

A Rare Association of Fahr's Syndrome and Hashimoto's Encephalopathy

Case Report

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Abstract

Background: Steroid Responsive Encephalopathy Associated with Autoimmune Thyroiditis (SREAT), also known as Hashimoto encephalopathy, is a rare but treatable cause of rapidly progressive dementia. Coexistence with Fahr's syndrome, an idiopathic bilateral basal ganglia calcification, is exceptionally uncommon and poses a diagnostic challenge.

Case Presentation: We report a 70-year-old female who presented with a 6-month history of insidious onset behavioral changes, psychosis, cognitive decline, and movement abnormalities including bilateral rest tremors. Laboratory investigations revealed elevated anti-thyroid peroxidase (anti-TPO) antibodies, thyroiditis on ultrasound, elevated parathormone, and vitamin D deficiency. Neuroimaging showed coarse bilateral calcifications in the basal ganglia, dentate nuclei, and occipital gyri consistent with Fahr's syndrome. Serum autoimmune encephalitis markers were negative. The patient responded dramatically to intravenous pulse steroids and rituximab, with marked clinical improvement and cognitive recovery.

Conclusion: This case highlights the importance of considering SREAT in rapidly progressive dementia even when imaging shows typical Fahr's syndrome calcifications. Early recognition and immunosuppressive treatment can lead to significant clinical improvement in this potentially reversible disorder, underscoring the need for vigilance in atypical presentations.

Keywords: SREAT; Rapidly progressive dementia; Fahr's syndrome

Introduction

Rapidly progressive dementia refers to cognitive decline with fast progression which is commonly considered to be less than 1 or 2 years. The etiology includes immune-mediated, infectious, metabolic encephalopathies which are treatable as well as prion diseases and atypical rapid presentations of more common neurodegenerative diseases. Among these, Steroid Responsive Encephalopathy Associated with Autoimmune Thyroiditis (SREAT), also known as Hashimoto encephalopathy, is a rare but important treatable cause characterized by subacute cognitive decline, psychiatric manifestations, elevated thyroid autoantibodies, and a favorable response to immunosuppressive therapy.

Fahr's syndrome, defined by bilateral basal ganglia and other brain region calcifications, is an uncommon neurodegenerative disorder classically associated with movement disorders and cognitive impairment. The coexistence of Fahr's syndrome and SREAT is rare and poses a diagnostic dilemma, especially when imaging abnormalities suggest a neurodegenerative process.

This case report describes an elderly female patient presenting with rapidly progressive dementia and neuropsychiatric symptoms, found to have elevated anti-thyroid antibodies and classic basal ganglia calcifications consistent with Fahr's syndrome.

Case report

A 70-year-old female had presented with complaints of insidious

onset slowness of activities for 2 years and behavioural disturbances which started as episodes of anger outbursts, usage of foul language, auditory and visual hallucinations, delusion of persecution for 6 months. There was no history of fever, headache and seizures. The patient also had history of irrelevant talks, throwing up of objects and later developed way finding difficulty. Later on, the patient became apathetic requiring assistance for all activities of daily living.

On examination -The patient was awake, irritable, not cooperative, disoriented with lack of attention. MMSE-6. She was afebrile. without signs of meningeal irritation. Both pupils were equal, 3mm and reacting to light. She was moving all 4 limbs with a power of 4/5. However, they were rigid and bradykinetic. All DTRs were brisk with bilateral extensor plantar response. There was bilateral rest tremor of hand (left>right).

On investigation -The patient had normal hemogram with normal liver and renal function test.

MRI brain was suggestive of coarse calcification in bilateral basal ganglia, dentate nuclei and occipital gyri. She had low normal serum calcium level. With elevated parathormone levels and vit-D3 deficiency. Her TSH- 5.4 iu/L. CSF analysis showed acellular smear with elevated protein level of 97 g/dl.

EEG showed a normal record

In view of subacute onset of behavioural symptoms. Serum markers of autoimmune encephalitis were tested which turned negative. However Anti TPO antibodies were 917 IU/ml with elevated ESR and C-Reactive protein. Ultrasound scan of neck was suggestive of thyroiditis.

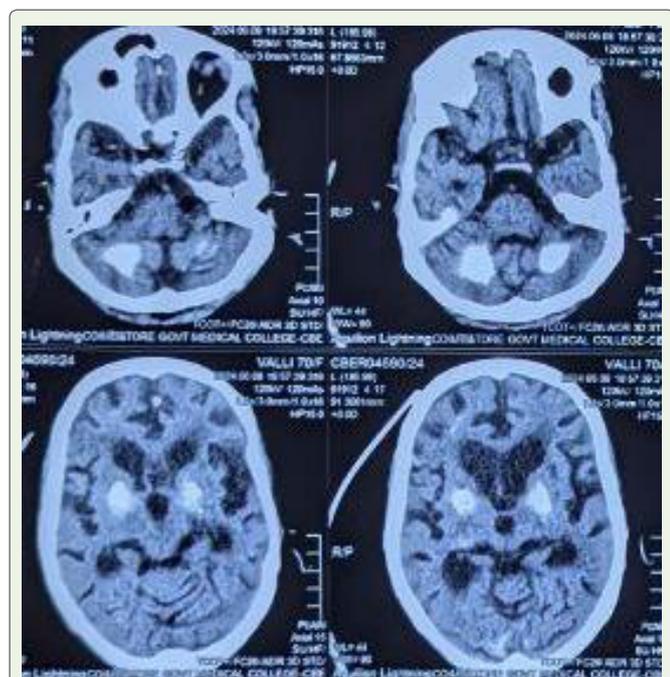


Figure 1: CT- brain showing bilateral symmetric calcification of basal ganglia and dentate nucleus of cerebellum

