Oral Rehabilitation of Pseudohypoparathyroidism: Case Report

PSÖDOHİPOPARATİROİDİZMLİ BİR HASTADA ORAL REHABİLİTASYON

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Abstract

Objective: Hypoparathyroidism is an endocrine based metabolic disorder and characterized by low serum levels of parathormone and hypocalcaemia. However, pseudohypoparathyroidism is characterized by high serum levels of parathormone and hypocalcaemia. In this study, we report a 20-year-old Turkish male with pseudohypoparathyroidism and discuss his clinical, radiographic and laboratory test characteristics together with his oral manifestations. We also discuss the prosthetic management of treatment.

Case Report: Clinical examination showed that the patient with pseudohypoparathyroidism had a short stature, round face, low nasal bridge, short neck, z deformity on hands and feet deformity and weakness. Biochemical tests showed hypocalcaemia (8.1 mg/dL) and high serum levels of parathormone (344 pg/ml). Bone mineral density (BMD) studies revealed clear osteopenia (BMD: 0.757) from L1-L4 region, and clear osteoporosis (BMD: 0.507) from total BMD results.

The oral examinations showed limited mouth opening and the maxilla was quite small as compared with the mandible. The dental radiographic evaluations showed pseudoanadontia, malformed dental roots and lack of periodontal spaces. In addition, there were a lot of unerupted teeth in both jaws, except lower left central incisor. We also determined several internal resorptions in unerupted teeth. There were large pulp chambers located primarily in lower molar teeth.

Conclusion: We believe that, for better prosthetic rehabilitation of the patient, orthodontic treatment of unerupted teeth was quite impossible due to lack of enough space. Advanced dental surgery techniques were refused by the patient. We refrained from extraction of unerupted teeth to preserve alveolar ridge levels of the patient. Finally, the patient's masticatory and speech functions and aesthetics were restored with conventional complete dentures.

Key Words: Pseudohypoparathyroidism; tooth, unerupted

Özet

Amaç: Hipoparatiroidizm endokrin kaynaklı metabolik bir hastalıktır; parathormonun serum da düşük seviyede oluşu ve hipokalsemi ile karakterizedir. Psödohipoparatiroidizmde ise parathormonun serum seviyesi yüksektir ve hipokalsemi mevcuttur. Bu çalışmada psödohipoparatiroidizmli 20 yaşında erkek hasta rapor edilmiş ve klinik, radyolojik ve laboratuvar bulguları ağız belirtileri ile birlikte değerlendirilmiştir. Ayrıca tedavinin protetik başarısı da ele alınmıştır.

Olgu Sunumu: Klinik incelemelerde psödohipoparatiroidizmli hastada kısa boy, yuvarlak yüz, basık burun kemiği, el ve ayaklarda z deformiteler ve güçsüzlük tesbit edilmiştir. Biyokimyasal testlerde hipokalsemi (8.1 mg/dl) ve parathormonun serum düzeyinde yükseklik (344 pg/dl) görülmüştür. Kemik mineral yoğunluğu (BMD) çalışmalarında L1 ve L4 bölgesinde osteopeni (BMD: 0.757) ve toplam BMD sonucunda ise osteoporozis (BMD: 0.507) ortaya çıkmıştır.

Oral incelemede ağız açıklığının sınırlı olduğu ve maksillanın mandibuladan oldukça küçük şekillendiği görülmüştür. Panoramik radyografta; psödoanodonti, diş köklerinde şekil bozukluğu ve periodontal aralıkta azalma tespit edilmiştir. Ek olarak her iki çenede de alt santral kesiciler haricinde sürmemiş dişler bulunmuştur. Aynı zamanda sürmemiş dişlerde çok sayıda internal rezorpsiyon saptanmış ve alt molar dişlerde geniş pulpa odaları görülmüştür.

Sonuç: Hastanın protetik tedavisinin daha iyi planlanması için, periodontal aralıktaki daralma sebebiyle sürmemiş dişlere ortodontik tedavi uygulanması hemen hemen imkansız görülmüştür. Gelişmiş cerrahi teknikler ise hasta tarafından reddedilmiştir. Alveolar kret seviyesini korumak için sürmemiş dişlerin çekiminden kaçınılmıştır. Sonuç olarak hastanın çiğneme, konuşma ve estetik fonksiyonları konvansiyonel tam protezle düzeltilmiştir.

