Vitamin B12 Deficiency Presenting As Food Faddism and Rapidly Progressive Dementia

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Abstract

Vitamin B12 deficiency can affect the nervous system at all levels, from central to peripheral. However, when patients present with rapidly progressive dementia, inflammatory or infective causes are considered more likely. Our patient presented with a rapidly progressive Fronto Temporal syndrome who had a unique food faddism for non-vegetarian food from his vegetarian habit. He had a very poor MMSE score of 7/31, indicative of poor outcome generally. Our patient had extensive hyper pigmentation of skin of the back, both legs and extensor surface of the upper limb, and lower limbs and axilla but not the classical knuckle pigmentation. Investigations showed megaloblastic anemia with hyper-segmented neutrophils; Vitamin B12 was 297pg/ml with normal homocysteine levels. MRI Brain showed bilateral T2W and FLAIR periventricular hyperintensities, dorsal column signal changes in the cervical spine. With six weeks of treatment, his MMSE improved to 30/31, and he resumed routine work. It was interesting to note the new food faddism to non-vegetarian food and less typical skin changes.

Keywords: Rapidly progressive dementia, Vitamin B12 deficiency, Generalized hyperpigmentation, Food faddism.

Key message: Rapidly progressive dementia with diffuse hyperpigmentation and New Food faddism can occur with B12 deficiency associated cognitive decline.

Introduction

Rapidly progressive dementias are classified as Prion-associated and non Prion associated cognitive decline. A wide spectrum of conditions, including degenerative ones, can present with the rapid course. However, a high degree of suspicion is needed to categorize the reversible ones as life-saving.[1] Toxic-metabolic causes account for 2 to 4% of all RPD depending upon the study and age group.[2,3,4,5] Only a few case reports are available regarding Vitamin B12 deficiency presenting as rapidly progressive dementia.[4,5] Vitamin B12 is generally found in animal products and is the only vitamin stored in the liver for a long time. It has an important role in forming myelin and Nucleic acid synthesis and methylation for converting Homocysteine to Methionine by methionine synthase and S-adenosylmethionine (SAM). S adenosyl homocysteine (SAH) occurs in a deficient state, inhibits methylation, and increases homocysteine, causing damage to vascular endothelium and inhibition of NMDA receptors. These contribute to neuropsychiatric disorders including schizophrenia, Dementia, Fatigue, depression, and developmental problems in children.

Patient Presentation

61-year-old tailor, educated till 10th standard, married, right-handed person, vegetarian presented with severe loss of executive function, lack of initiative, and new onset of obsession for Non-vegetarian food, followed by recent memory impairment, naming defect, and double incontinence of six months duration. He had six brief episodes of unexplained syncope and became bed-bound 15 days before admission. There was no history of hallucinations, delusions, alteration in sensorium. No history of fever, joint pains,

arthralgia, dry mouth, rash, mucocutaneous ulcers, myoclonic jerks, infections, substance abuse, or other systemic diseases. General physical examination showed hyper pigmentation over the whole back, both legs, and extensor surface of the upper limb and axilla (Figure 1A, 1B). The patient was conscious but apathetic and inattentive. HMSE: 7/31 (Orientation in Time 0/5, Place 0/5. Registration 2/3, Attention and Calculation 0/5, Recall 0/3, Naming 2/2, reading 1/1, writing 0/1, copying 0/1, 3 step command 2/3). On detailed mental status, the examination had features of frontal and temporal lobe involvement. Cranial nerves, tone, and power of all limbs, sensory and cerebellum-normal, brisk deep tendon reflexes with right plantar withdrawal response. Gait was cautious but uncharacterized. His investigations revealed the following: Hemoglobin-11.1grm/dl, WBC count 8.8 K/ul, MCV118.7fl, peripheral smear: dimorphic blood picture with hyper-segmented neutrophils, serum Vitamin B12-297 (180-914 pg/ml), serum Homocysteine 9.12 (<15 umol/L). ESR, Thyroid functions, hepatic and renal function tests, electrolytes, autoimmune antibody, vasculitis and paraneoplastic profile, CSF analysis with chronic meningitis workup, HIV, VDRL, and autonomic function tests were normal. EEG was normal. MRI of the Brain showed bilateral T2W and FLAIR periventricular hyperintensities. Dorsal column signal changes in the cervical spine were noted in MRI of the spine (Figure 2). In view of skin changes, hematological and imaging findings suggestive of nutritional deficiency patient was investigated further in spite of subtle biochemical evidence with upper GI endoscopy followed by a biopsy, which showed features of duodenitis. So, diagnosis of rapidly progressive dementia secondary to Vitamin B12 deficiency was considered, and he was treated with Vitamin B12 1000 mcg daily for ten days, followed by a weekly dose. In six weeks patient resumed normal life, skin changes improved, and HMSE improved to 30.