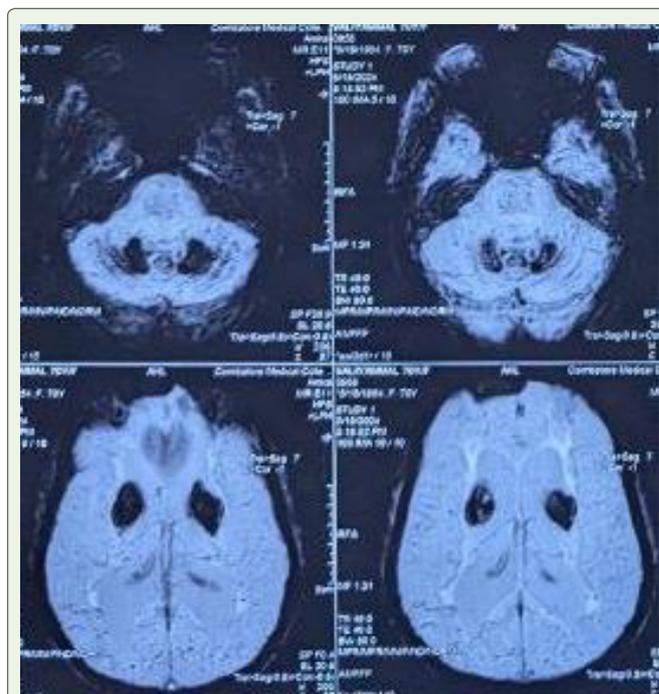


Figure 2: MRI -SWI revealing bilateral symmetric hypointense signals (dark spots) from basal ganglia and dentate nucleus of cerebellum

A diagnosis of Hashimoto Encephalitis was considered

The patient was treated with Intravenous Injection. Methylprednisolone 1 gm for 5 days along with vitamin D3 and calcium supplements and also with Inj. Rituximab 1gm 1 week after pulse steroid course.

5 days after the rituximab dose the patient’s cognition started improving. Her MMSE score improved to 27 at the time of discharge. However, there was no improvement with respect to her motor disability. she was discharged on tapering doses of steroid

Discussion

Most SREAT case reports describe a female predominance with a wide age range, often presenting with subacute or chronic neuropsychiatric symptoms, cognitive decline, psychiatric disturbances, and occasionally movement disorders such as myoclonus or tremor.

High anti-TPO titers are a hallmark in nearly all SREAT cases, even in the presence of normal or slightly abnormal thyroid function tests [1-3]. CSF often shows elevated protein without pleocytosis. [3]

Imaging varies: While many cases report normal MRIs or nonspecific changes, some describe white matter changes or atrophy. Unusually, this case features coarse calcifications in basal ganglia, dentate nuclei, and occipital gyri, meeting radiological criteria for Fahr’s syndrome.[1,4]

It is quite challenging to distinguish Fahr’s calcifications from age-related changes or other metabolic causes. However, the coexistence of autoimmune thyroiditis and Fahr’s syndrome points to possible metabolic or autoimmune overlap.[4-7]

Fahr's syndrome presents with insidious onset movement disorder like tremor, bradykinesia, rigidity and also cognitive decline and psychosis particularly late in onset, as in our patient.

The literature highlights diagnostic dilemmas when metabolic or degenerative findings (e.g., Fahr's calcifications) coexist, sometimes confounding initial impressions or delaying immunotherapy.[1,4,5]

The rapid progress in cognitive decline and psychosis prompts to look for other possible causes to exclude.

SREAT remains a diagnosis of exclusion, based on characteristic clinical syndrome, high antithyroid antibody titers, and steroid responsiveness.[1-3] Other treatable RPDs such as voltage-gated potassium channel antibody encephalitis and NMDA-R encephalitis present with slightly different syndromes and biomarkers.[8,9]

Most reports cite rapid and dramatic improvement with steroids as highly suggestive of SREAT. Immunosuppressants or biologics (such as rituximab) are reserved for refractory cases.[1-3]

The discussed patient responded well to pulse steroid and rituximab, paralleling outcomes in published series, emphasizing the treatable nature of SREAT despite atypical imaging.[1]

Fahr's syndrome, with basal ganglia calcification and elevated parathormone, has occasionally been reported alongside autoimmune thyroiditis (chronic lymphocytic thyroiditis). Few case reports highlight this overlap, hinting at shared autoimmune or metabolic pathways.[1,5-7]

Conclusion

This case underscores that steroid-responsive encephalopathy associated with autoimmune thyroiditis (SREAT) should be considered in rapidly progressive dementia, even in the presence of basal ganglia calcifications suggestive of Fahr's syndrome. The

unique radiological overlap highlights possible coexistence of neurodegenerative and autoimmune-metabolic processes in elderly patients. High anti-thyroid antibody titers and dramatic steroid responsiveness are key diagnostic clues for SREAT, despite atypical imaging findings. Timely institution of immunomodulatory therapy, including steroids and biologics, can lead to significant neurological recovery in such atypical presentations.

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