Anahtar Kelimeler: Psödohipoparatiroidizm, sürmemiş diş

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Hypoparathyroidism is an endocrine based metabolic disorder and characterised by low serum levels of parathormone (PTH) and hypocalcemia. However, pseudohypoparathyroidism (PsHP) is characterized by high serum levels of PTH and hypocalcemia. The phenotypic

characteristics present in PsHP are: short stature, obesity, round face, shortening of the metacarpus and metatarsus, short neck, low nasal bridge and mental retardation.[1-3]

The term PsHP has been used to describe a group of disorders with biochemical and clinical features of hypoparathyroidism but with target organ resistance to PTH as indicated by elevated levels of PTH. In the two major forms of this disorder (PsHP type Ia and PsHP type Ib), correction of hypocalcemia usually results in supression of PTH levels but fails to correct target organ resistance to the hormone.[4]

PsHP is divided into 2 main types. Type I is characterised by the low level or absence of renal CAMP production in response to PTH. Type II responds to PTH with normal increase in urinary CAMP but show absent or subnormal phosphaturic response.[5]

Hypocalcemia usually follows an asymptomatic course and diagnosis depends on recognizing its clinical symptoms. The clinical symptoms of hypocalcemia are commonly neurological. Severe hypocalcemia can cause laryngospasm and generalized convulsion. The most specific syndrome is tetania.[3,6]

The oral manifestations of PsHP found in the literature include aplasia and enamel hypoplasia, late tooth eruption, and enlarged radicular channels susceptible to caries, malformed teeth, presence of severe periodontal disease with gingivitis.[1,7-11]

The main goal of therapy in PsHP patients is to restore serum calcium and phosphorus. These patients require regular monitoring of serum calcium, phosphorus and renal function tests.[12]

Case Report

A 20-year-old male patient was referred with history of thigh and neck contraction, tingling in the ear ear, lack of hearing and anadontia to Gülhane Military Medical Academy (GMMA) Department of Endocrinology. The patient was diagnosed with PsHP type Ib.

Physical examination showed: stature of 160 cm; weight of 44 kg. Biochemical tests showed hypocalcemia (8.1 mg/dL) and high serum levels of PTH (344 pg/ml). On thyroid sonography, diffuse and hyperplasic thyroid gland was found.

Clinical examinations revealed that the patient had a short stature, round face, low nasal bridge and short neck. The patient had deformity in hands and foots deformity and weakness, in contrast to the obesity which is usually seen in PsHP.

Radiographic findings showed bilateral long 1st, 3rd and 4th metacarpus and also bilateral long 2nd metatarsus (Figure 1, 2). Bone mineral density (BMD) studies revealed clear osteopenic (BMD: 0.757) in L1-L4 region and clear osteoporotic (BMD: 0.507) in total BMD data.

The patient was sent to GMMA Center of Dental Sciences, Department of Oral Diagnosis and Radiology in order to be examined for unerupted teeth and difficulty in chewing. Intraoral and radiographic examination (panoramic radiographs) were made for dental findings. The oral

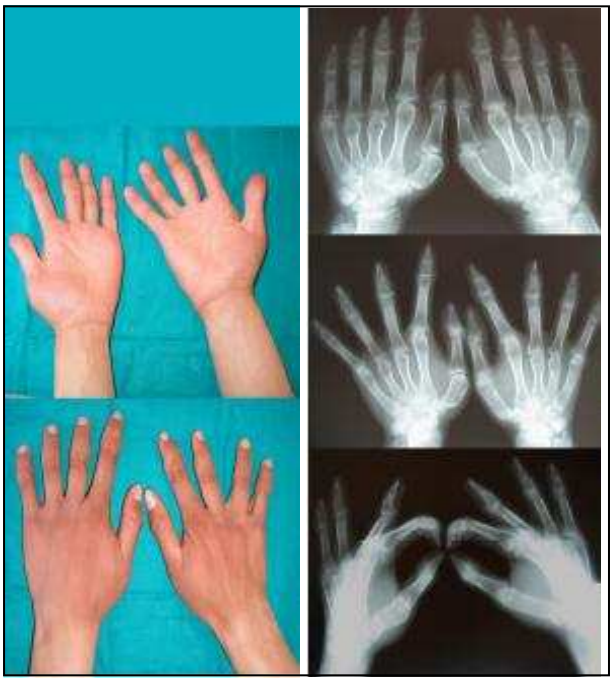

Figure 1. **a-** Patient's hands clinical view.
b- Patient's hands appearance in radiography.

Figure 2. **a-** Patient's feet clinical view.
b- Patient's feet appearance in radiography.

Figure 3. Intraoral view of the patient.

examinations showed limited mouth opening and maxilla was quite small compared to the mandible (Figure 3).

The dental radiographic evaluations showed pseudoanadontia, malformed dental roots and lack of periodontal spaces (Figure 4). In addition, there were a lot of unerupted teeth in both jaws, except lower left central incisor. We also determined several internal resorptions in unerupted deciduous teeth. There were large pulp chambers located primarily in lower molar teeth. The patient's masticatory and speech functions and aesthetics were restored with conventional complete dentures in GMMA Centre of Dental Sciences, Department of Prosthetic Dentistry (Figure 5, 6).

Figure 4. Panoramic radiograph showing pseudoanadontia, malformed dental roots and lack of periodontal spaces.

Figure 5. Frontal and sagittal view of the patient's face contour seen before treatment and after treatment.

