present with macrocephaly and/or cerebral white matter changes; adrenoleukodystrophy, Canavan disease, megalencephalic leukoencephalopathy with subcortical cysts, metachromatic leukodystrophy and multiple sclerosis [6]. Conclusive diagnosis regarding the present case was already brought by MRI and genetic mapping (Fig 5, 6) [7].
Dentists, especially pediatric dental practitioners play a crucial role in case of Infantile AD as these children often present with dental caries, hypoplasia, gingival and periodontal pathologies, medication-induced gingival hyperplasia, and delayed healing. Seizure episodes in AD may also cause trauma to the tongue, mucosa thereby resulting in bleeding. Parents should be educated about the medical condition, oral hygiene maintenance instructions and informed about the treatment plan. Mouth-guards should be used in those children with uncontrolled epilepsy. For those taking food orally, cariogenic foods and beverages should be avoided. Topical fluorides and sealant application should be recommended in these patients, sugar-free medications should be preferred and fluoridated toothpastes should be used daily [8].

Conclusion
Early diagnosis is essential for Alexander disease as treatment is only symptomatic and supportive, including attention to general care and nutritional requirements, antibiotic treatment for intercurrent infection, antiepileptic drugs to control seizure, assessment of cognitive impairment and learning disabilities, and physical therapy as needed.
The oral health care in these children should be more focussed towards a preventive approach rather than definitive treatment as management of such patient in OPD becomes difficult.

Figures

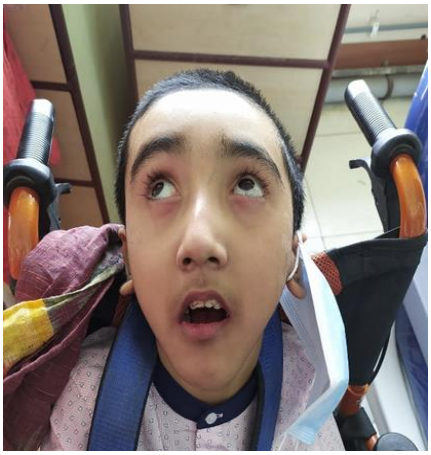

Fig 1: Extraoral features

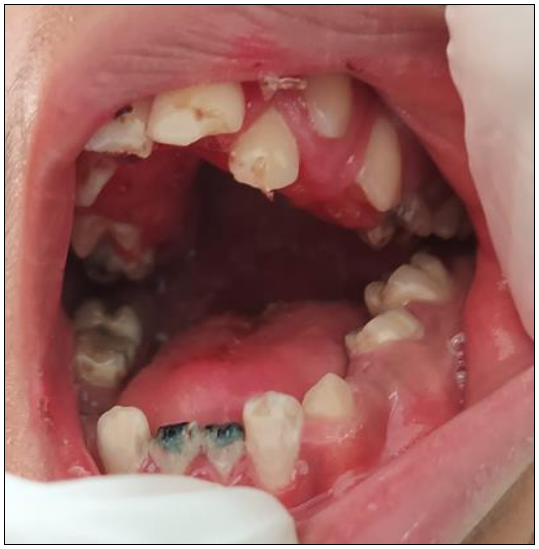

Fig 2: Intraoral features

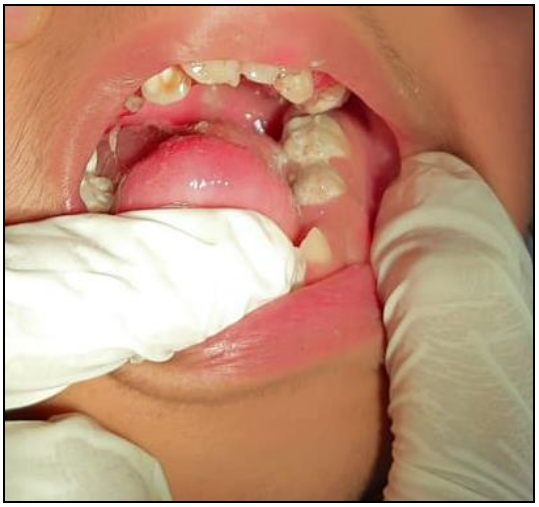

Fig 3: GIC restoration wrt 74, 36

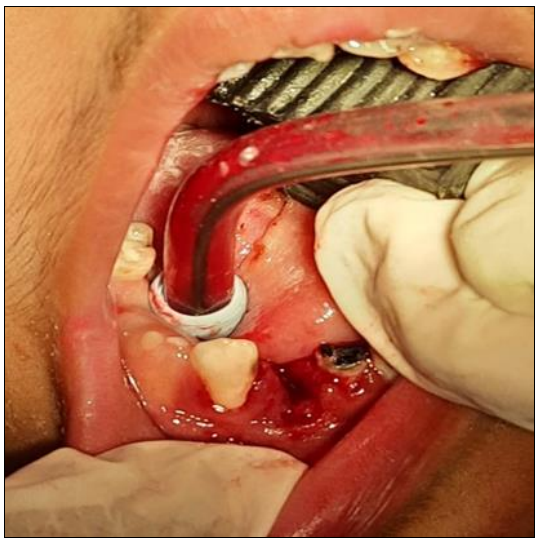

Fig 4: Extraction wrt 41

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