Case Report

Prosthetic rehabilitation of a child affected with Albright’s hereditary osteodystrophy: a case report

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ABSTRACT

Albright’s hereditary osteodystrophy is a rare metabolic disease resulting from an inability of the kidneys and bones to respond appropriately to circulating parathyroid hormone (PTH), the principal regulator of calcium and phosphorous levels in the body. The deranged parathyroid hormone levels along with the disturbed metabolism of calcium and phosphorous significantly affects the development and calcification of the dental hard tissues along with the bone matrix. Oral manifestations include multiple impacted and aplastic teeth, tooth hypoplasias, small crowns with blunt roots, thin enamel with large pulp chambers, malocclusion and premature loss of teeth resulting from caries. The present case report highlights the essential dental findings of the child affected with Albright’s osteodystrophy along with highlighting the complete rehabilitation of the case using a combination of maxillary overdenture and mandibular partial prosthesis.

Keywords: Pseudohypoparathyroidism, Hypoplasia, Missing teeth

INTRODUCTION

Albright’s hereditary osteodystrophy (AHO) also referred to as pseudohypoparathyroidism (PHP) is a rare hypocalcemic disorder which was first described by Fuller Albright in 1942.¹,² The condition itself is a rare one with a prevalence of 0.7 in 100 and results from mutation of GNAS1 protein mapped to q13.11 region of chromosome 20.³ The result is an end organ resistance of the kidneys and bones to the peripheral action of parathyroid hormone (PTH). As a result, the parathyroid hormone is secreted in excess, with a consequent increase in serum parathyroid levels and hyperplasia of the parathyroid glands. The kidneys however fail to respond to the elevated circulating levels of PTH, resulting in hypocalcaemia and hypophosphatemia.⁴,⁵ Systemic manifestations of AHO include short stature, thick short necks, round faces with greasy skin, considerably reduced length of the 3rd and 4th fingers and toes along with dimpling of the corresponding knuckles.⁶⁻⁸ Epiphyseal closure in advance of age and occasional patches of soft tissue calcifications may be seen. Basal ganglia calcifications, frontal bossing and thick calvarial lining referred to as "hyperostosis frontalis interna" have also been reported.⁹⁻¹¹ Oral manifestations are common and seen in more than one-third of cases with hypoparathyroid conditions.¹⁰ The manifestations mimic those of idiopathic hypoparathyroidism, including multiple impacted and aplastic teeth, hypoplastic pitting, small crowns with blunt roots, thin enamel with large pulp chambers, “dagger-shaped”
pulp stones, malocclusion and premature loss of teeth resulting from caries.\textsuperscript{6,8,10} Jensen et al in an excellent review described the medical and dental findings in eleven patients affected with hypoparathyroidism and pseudohypoparathyroidism. All the reviewed patients presented with tetanic and epileptic manifestations. Likewise dental manifestations were seen in all cases except one with disturbance in tooth eruption, root defects, hypodontia and enamel hypoplasia being observed in more than 2/3rd of the evaluated cases. The authors further highlighted that the above-mentioned disturbances were most severe and frequent in the pseudohypoparathyroid group as compared to hypoparathyroidism.\textsuperscript{19} Like wise Ritchie et al in 1965 described dental findings associated with AHO in 4 patients along with highlighting the histochemical features of the dentine in the form of irregularly calcified matrix, with dentinal tubules exhibiting sharp bends near the pulpal and amelodentinal junctions. The dental findings were summarized as small crowns with thin hypoplastic enamel, short roots with blunt apices, and large pulp chambers with numerous calcified deposits. Dental aplasia and delayed eruption were also common.\textsuperscript{6} Management of AHO varies from rehabilitation of hypoplastic teeth using direct composite or esthetic celluloid crowns, removal of impacted teeth and placement of implants to fabrication of complete dentures or overdentures for missing teeth and functional rehabilitation.\textsuperscript{7} The oral and clinical aspect of the condition has been described by numerous authors however it is the detailed management of AHO on which the literature is sparse.\textsuperscript{11,6,9}

The present case report aims to highlight the prosthetic management a child affected with AHO associated with numerous unerupted teeth using overdenture and mandibular partial denture along with the detailed description of associated dental and medical findings.