Figure 6. **a-** Frontal and sagittal view of the old prosthesis before treatment.
b- The clinical view of the patient with new prosthesis after treatment.

Discussion

Many inherited disorders have oral manifestations which can be detected on dental radiographs as alterations in the morphology or chemical composition of the teeth. Thus, the dentist may be the first to detect disorders of development and metabolism of importance to the general health of the patient and his family.[7]

While the metabolic and genetic disorders are usually established by endocrinologists and medical geneticists, oral symptoms have to be taken into consideration by dental professions. With simple conservative and prosthetic treatments, patient's life comfort can be dramatically increased. Eleven patients with idiopathic hypoparathyroidism and PsHP were examined orally and medically by Jensen[10] 1981. They have demonstrated dental anomalies in all patients but one. Enamel hypoplasia was observed in 6 cases, disturbances in tooth eruption in 8, root defects in 5 and hypodontia in 7. Dental anomalies were more frequent than expected from the literature, probably because the dental aspects of hypoparathyroid disease often have been overlooked. In the present material, the above-mentioned disturbances were most severe and frequent in the pseudohypoparathyroid group. Hypoparathyroid conditions are highly invalidating, easily accessible to treatment, but often undiagnosed for years.[10]

In Lagarde's study,[8] 2 premolars removed from a 14 year old girl, with suspicion of PsHP have been studied, using several of the correlated techniques applied to the study of calcified tissues (light microscopy, microradiography and scanning electron microscopy). Enamel gross and micro hypoplasia, hypomineralization of the enamel surface and dentin hypocalcification were similar to dental abnormalities observed in PsHP. Microradiography showed for the first time calcifications present in blood vessels of the dental pulp. Light microscopy and SEM revealed dystrophic globular calcifications within enamel hypoplastic pits.[8]

In the case reported above, we determined pseudoanadontia, malformed dental roots, lack of periodontal spaces, internal resorptions in unerupted deciduous teeth and large pulp chambers located primarily in lower molar teeth.

We believe that, for better prosthetic rehabilitation of the patient, orthodontic treatment of unerupted teeth was quite impossible due to lack of enough space. Advanced dental surgery techniques were refused by the patient. We refrained from extraction of unerupted teeth to preserve alveolar ridge levels of the patient. Finally, patient's masticatory and speech functions and aesthetics are restored with conventional complete dentures. It was observed that, after one year of follow up for three-month periods, the patient was adapted to his dentures well both esthetically and functionally. Today, dental implants are common treatment alternatives in rehabilitation of lost teeth and anadontia. However, dental implantology to prevent with several problems, such as systemic disorders (PsHP) and economic factors.

REFERENCES

1. Gomes MF, Camargo AM, Sampaio TA, Graziozi MA, Armond MC: Oral manifestations of Albright Hereditary Osteodystrophy: A Case Report. Rev Hosp Clin Fac Med Sao Paulo 57: 161, 2002
2. Lavin N: Manual of Endocrinology and Metabolism.Third Edition. Philadelphia, Lippincott Williams & Wilkins, 2002, p.316-9
3. Berkow R, Fletcher A: The Merck Manual of Diagnosis and Theraphy. Rahway, Division of Merck &Co., Inc., N.J., 1992, p. 1005-7
4. Bastepe M, Frohlich LF, Hendy GN, Indridason OS, Josse RG, Koshiyama H, et al: Autosomal dominant pseudohypoparathyroidism type Ib is associated with a heterozygous microdeletion that likely disrupts a putative imprinting control element of GNAS. J Clin Invest 112:1255, 2003
5. Vasicek TJ, McDevitt BE, Freeman MW, Fennick BJ, Hendy G, Potts JT Jr, et al: Nucleotide sequence of the human PTH gene. Proc Natl Acad Sci USA 80:2127, 1983
6. White IL: Hyperparathyroidism, hypercalcemia and the otolaryngology head and neck surgeon. Laryngoscope 94: 742, 1984
7. Witkop CJ: Clinical aspects of dental anomalies. Int Dent J 26:378, 1976
8. Lagarde A, Kerebel LM, Kerebel B: Structural and ultrastructural study of the teeth in a suspected case of pseudohypoparathyroidism. J Biol Buccale 17: 109, 1989
9. Jones KL: Smith's Recognizable Patterns of Human Malformation. 4th ed. Philadelphia, WB Saunders Company, 1988, p. 394
10. Jensen SB, Illum F, Dupont E: Nature and frequency of dental changes in idiopathic hypoparathyroidism and pseudohypoparathyroidism. Scand J Dent Res 89:26, 1981
11. Brown MD, Aaron G: Pseudohypoparathyroidism: Case report. Pediatr Dent 13:106, 1991
12. Sajitha S, Krishnamoorthy PN, Shenoy UV: Psedohypoparathyroidism in newborn-a rare presentation. Indian Pediatr 40:47, 2003