DE GRUYTER OPEN

CASE REPORTS

Fahr's Syndrome and Secondary Hypoparathyroidism

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A typical case of Fahr's syndrome is described in a 76-year-old Brazilian female who underwent a total thyroidectomy three decades ago. Six years before the current admission, she started with generalized tonic-clonic seizures. Associated disorders involved extra-pyramidal, cognitive, nocturnal terror and mood changes. With suspicion of hypocalcemia due to secondary hypoparathyroidism, laboratory determinations confirmed the diagnoses. Furthermore, imaging studies of the central nervous system detected multiple calcifications, with characteristic distribution of Fahr's syndrome. Clinical management was successful.

Key words: Fahr's syndrome, hypocalcemia, hypoparathyroidism, thyroidectomy.

INTRODUCTION

Fahr's syndrome or bilateral striatopallido-dentate calcinosis is a condition of sporadic or familial occurrence [1-4]. The reported prevalence of this syndrome is less than 0.5%, but it seems underestimated [3]. Individuals between 30 and 60 years constitute the more commonly involved group [2, 3]. Hypocalcemia and hypoparathyroidism play a major role in calcifications of the central nervous system associated with the neurological and psychiatric features of the syndrome [1-4]. Diagnostic challenges are related to the lack of indicative symptoms in the early phase, and confirmatory data depends upon laboratory and imaging studies of the brain structures. The aim of this case study is to enhance the suspicion index of primary care workers.

CASE REPORT

A 76-year-old Brazilian female was admitted presenting with vomiting and syncope followed by generalized tonic-clonic seizure. She underwent a total thyroidectomy 30 years before, because of a benign goiter, and convulsions appeared six years after surgery, with gradually shorter periods inter crises. Other antecedents were epilepsy, extra-pyramidal symptoms, cognitive impairment, and nocturnal terror and mood changes. There was no history of similar cases in her family, and she denied use of alcohol or tobacco. She had been under regular use of levothyroxin, levodopa, phenobarbital, trazodone, rosuvastatin, and acetylsalicylic acid. More recently, she started taking amiloride hydrochloride for blood pressure control. On admission, she was confused and in postictal state (Glasgow scale: 14), with normal sensibility, motricity and cranial nerve functions. The rest of clinical examination was considered unremarkable. Laboratory results (normal range in parenthesis) revealed calcium 5.0 (8.1-10.4) mg/dL, magnesium 1.4 (1.4-2.0) mEq/L, potassium 3.2 (3.5-5.0) mEq/L, sodium 132 (136-145) mEq/L, creatinine 1.3 (0.6-1.2) mg/dL, urea 50 (15-45) mg/dL, and parathormone 3 (16-87) pg/mL. Furthermore, the images of non-contrast brain computerized tomography (CT) revealed multiple bilateral calcifications involving the basal ganglia, thalamus, dentate cerebellar nuclei, and the centrum semiovale (Figure 1). Additionally to diuretic withdrawal, she received intravenous solution of calcium gluconate, and her clinical course was characterized by progressive improvement of the neuropsychiatric manifestations. After the hospital discharge, she is under outpatient control and taking oral calcium supplements.

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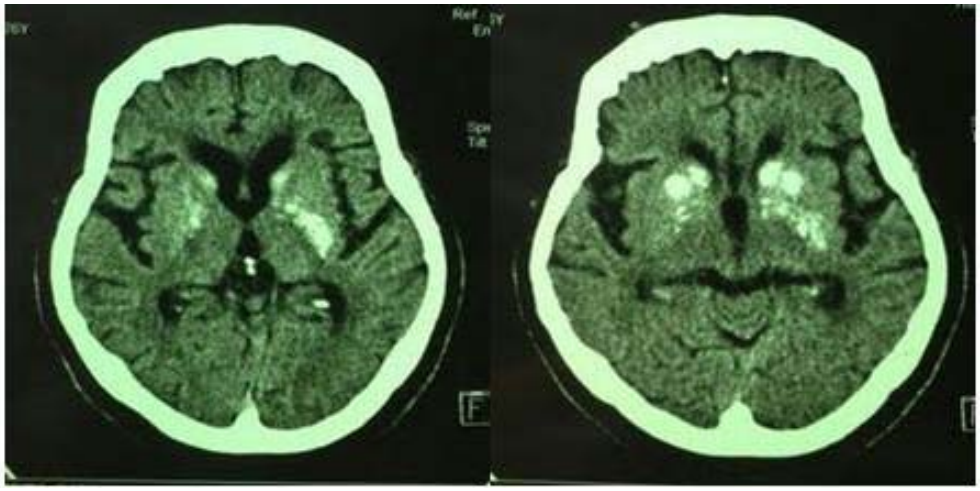

Figure 1. Non-contrast CT of the brain showing images of bilateral calcifications involving structures of the basal ganglia, thalamus, centrum semiovale, and dentate cerebellar nuclei.