**CASE REPORT**

An 11 yr old male child reported to the Unit of Pedodontics and Preventive Dentistry with the chief complaint of numerous missing teeth associated with difficulty in eating and speaking. The child also had concerns with the prominence of lower jaw. On seeking the detailed history it was found that only the anterior primary anterior teeth had erupted and the eruption was significantly delayed with the primary mandibular incisor erupting at 1.5 yrs of age. The eruption of permanent teeth was also significantly delayed with the permanent mandibular incisors erupting at 9 years of age. Posterior maxillary and mandibular teeth, however had failed to erupt both in the primary and the permanent dentition. Past medical history evaluation revealed that the child was born of a non-consanguineous marriage and at about 2 months of age had recurrent episodes of seizures which were attributed to the low level of serum calcium. At the same time, serum phosphorous and PTH levels were also severely elevated with normal Vitamin D levels. Initial biochemical investigations revealed severe hypocalcaemia (5.3 mg/dL, normal 9-11 mg/dL), elevated phosphorous levels (11.50 mg/dL, normal 4.50-6.70 mg/dL) along with high serum levels of parathormone (372 pg/ml, normal 10-69 pg/ml). Bone mineral density (BMD) studies revealed generalized osteopenia and clear osteoporosis. Furthermore, radiographic examination revealed bilateral hypoplastic 4th phalanges along with deformity of the left ring and index finger which is a consistent feature of Albright’s hereditary osteodystrophy. The intellectual development of the child was however normal. A detailed work-up was then carried out and was final diagnosis of AHO was established. Since then the focus of medical therapy has been on maintaining optimum serum levels of calcium and phosphorous with daily Calcium (1500 mg/day) and vitamin D supplements (50 micro gm/day). Along with that CT scans are done to estimate the extent of calvarial and basal ganglion calcification.

Oral examination of the patient revealed various partially erupted and hypoplastic permanent maxillary incisors along with a partially erupted maxillary right molar which were carious (Figure 1a-e). The gingival mucosa corresponding to the mandibular incisors was inflamed (Figure 1a). Dental cross-bite was also seen corresponding to the right maxillary and mandibular side along with a developing skeletal Class III malocclusion (Figure 1a).

![Figure 1: (a) Pre-operative intra-oral frontal view revealing multiple missing permanent teeth along with hypoplastic maxillary incisors and complete anterior crossbite; (b) Maxillary occlusal view revealing partially erupted carious first permanent molar; (c) Mandibular occlusal view (d, e) Right and left Lateral views.](image-url)
Panoramic view revealed presence of multiple impacted primary and permanent teeth especially in the posterior maxillary and mandibular region (Figure 3a). None of the teeth from the permanent series was congenitally missing. The teeth were however impacted with varying thickness of overlying bone. The roots were short and blunt with widened pulp chambers. Along with the panoramic view a lateral cephalogram also was taken to assess the maxillo-mandibular relation with the cranial base, revealing a vertical growth pattern of the child (Figure 3b). Baseline models were also recorded.

Prior to making the impression for denture fabrication, the partially erupted molar was restored using preventive resin restoration employing a microfilled resin and oral hygiene measures were instituted especially corresponding to the inflamed mandibular mucosa. This was followed by making of the primary impressions using alginate (Mariflex™ Type II Normal setting Alginate, Septodont, France) and fabrication of a special tray. All the peripheral borders of the denture base were moulded using a functional technique followed by a final impression to adequately register the tissue details. Bite was registered with an approximate clearance of 3mm between the anterior segments so as to provide adequate space between the denture bases for free growth of the maxilla. Once teeth setting were complete, a final trial was taken. The final prosthesis was processed using heat activated poly methyl meth acrylate followed by finishing, polishing and insertion. For additional retention and stability, stainless steel wire clasps was provided on the incisor. Post insertion instructions and oral hygiene instructions were provided both to the patients and the parents. They were also told about the need for frequent adjustments that would be necessary once the permanent teeth start erupting.