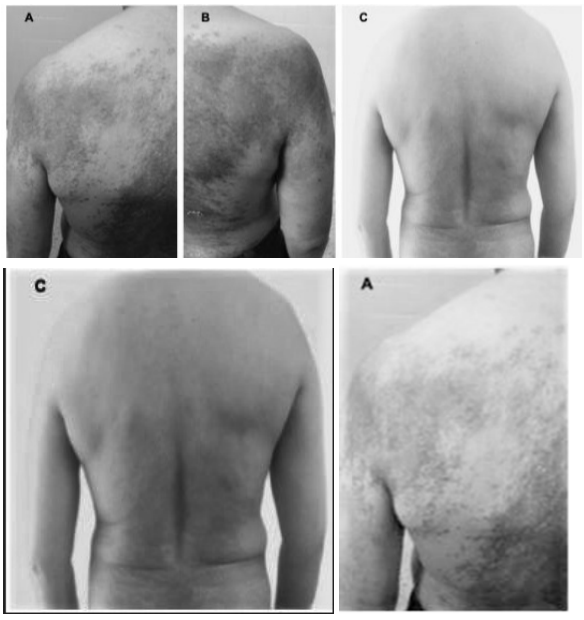

Figure 1: *Dermatological clues: At presentation: hyperpigmented macular lesions over back and extensor surface of upper limbs (A and B). At 6 months follow up: improvement of lesions (C).*

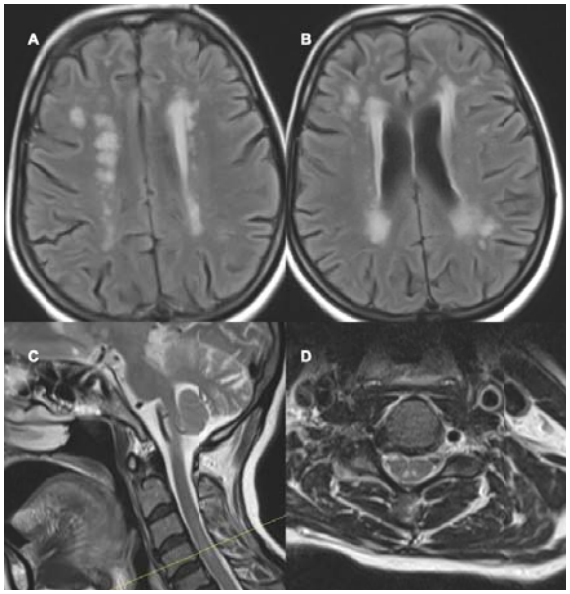

Figure 2: *Imaging clues: MRI of Brain: T2 FLAIR Axial and sagittal sections show bilateral periventricular linear hyperintensities as well as rounded lesions (A and B). MRI of spine: posterior column hyperintensities in cervical cord with incomplete inverted V sign (C and D).*

Discussion

All dementias are not due to irreversible cause[2]. It is important to look for reversible causes in all patients, more so if they are rapidly progressive.[6,1] Infections and autoimmune encephalitis attribute to most of these cases; other aetiologies include vasculitis, toxic, metabolic, and nutritional.[1,2,3] As a cause of dementia, nutritional deficiency is reported in 4% of dementias in a large cohort of patients.[3,2,7]

The domains of cognition affected in patients with B12 deficiency are attention, Mood, Executive functions, Memory, and closely imitates Frontotemporal dementia.[8,9] Eating behavior like a preference for sweets, lack of satiety, eating several times, eating from other person plate and lacking table manners, Gluttony, etc. is common in semantic variant Fronto-Temporal Dementia (FTD), but food faddism are not described in non-degenerative dementia[10] and is a unique feature in our patient with a special interest in non-vegetarian food in a previously vegetarian patient. The term food faddism is described as a tendency to exaggerate or eliminate certain foods for presumed benefits. Degeneration of insular, anterior temporal, mesolimbic areas is believed to play a role in sensory food perceptions and its semantic association and programming appropriate behavior.[11]

Hyperpigmentation of the skin is the common manifestation of Vitamin B12 deficiency in children.[12] Cutaneous hyperpigmentation was noticed in 19% of patients with Vitamin B12 related neurological disorders; however, generalized hyperpigmentation was reported in a few cases reports.[13] The current study patient had also developed generalized hyperpigmentation, which responded significantly to Vitamin B12 therapy (Figure1). Cobalamin deficiency results in increased melanin synthesis and hyperpigmentation due to reduced inhibition of tyrosinase.[14]

Vitamin B12 deficiency-related spinal cord imaging abnormalities are well described, but only a few case reports describe brain lesions. Brain MRI may show periventricular T2, Flair hyperintensity sometimes discrete periventricular and subcortical lesions mimicking other demyelinating disorders.[15,16] Co-occurrence of brain and spine lesions is described only in a few case reports. The present study patient had both brain and spinal cord lesions, similar to previous reports (Figure 2).

Usually, the diagnosis is confirmed on biochemical assays with serum Vitamin B12, homocysteine, and methylmalonic acid levels. Elevated homocysteine and methylmalonic acid have high sensitivity even in the presence of normal Vitamin B12 levels.[15] The present patient laboratory investigations revealed anemia with raised MCV, dimorphic blood picture, and hyper-segmented neutrophils consistent with Vitamin B12 deficiency. However, homocysteine was normal, with Vitamin B12 in the lower range of normal. In spite of subtle biochemical findings, diagnosis of Vitamin B12 deficiency was considered due to constellation of symptoms, generalized hyperpigmentation, consistent hematological and imaging findings. Vegetarian diet and duodenitis were attributed as the cause in our patient. The patient clinically entirely improved to vitamin B12 supplements and resumed work confirming the diagnosis.

Conclusion:

Our patient presented with features of rapidly progressive dementia and radiological evidence of both brain and typical spinal cord involvement of vitamin b12 deficiency. He showed a unique new-onset food preference to the non-vegetarian diet, which is uncommon in vegetarians and non-degenerative dementias. His skin changes were also extensive and not the typical knuckle pigmentation. This patient is reported to highlight that altered food preferences and food faddism is not a unique

feature of degenerative Dementias alone and always should look for a treatable cause.

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