İDİYOPATİK HİPOPARATİROİDİZM VE EPİLEPSİ

IDIOPATHIC HYPOPARATHYROIDISM AND EPILEPSY

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Özet

Bu yazıda epilepsi tanısı ile takip edilmekte ve difenilhidantoin tedavisi almakta iken hipokalsemi, çok düşük parathormon düzeyleri ve bilgisayarlı tomografide bazal gangliyonlarda bilateral simetrik kalsifikasyonların saptandığı bir idiyopatik hipoparatiroidizm vakası sunulmuş ve idiyopatik hipoparatiroidizmin etyopatogenezi, klinik görünüm ve tedavisi literatür bilgileri eşliğinde gözden geçirilmiştir.

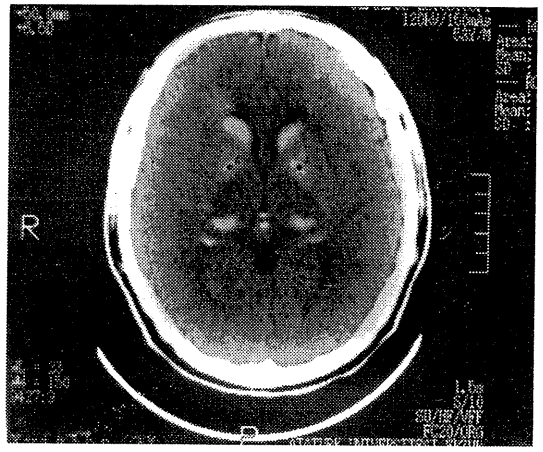
Anahtar kelimeler: *İdiyopatik hipoparatiroidizm, Epilepsi, Difenilhidantoin*

Summary

In this paper, one case of idiopathic hypoparathyroidism with the initial diagnosis of epilepsy under diphenylhydantoin treatment had hypocalcemia, very low parathyroid hormone level, bilateral symmetric calcification in the basal ganglia on the computerized tomography was presented and clinical presentation and treatment of idiopathic hypoparathyroidism related to recent literature were reviewed.

Key words: *Idiopathic hypoparathyroidism, Epilepsy, Diphenylhydantoin*

Figure 1. *Calcifications in the Basal Ganglions are Seen Bilaterally on the Brain CT*



Introduction

Hypoparathyroidism (HP) generally results from decrease in either the secretion of parathyroid hormone (PTH) or its peripheral effect. Tetany, cataract, neuropsychiatric disorders, epilepsy and congestive heart failure are the most frequently seen clinical signs and hypocalcemia is the cause of many of these symtoms (1). Idiopathic HP (IHP) is a rare disorder that can be seen either as familial or sporadic. A variety of clinical forms has been observed in patient's with IHP. Commonly, intracranial calcification and neuropsychiatric symptoms and signs occur together. Convulsions and tetany being more common, extrapyramidal and cerebellar manifestations, emotional lability, memory disorders, mental retardation, organic brain syndrome and functional psychosis are the neuropsychiatric disorders frequently found (2-4). Fahr syndrome is a clinical picture with bilateral calcifications in the striopallidodentate system, progressive dementia, convulsions and extrapyramidal manifestations and it has been reported to have been observed in 70-80 % of patients with HP (4). Etiology is not known.

Case

A 43 year old male patient with the diagnosis of epilepsy was admitted to our clinic. It was understood that the patient has had epilepsy for twenty years, has been regularly using 300 mg/day of phenytoin orally, for the past 5 years and has suffered 5 generalised tonic clonic type epileptic seizures in the past 2 days. In the evaluation of the patient, he was in good clinical condition, conscious, oriented and cooperative. Bilaterally, Chovastek's sign was noted to be positive. The Trousseau sign on the other hand was negative. No other pathologic sign was found. Calcifications in the basal ganglions were seen bilaterally on the computed tomography (CT) (figure 1). The obtained serum levels of calcium: 5.1 mg/dl (8.5-11.5 mg/dl), phosphate: 8.3 mg/dl (2-5 mg/dl), PTH: <1.00 pg/ml (12.0-72.0 pg/ml) and magnesium: 1.5 mEq/L (1.3-2.1 mEq/L) were noted. The other routine biochemical tests, hemogram, urogram, thyroid function tests, electrocardiogram, chest radiogram were evaluated to be normal. In the electroencephalogram, apart from slowing in the theta frequencies here and there, no pathology was found. The patient was diagnosed to have IHP and a antiepileptic treatment was stopped and treatment with 1,25 (OH)2 D3 and calcium was started. Also anti-edema treatment was administered for 3 days. During the patient's stay in our clinic, the serum calcium levels increased gradually and the serum level on the 19th day of treatment was 8.3 mg/dL. The patient suffered no seizure during his stay in our clinic. The patient was discharged with 1,0 mcg/d calcitriol and 1000 mg/d ionised calcium orally. Also the other members of the family underwent a brain CT, serum PTH and serum calcium levels and no pathology was found in either of them.