Post the rehabilitation the recall was done initially at 1 week. This was followed by a recall at 1 month, 3 months and every 6 months. At every recall appointment other than the denture base adjustment special attention was given to the partially erupted maxillary molar. At one year of follow-up the patient showed excellent compliance with the prosthesis with adequate retention (Figure 2 a-c). Improvement in speech was also noticed. The parents reported improvement in masticatory efficiency with preference changing from soft consistency food to that with more fibrous consistency.

**DISCUSSION**

Albright’s hereditary osteodystrophy is a rare metabolic disease resulting from a disturbance in the peripheral action of parathormone on the end organs, namely kidney and the bones. The end result is an inability of the body to respond appropriately to circulating parathyroid hormone (PTH), the principal hormonal regulator of the developing class III malocclusion and placement of implants. The placement of implants was ruled out due to the presence of multiple impacted teeth (Figure 3a). Furthermore, owing to the lack of adequate anchorage it was decided to provide adequate clearance between the maxillary and mandibular denture bases such that the maxilla is free to continue with its natural growth. Extraction of impacted primary teeth was ruled out to prevent undue harm to the permanent teeth and to maintain the alveolar ridge height. It was decided to fabricate a maxillary overdenture, extending over the right maxillary molar after providing adequate clearance for tooth eruption to aid in retention with a mandibular partial denture.
calcium and phosphorous levels in the body. \textsuperscript{1,4,11} Clinically the disease is similar to idiopathic hypoparathyroidism which is also characterized by a deficiency of circulating parathyroid hormone resulting in low serum calcium and high phosphorus concentrations. As the serum calcium and phosphorous levels are intricately associated with the development and calcification of dental hard tissues significant oral manifestations are seen associated with AHO in the form of enamel hypoplasias, pitting and discoloured enamel, small crowns and short blunted roots, enlarged pulp chambers with multiple pulp stones, thin enamel, multiple impacted and missing teeth, congenitally missing premolars, short wide jaws with propensity for malocclusions and early loss of teeth due to caries. \textsuperscript{4,6,11}

Traditionally dental management has been in the form of rehabilitation of the existing dentition using stainless steel and celluloid crowns along with replacement of missing teeth with either complete dentures or overdenture. Extraction of impacted teeth may be done but is often associated with significant morbidity. Placements of implants are indicated in cases with congenitally missing teeth but may not serve as a feasible option in cases where permanent teeth are impacted. Malocclusions are frequently encountered in cases with AHO owing to disproportionate jaw size. Management varies from surgical intervention for the malocclusion to interceptive procedures based on the severity of malocclusion and age of the child. \textsuperscript{7,9,12}

Early prosthetic rehabilitation for cases of AHO is often desirable from the child’s functional, esthetic and psychological point of view. Conventional prosthodontic rehabilitation in young patients is however challenging owing to the anatomical abnormalities of the mixed dentition, minimal denture bearing area, along with the compliance issues with the lengthy chair side procedures associated with denture fabrication. Moreover, in young patients, denture bases should lateral growth of the jaw for which special measures have to be taken into consideration in the form of incorporation of expansion screws to provision of flexible dentures. \textsuperscript{13}

For the present case a combination of maxillary overdenture with a mandibular partial denture was chosen. As the child had numerous impacted primary and permanent teeth in the posterior segment implant placement was not considered feasible. For the malocclusion in the form of anterior dental cross bite, no corrective appliance or expansion screw could however be incorporated owing to the lack of adequate retentive surfaces and anchorage units. The orthodontist however suggested anterior incisal clearance to allow for maxillary growth which was incorporated into the prosthesis. The present approach allowed for a normal physiological development, development of correct masticatory function which was evident in terms of improvement in quality of food intake along with improved esthetics and phonetics. All these factors ultimately contributed to the enhanced social acceptance and self-worth along with ensuring better psychological development which is especially important for young growing patients affected with AHO.

**CONCLUSION**

Albright’s hereditary osteodystrophy is a rare developmental disorder with severe oro-facial manifestations. Successful management requires a multidisciplinary approach inclusive of a paediatric dentist, orthodontist, prosthodontist and an oral-maxillofacial surgeon. The goal should be improvement in overall patient’s health in terms of masticatory and speech efficiency along with increase in self-confidence.

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**REFERENCES**
