中文題目：續發性副甲狀腺低下症引發 Fahr 氏症候群

英文題目：Fahr's syndrome induced by secondary hypoparathyroidism

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Introduction

Fahr's syndrome is a rare disease which is characterized by symmetric deposition of calcifications over bilateral basal ganglia. Besides, areas of brain that control movement such as thalamus, dentate nucleus, cerebral cortex and cerebellum may also be involved. Thus patients with this syndrome can present neuropsychiatric symptoms, extra pyramidal symptoms and even cerebellar dysfunction, dementia and seizure.

Case presentation

The 52 years-old female was sent to our emergency room due to seizure attack. This patient had history of thyroidectomy due to thyroid nodule at the age of 24 under thyroxine treatment and major depression diagnosed at age 49 under Sertralin control. Her family found that the patient had hand tremors, progressive bradykinesia and gait instability since two months ago. However, comatous consciousness and general convulsion occurred on the admission day. Then she was sent to our emergency room. At there, her vital signs are unstable as tachycardia and tachypnea (BP-195/112mmHg, T.P.R-36.5℃, 124/min, 26/min). Due to persisted seizure and desaturation, intubation was done and sedation with Midazolam was used. Laboratory examination revealed hypocalcemia (3.9mg/dl), hyperphosphatemia (5.8 mg/dl), no hypomagnesemia (1.7mg/dl) and normal Creatinine level. Brain computed tomography showed multifocal calcifications in the bilateral basal ganglia, thalami, cerebral and cerebellar hemispheres and mild brain atrophy. After admission, anticonvulsant as Levetiracetam was used. Lumbar puncture showed no CNS infection and autoimmune screening tests were unremarkable. Intact parathyrin and 25-OH Vitamin D were checked and disclosed very low level of iPTH (<3 pg/ml) but normal range of 25-OH Vit D (32 ng/ml). Under the suspicion of Fahr's syndrome caused by secondary hypoparathyroidism complicated with seizure attack, intravenous calcium gluconate and oral calcitriol were given and blood calcium and phosphate level were monitoring. Sedation was hold at the third day of admission and no more seizure attack. Her consciousness improved gradually and IV calcium supplement was shifted to oral calcium acetate at the fifth day. Endotracheal tube was removed at the seventh day and she was discharged at the ninth day under normal range of blood calcium and phosphate.

Discussion

Patients with Fahr's syndrome most present with extrapyramidal, and

neuropsychiatric signs, dementia and seizure which are caused by diffuse, symmetric calcifications of bilateral basal ganglia, thallumus, nucleus pallidus, cerebral cortex, subcortical white matter and the dentate nucleus of the cerebellum. Fahr's syndrome is caused by several etiologies, including (1) abnormalities of calcium-phosphate metabolism such as hypoparathyroidism, (2) infections of brain, like Toxoplasmosis, CMV, and Acquired Immune Deficiency Syndrome (AIDS), (3) Autoimmune disease as systemic lupus erythematosus. In this case, secondary hypoparathyroidism is the etiology of this syndrome. Thus our treatments were tried to control symptoms such as seizure and correct phosphate and calcium levels including oral calcitriol supplementation and intravenous calcium.

Conclusion

Fahr's syndrome is a progressive neuropsychiatric disorder. Many etiologies could cause this syndrome. Timely recognition and management of underlying cause are recommended to prevent disease progression.