Discussion

HP is characterised by hypocalcemia and immeasurably low serum PTH levels (1,3). It was seen on the brain CT that bilateral symmetric calcifications were primarily seen the basal ganglia, the dentate nucleus of the cerebellum and the white matter of cerebrum and cerebellum. The globus pallidus in the basal ganglions are the most affected (5). The mechanism of the development of calcification is not known. It is postulated that local factors play an important role in the mechanism of calcification (4). Morgante et al (4) have reported 3 cases of Fahr Syndrome which had suffered meningoencephalitis in childhood. During their hospitalization, one patient died because of generalized status epilepticus. In the autopsy examination of the patient, it was found that there were generalised non-arteriosclerotic perivascular calcifications, especially in the basal ganglions. Therefore, the previous disease thought to have brought about the gliovascular changes have been postulated to have made easy the deposition of calcium in the basal ganglions. It has been brought to

attention that since the period between the inflammatory process and the neurologic manifestation is long, frequently, this inflammatory process can be overlooked. However, it has not been proven whether the gliovascular changes in the basal ganglions are a result of cerebral calcifications. We could not identify an inflammatory period in our patient. Duran et al (6) proposed that the hypomagnesemia is responsible for the decrease in the synthesis and secretion of PTH. We observed that the serum magnesium levels in our patient to be within normal limits. Huddle and Ally (1) followed up 11 patients with IHP for a period of 7 years and reported that of these 11 patients, 4 had epilesy. They brought to attention that in some patients, the symptoms may begin years before the diagnosis of IHP is made and that in patients with epilepsy, phenytoin treatment can suppress the hypocalcemic symptoms. It is thought that epilepsy in patients with IHP is the result of increased neuronal irritability due to hypocalcemia (1). The other causes that should be taken into attention in the differential diagnosis of bilateral symmetric calcifications include HP, pseudo HP, hyperparathyroidism, radiotherapy, intrathecal methotrexate and cytosine arabinoside with radiotherapy, Kearns-sayre syndrome, cockayne syndrome, lipoid proteinosis, mitochondrial myopathy, lactic asidemia, Down's syndrome, hydrotic ectodermal dyplasia and mental retardation, dyskeratosis congenita and post encephalitic-like disease (5). IHP by autosomal domimant transmission has been reported in a lot of cases. Calcifications in the basal ganglions in asymptomatic patients is known (7). Therefore, the families of patients found to have IHP should be screened. Treatment in the early phases of IHP with 1-α-hydroxy vit D3 and calcium has been reported to protect against most of the long term complications of hypocalcemia and also reverse some of the already present complications (1,3). Kistler (8) report ed that the calcifications in IHP, are not a rarely seen condition, however, due to its appearance in a variety of clinical pictures has been proposed as the cause of its being frequently overlooked. Finally, the appearance of symptoms of IHP years before diagnosis and treatment with phenytoin-like anti-convulsive drugs which suppress hypocalcemic symptoms, especially in patients with isolated epileptic manifestations makes IHP diagnosis difficult. Therefore, IHP should be suspected in all patients diagnosed with epilepsy and follow up all those patients diagnosed as having epilepsy by periodic measurements of serum calcium levels and brain CT. It shouldn't be forgetten that early diagnosis and treatment, protects against the late complications of hypocalcemia.

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