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A case of pseudotumor cerebri associated with idiopathic hypopara-thyroidism and vitamin B12 deficiency

İdiopatik hipoparatiroidizm ve B12 eksikliğinin eşlik ettiği bir psödotümör serebri vakası

Pınar AKGÜL[1] Müge ÖZSAN[2] Remzi YİĞİTER[3] Mustafa YILMAZ[4] Aylin AKÇALI[3] Münife NEYAL[3] Mehmet Ali ELÇİ[5] Sırma GEYİK[3]

[1]Dr. Ersin Arslan State Hospital, Clinic of Neurology, Gaziantep, Turkey
[2]Niğde State Hospital, Clinic of Endocrinology, Niğde, Turkey
[3]Gaziantep University Faculty of Medicine, Department of Neurology, Gaziantep, Turkey
[4]Harran University Faculty of Medicine, Department of Neurology, Şanlıurfa, Turkey
[5]Hatay Kırıkhan State Hospital, Clinic of Neurology, Hatay, Turkey

Abstract

Idiopathic hypoparathyroidism is a heterogeneous disorder rarely involved in the etiology of pseudotumor cerebri, and characterized by low levels of parathormone accompanying hypocalcemia, hyperphosphatemia and vitamin D deficiency. A 38-year-old female patient was admitted to our clinic with the complaints of headache and blurred vision. Her neurological examination revealed bilateral papilledema which was more prominent in the left eye. Cranial magnetic resonance imaging findings of the patient were within normal ranges and cerebrospinal fluid (CSF) pressure was 270 mmH_2O which was measured through a lumbar puncture (LP), then the patient was diagnosed with pseudotumor cerebri. Laboratory assessment revealed low parathormone (PTH) and vitamin D levels accompanying hypocalcemia and hyperphosphatemia; and the patient was diagnosed with idiopathic hypoparathyroidism. In her follow-up, no decline was observed in CSF pressure and in the repeated laboratory assessments, a decrease in the vitamin B12 level was observed, thus vitamin B12 supplementation was added to her treatment. In control LPs, CSF pressure (190 mmH_2O) decreased as the blood calcium (Ca) and vitamin B12 levels increased.

Keywords: Pseudotumor cerebri, vitamin B12 deficiency, idiopathic hypoparathyroidism.

Öz

İdiopatik hipoparatroidizm, düşük parathormon seviyesi ile birlikte hipokalsemi, hiperfosfatemi ve D vitamini eksikliğinin görüldüğü ve nadiren psödötümör serebri etyolojisinde yer alan heterojen bir hastalıktır. Otuz sekiz yaşında kadın hasta kliniğimize baş ağrısı ve bulanık görme şikayeti ile başvurdu. Yapılan nörolojik muayenede solda daha belirgin olmak üzere bilateral papil ödemi mevcuttu. Hastanın kranial görüntülemesi normal sınırlarda olup yapılan lomber ponksiyonda (LP) beyin omurilik sıvı (BOS) basıncı 270 mmH_2O olarak ölçüldü ve psödötümör serebri tanısı konuldu. Hastanın laboratuvar tetkiklerinde; düşük parathormon (PTH) ve vitamin D düzeyi ile birlikte hipokalsemi, hiperfosfatemi saptandı ve idiopatik hipoparatrodizm tanısı konularak tedavisi düzenlendi. Takiplerde BOS basıncının düşmediği gözlendi ve tekrarlanan laboratuvar tetkiklerinde vitamin B12 düzeyinde düşme olduğu saptanarak tedaviye vitamin B12 eklendi. Yapılan kontrol LP'lerde hastanın kan kalsiyum ve vitamin B12 düzeyindeki artışa paralel olarak BOS basıncının düştüğü (190 mmH_2O) gözlendi.

Anahtar Sözcükler: *Psödotümör serebri, vitamin B12 eksikliği, idiopatik hipoparatiroidizm.*

Introduction

Idiopathic hypoparathyroidism is a heterogeneous group of disorders characterized by low parathormone (PTH) levels accompanied by hypocalcemia and hyperphosphatemia, and rarely involved in the etiology of pseudotumor cerebri (1).

Corresponding Author: Mehmet Ali ELÇİ
Gaziantep University Faculty of Medicine, Department of Neurology, Gaziantep, Turkey

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Pseudotumor cerebri is a syndrome characterized by an increase in the intracranial pressure and occurs without evidence of intracranial mass lesion or hydrocephalus (2-4). This syndrome was first described by Quincke in 1897 and named as "serious meningitis" (4). The etiology of this syndrome is not well-known (3-5). Some factors have been suggested to trigger the idiopathic pseudotumor cerebri or even involved in its etiology. The most common factors are obesity, pregnancy, menstrual disorders, Addison's disease, hyper- or hypovitaminosis A steroid treatment or its cessation, tetracycline

treatment, hypoparathyroidism, and iron deficiency anemia (2,6-7). Moreover, vitamin B12 deficiency has also been reported to cause pseudotumor cerebri in the literature (8). In the present case report, we described a clinical picture of pseudotumor cerebri caused by vitamin B12 deficiency and idiopathic hypoparathyroidism and vitamin D deficiency.

