

Case Report on a Misdiagnosed Case of Subcortical Vascular Dementia – the Importance of Sound Knowledge of Psychiatry With Proper History Taking

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Abstract - Psychiatric symptoms at presentation may often be missed, if not suspected or specifically explored. A missed psychiatric diagnosis may lead to dire consequences in terms of poor quality of life and function for the patient, affecting overall quality of healthcare provided. This lady presented with depressive symptoms after multiple strokes and was initially diagnosed as post stroke depression. However, after it was observed that she did not show any improvement in symptoms despite being on antidepressants, subsequent further investigations revealed a history more suggestive of subcortical vascular dementia. Consequently, detailed neuropsychological and neuropsychiatric assessments, including NUCOG, and relevant investigations including MRI brain scans were performed suggesting a diagnosis of vascular dementia. This case illustrates that an insufficiently thorough assessment and treatment process results in unnecessary morbidity, prolongs duration of illness, and increases social and occupational dysfunction to the patient. Hence, it further underscores the need to perform a thorough history, physical examination and relevant investigations to ensure organic aetiologies are ruled out in clients with relevant sociodemographic and clinical risk factors.

Keywords: dementia; depression; post stroke; neuropsychiatry; organicity

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Introduction

The diagnosis and treatment of psychiatric disorders are often not as straightforward as their medical or surgical counterparts. Psy-

chiatric symptoms at presentation may often be missed, if not suspected or specifically explored. A missed psychiatric diagnosis may lead to dire consequences in terms of poorer quality of life and functioning for the patient, affecting the care given overall.

This case highlights several pertinent issues. Firstly, the importance of some knowledge of psychiatry with thorough history taking is underscored. Secondly, management of this case

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highlights the importance of multidisciplinary involvement from other specialties and hospitals in coming to a more accurate diagnosis, by understanding the limitations of the primary treating team, in order to give the best treatment. Thirdly, this case report reiterates how crucial it is to formulate an accurate diagnosis from a quality of life point of view, as in this case, an accurate diagnosis allowed her to be medically boarded for permanent disability benefits.

Case Report

J is a 38-year-old lady with a twelve-year history of metabolic syndrome. She was diagnosed with hypertension, diabetes mellitus and high cholesterol since she was 26 years old at a district health clinic. She was pre-morbidly obese, with a genetic predisposing factor, namely maternal hypertension. She frequently defaulted going for her appointments at the district health clinic and was not consistently taking her medications, consequently suffering from two strokes.

The first episode of stroke was in October 2014 at the age of 32. J came to the emergency department of a tertiary hospital, complaining of left sided weakness for two days. A few days prior she felt well albeit a little tired which she felt was due to her heavy workload at school. Upon further questioning, there was no blurring of vision, seizure, respiratory or urinary tract infection. She did not have any symptoms suggestive of an acute coronary syndrome like chest pain, palpitations, shortness of breath, orthopnoea, paroxysmal nocturnal dyspnoea or generalised shortness of breath. She was never prescribed oral contraceptive pills, nor was she a cigarette smoker. Due to religious obligations, she did not consume alcohol however she did admit her diet control was bad, eating fast food on most days with carbonated drinks. She reported no involvement in physical activities.

Her physical examination revealed a young Malay lady with a Body Mass Index (BMI) of 40, she did not appear jaundiced nor did she have any clubbing. There was no facial asymmetry or slurring of speech. Her Cardiovascular, respiratory and abdominal examinations were normal. There was no carotid bruit. However, on examination of the Central Nervous System (CNS), power through the left side of her body was 4/5, otherwise the tone, reflexes and sensations were all intact and normal bilaterally. Cerebellar signs were all negative and all her cranial nerves showed no abnormalities. Her blood pressure was recorded high; however, Computed Tomography (CT) of the brain reported no obvious ischemia. Blood investigations done showed that fasting blood sugar and fasting lipid profile was high. Other investigations (full blood count, renal profile, liver profile, coagulation factors, thyroid function test, cardiac enzymes, arterial blood gas analysis, erythrocyte sedimentation rate and viral screening tests) were normal. Her electrocardiogram showed T inversion in lead III only. She was seen by a physician and given the diagnosis of 'hypertensive emergency with cerebrovascular accident'. She was admitted for 3 days only. During her stay in the hospital she was given Amlodipine 10 mg once a day (OD), Aspirin 10 mg OD, Simvastatin 40 mg OD, Metformin 500 mg twice a day (BD) and Gliclazide 40 mg BD. Prior to discharge, a physiotherapist saw her and advised her on the importance of exercise. A dietician also sat with her to discuss about strict diabetic diet. She was then discharged to be reviewed back at the district health clinic. She was compliant for two months however, over the next few months she defaulted her appointments at the health clinic and had poor adherence to her medications.