DISCUSSION

The woman herein reported had clinical and imaging features of Fahr's syndrome. Main symptoms include extra-pyramidal and cerebellar disorders, cognitive impairment, epileptic seizures, and psychiatric changes [2-4]. Complementary data include hypocalcemia and hypoparathyroidism, and pathognomonic images of computed tomography are bilateral calcifications of basal ganglia, thalamus, centrum semiovale, and dentate cerebellar nuclei [1-4]. Clinical features of the syndrome often appear in people with 30-60 years of age [2, 3]. Hypocalcemia is found in chronic renal failure, deficiency or resistance to vitamin D, hungry bone syndrome, hypoalbuminemia, hypomagnesemia, hypoparathyroidism, malignant diseases, pancreatitis, pseudohypoparathyroidism, and rabdomyolisis [5]. Acute hypocalcemia can cause neurological symptoms of variable intensity, from mild paresthesias up to tetany, and chronic hypocalcemia may evolve unsuspected because of the absence of symptoms [5,6].

Hypoparathyroidism may be associated with parathyroid developmental defects; autoimmune, genetic or infiltrative diseases; drugs, radiation, surgery or tumors; dysfunction in receptors or sensors of calcium, hypomagnesemia, maternal hyperparathyroidism, and parathormone mutation [5]. Secondary hypoparathyroidism and chronic hypocalcemia give origin to calcifications in the basal ganglia and other regions of the central nervous system [1-7]. These changes contribute to the origin of neurological and psychiatric symptoms including extrapyramidal disorders, convulsions, emotional and behavioral disturbances [1-4, 6, 7].

The patient herein described presented with classical manifestations of the Fahr's syndrome, which developed as a late consequence of iatrogenic hypoparathyroidism secondary to total thyroidectomy. Moreover, the brain images disclosed by CT evaluation were characteristic of this scarcely reported syndrome [1-4, 6, 7]. The diagnosis of hypoparathyroidism was established by the exceeding low serum levels of the parathormone [5]. Postoperative hypoparathyroidism is a transitory condition in the majority of cases, and functional recovery occurs between ten days and six months after thyroidectomy; nevertheless, some individuals have permanent disturbances of calcium metabolism [5, 7]. In this patient, hypoparathyroidism was due to removal or devascularization of the glands [5]. Late-onset hypoparathyroidism must merit special attention after completion thyroidectomy [7]. Gulcelik *et al*. compared complication rates after total and completion thyroidectomies in Turkish patients with benign or malignant conditions. They found 20.7% of temporary and 4.4% of permanent hypoparathyroidism after completion thyroidectomies, and these rates were higher than previously described in literature (3-15% and 0-3.5%, respectively) [7]. Individuals with Fahr's syndrome can evolve with nonspecific features for a long time, and they might be under risk of severe complications, including cardiomyopathy [1]. Mechanisms of disease involve changes in blood flow and

cellular transport; precipitation and mineralization of serum proteins, and persistent inflammatory processes. Brain lesions may show deposits of calcium; other minerals and lipids, and demyelinization [2]. Prevention of postsurgical hypothyroidism includes parathyroid autotransplantation [3]. Oral supplements of calcium and vitamin D prevent late consequences of hypocalcemia, and the serum calcium level should be maintained between 8.0 and 8.5 mg/dL [1, 2, 5].

CONCLUSION

Fahr's syndrome is an uncommon condition, which seems underestimated. Because of non specific symptoms, it evolves unsuspected and may cause diagnostic pitfalls for primary care workers.

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Un caz tipic de sindrom Fahr este descris la o pacientă în vârstă de 76 de ani de sex feminin care a suferit o tiroidectomie totală cu trei decenii în urmă. Pacienta a acuzat, cu şase ani înainte de internarea curentă, convulsii tonico-clonice generalizate. Pacienta acuza sindrom extrapiramidal, anxietate, tulburări de comportament şi disfuncţie cognitivă. Analizele de laborator au confirmat diagnosticul având în vedere suspiciunea de hipocalcemie datorată hipo-paratiroidismului secundar. Studiul imagistic al sistemului nervos central a relevat multiple calcificări cu distribuţii caracteristică sindromului Fahr. Pacienta a fost tratată cu succes terapeutic.

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