Case Report

A 38-year-old female patient was admitted with the complaints of headache started a month ago and worsened gradually and blurred vision in the left eye developed 15 days ago. The compressive pain was located in the right half of the head and partially responsive to pain killers. Mild nausea was accompanying the headache but no vomiting, photophobia or phonophobia were present. In neurological examination, bilateral visual acuity was 20/100, and confrontational visual fields were normal and in ophthalmologic examination, bilateral papilledema was detected which was more prominent in the left eye. Other physical and neurological examination findings of the patient were normal; as well as the results of cranial magnetic resonance imaging (MRI) and MR venography which were performed with the suspicion of increased intracranial pressure syndrome. Cerebrospinal fluid (CSF) pressure was measured as 270 mmH_2O through lumbar puncture (LP); the fluid was clear and no cells were observed on direct microscopy. No pathological finding was detected in the biochemical, serological and cytological examination of CSF. Biochemical analysis revealed a calcium (Ca) level of 5.5 mg/dL, a phosphorus (P) level of 4.5 mg/dL, a magnesium (Mg) level of 1.5 mg/dL, an alkaline phosphatase (ALP) level of 130 U/L and other parameters were within normal ranges. In hormone tests, PTH was <3 IU and vitamin D3 was 8 IU, whereas the levels of vitamin B12 and folic acid, and the results of thyroid function test, vasculitis test, diagnostic tests for autoimmune hypopara-thyroidism, serologic tests, and whole-body scintigraphy were within normal ranges.

The diagnosis of idiopathic hypoparathyroidism and vitamin D deficiency was established by the Department of Endocrinology and Metabolism, Gaziantep University Faculty of Medicine. In the treatment of the patient, calcium carbonate (5000 mg/day), 1-alpha-hydroxyvitamin D3 (0.5 µg/day), calcitriol (1.0 µg/day), and acetazolamide tablets (750 mg/day tablets) were initiated. In control LPs, CSF pressure was found to be increased (290 mmH_2O) and papilledema was persisting. The dose of acetazolamide was increased to 1500 mg/day and blood tests were repeated. These tests revealed a Ca level of 7.5 mg/dL, a vitamin D3 level of 12 IU, a PTH level of <3 IU, a hemoglobin (Hb) level of 9.6 g/dL, a mean cell volume (MCV) of 92.7 fL, a serum iron level of 39 ug/dL, an iron binding capacity of 345 ug/dL, reticulocyte percentage of 0.56%, and a vitamin B12 level of 107 pg/mL and folic acid level was within normal ranges. Peripheral blood smear revealed no increase in the number of hypersegmented neutrophil; however, anisopoikilocytosis and hypochromia was present. Her MCV was within the normal range and inadequate reticulocytosis was present; thus, she was diagnosed with iron deficiency anemia accompanying pernicious anemia caused by vitamin B12 deficiency. In the treatment, ferrous sulfate tablets twice a daily and vitamin B12 ampoules (cyanocobalamin) once a daily were initiated. In the investigation of the etiology of anemia, her fecal occult blood test was negative; however, *Helicobacter pylori* (*H. pylori)* antigen was detected using the urea breath test and eradication treatment was initiated.

During the follow-up, no increase was observed in the hemoglobin levels despite the adequate duration and doses of treatment, but an increase was observed in MCV levels. Routine biochemical analysis, blood count and hormone tests were repeated. Her serum Ca level was 10.2 mg/dL, vitamin B12 level was 100 pg/mL, Hb level was 10.4 g/dL, MCV was 93, serum iron level was 80 µg/dL, serum iron binding capacity was 273 ug/dL, PTH level was <3 pg/mL, and vitamin D3 level was 22 IU. According to the test results, her iron deficiency anemia was improved and thus her low level of hemoglobin was associated with vitamin B12 deficiency and replacement therapy for vitamin B12 deficiency was continued parenterally. Urea breath test was repeated and no *H. pylori* antigen was detected. Blood count and biochemical test results and vitamin B12 levels were found to be within normal ranges in the examinations performed after three months. Control LP was performed, CSF pressure was found to be normal and papilledema was found to be regressed on fundoscopic examination. Acetazolamide was tapered off.

Discussion

Idiopathic hypoparathyroidism is a heterogeneous group of disorders characterized by low PTH levels accompanied by hypocalcemia, hypomagnesemia, and hyperphosphatemia. Hyperphosphatemia reduces serum vitamin D levels by suppressing its production in the kidneys. Hypoparathyroidism and hypocalcemia may cause a calcification in the basal ganglia, extrapyramidal system symptoms, a papilledema, and an increase in the intracranial pressure. In hypoparathyroidism, pseudotumor cerebri occurring without the symptoms of hypocalcemia (Trousseau's and Chvostek's signs) is a rare condition and its pathogenesis is not well-known (1,9).

After several examinations, our case was diagnosed with idiopathic hypoparathyroidism and her treatment was

initiated accordingly. No decrease in the CSF pressure was observed despite the normal serum Ca level; thus, blood biochemical, hormone and vitamin tests were repeated. Serum vitamin B12 and iron levels were found to be low. Iron deficiency accompanying megaloblastic anemia was observed and her treatment was rearranged accordingly. However, her vitamin B12 level remained low despite a rapid increase in iron level. Although rarely reported in the literature, few cases emphasizing persistent vitamin B12 deficiency caused by hypocalcemia and vitamin D deficiency secondary to idiopathic hypoparathyroidism have been reported (9). In such cases, vitamin D replacement therapy should be initiated and its levels should be closely monitored in the treatment of hypoparathyroidism (9).

In such cases, serum Ca, P, PTH, vitamin D, and vitamin B12 levels and blood count values should be evaluated. Moreover, blood tests should be repeated as well as control LP. The current case was presented due to small number of similar cases reported in the literature and to the fact that an improvement was achieved in clinical condition by treating the underlying causes rather than administering classic anti-edema therapy.

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