Her second episode of stroke was in March 2015. She complained of sudden onset of left sided weakness and generalized headache for 4 days. The presentation of her symptoms were

similar to the first episode. On examination, there was facial asymmetry. Her Cardiovascular, respiratory and abdominal examination was normal. There was no carotid bruit. On examination of the CNS, there were significant changes compared to the first presentation. On her left side, the power of both her upper and lower limbs were 4/5 and the tone was significantly reduced compared to the right side. Reflexes and sensation were intact bilaterally. Her Cerebellar signs were negative, and all her cranial nerves showed no abnormalities. Her CT brain (plain and contrast) showed right recent multifocal infarcts with mass effect (infarcts seen in the right basal ganglia and head of the right caudate nucleus and corona radiata). Her blood investigations showed that her diabetes mellitus and cholesterol were poorly controlled however other investigations (full blood count, renal profile, liver profile, cardiac enzymes, arterial blood gas analysis, erythrocyte sedimentation rate and viral screening tests) were normal. Her electrocardiogram showed left ventricular hypertrophy. She was diagnosed with 'recurrent cerebrovascular accident with left hemiparesis'. She was discharged 3 days later, again with a referral to the district health clinic to follow up on her metabolic syndrome control and to see a physiotherapist. Her medications on discharge were Perindopril 8 mg OD, Bisoprolol 1.25 mg OD, Amlodipine 10 mg OD, Aspirin 15 mg OD, Simvastatin 40 mg OD, Metformin 1 gram BD and Gliclazide 40 mg BD.

J's referral to psychiatric services was in 2016. Her family medicine physician noticed she looked sad and promptly referred her for psychiatric assessment. She reported feeling low with some anhedonia and worthlessness; however, no other baseline investigations were done. Mental state examination revealed a young obese lady with superficial eye contact and rapport who was euthymic and had coherent and relevant speech. She was given the diagnosis of Post Stroke Depression and

was treated with Escitalopram 20 mg OD. Her subsequent follow up to the psychiatry department was regular and during each appointment she would say she was alright. She came alone and no other family member was present to give collaborative history, and hence her medications were maintained at Escitalopram 20 mg OD.

However, in 2018, a second opinion was requested as her employers felt she was posing a risk to staff and students. Her employers felt that she was not improving despite attending psychiatric appointments. She was working with pre-school children, and some of her students had complained of food poisoning after eating food cooked by her. Her employers also did not feel that she was depressed, but rather appeared nonchalant or apathetic.

Upon reassessment, it was discovered she had cognitive, emotional and behavioural impairment after her stroke, which was not identified previously. There was marked deterioration of function from her pre-morbid functioning. Prior to her stroke, she was someone who liked cleanliness, could cook very well, was hardworking and very capable in handling household chores and work demands. Since her stroke in 2015, she could not perform simple household tasks, and had poor judgement and poor self-care, especially with regards to personal hygiene and dressing. She was quite careless, apathetic with poor emotional responses, had poor judgement, and was impulsive (cooking and serving raw food to her family, irritable and harsh towards the students for no apparent reason). There was also perseveration behaviour (asking the same questions again and again), social inhibition and poor grooming (going out of the house with torn clothes or coming out naked from the bathroom after soiling herself) and had hyperorality. There were no movement disorders or aphasia. Mental State Examination revealed a young lady who was morbidly obese, dressed in T-shirt and pants, wearing a headscarf. She

had good eye contact with the interviewer but was smiling inappropriately at times. Her speech was coherent and relevant. She described her mood as 'normal', however her affect was blunted. She denied any thought or perceptual disturbances. There were no primitive reflexes. Her cognitive assessment showed poor attention and concentration (unable to do serial 7), concrete thinking with poor social awareness, and reduced personal/clinical judgement. She had poor insight as she was not aware of illness and unsure of attribution (stroke) of illness.

Investigations

CT of the brain (plain) on her first presentation of stroke was documented as reported, with no obvious ischemia noted. However, in March 2015, the CT of her brain (plain and contrast) showed right recent multifocal infarcts with mass effect (Infarcts seen in the right basal ganglia and head of the right caudate nucleus and corona radiata). A repeated CT of the brain (plain and contrast) in April 2018 demonstrated multifocal infarcts with early onset cerebral involution. In 2018, her Mini Mental State Examination (MMSE) scores were 25/50 and the Montreal Cognitive Examination (MOCA) scores were 15/30 which showed poor performance in executive function. A Neuropsychiatry Unit Cognitive Assessment Tool (NUCOG) was performed, demonstrating scores of 70/100 (10/20 for the memory domain, 12/20 for the executive function domain and 12/20 for the visuoconstructional domain). This objective assessment showed predominant memory impairment with clinically significant dysexecutive function, but with no significant behavioural issues.

In February 2019, A magnetic resonance imaging (MRI) of her brain showed cystic encephalomalacia of the Right Basal Ganglia extending to Right Corona Radiata as the sequelae of old infarcts in Right Lenticulostriate artery territory (perforating branches of Right

Middle Cerebral artery). There were also old infarcts of the contralateral Left Lenticulostriate artery territory involving the posterior limb of Left Internal Capsule that extended into Left Corona Radiata. In addition, confluent T2 and FLAIR hyperintensities indicating small vessel ischemia in the internal watershed areas of the brain bilaterally were present involving the corona radiata and centrum semi-ovale with additional internal watershed infarct on the left side. Cerebral volume loss is also evident, more prominently involving both frontal lobes (left more than right) consistent with involuntional changes from the old infarcts. The pattern of ischemia and old infarcts can be explained by the vascular compromise demonstrated on the Magnetic Resonance Angiography (MRA) of the Brain, which showed truncation of flow signal from the level of terminal Internal Carotid arteries (ICA) on both sides, suggesting occlusion at these levels in keeping with Moya-Moya syndrome. Consequently, Middle Cerebral Arteries showed poor flow signal bilaterally and the flow within Anterior Cerebral arteries were reconstituted from the collateral vessels of the Circle of Willis. In summary, the radiological findings were multifocal old infarcts and bifrontal cerebral atrophy with ischemia and infarcts within the internal watershed of the brain as a result of occlusion of terminal segments of Internal Carotid arteries on both sides, consistent with Moya-Moya syndrome. Co-relating with the clinical presentation of the patient, the radiological findings were supportive for Vascular Dementia.

Provisional and Differential Diagnosis

J's diagnosis from 2016 to 2018 was post stroke depression. Upon reassessment in 2018, her initial differential diagnosis was Frontal Lobe Syndrome. However due to her age (young onset) and multiple medical comorbidities, a Neuropsychiatrist from another tertiary hospital was consulted. Following the NUCOG, she was provisionally diagnosed with

Frontotemporal Dementia – Behavioural Variant with features of frontal lobe syndrome. The neuropsychiatrist suggested that an MRI of the brain be done to confirm the diagnosis.

Post-MRI, a multidisciplinary meeting with the neuropsychiatrist and neuroradiologist was conducted, and the final diagnosis post-meeting was Young Onset Dementia - Subcortical Vascular Dementia with Moya-Moya syndrome from occlusion of the terminal segments of both Internal Carotid arteries with involvement of both frontal lobes as a result of internal watershed ischemia and infarcts as well as multifocal old lacunar infarcts within the vascular territory of the perforating branches of the Middle Cerebral arteries, indicating insufficient flow within the brain supplied by the anterior circulation arteries.

Treatment

The patient and her family were carefully updated about the diagnosis during a family conference. She and her husband were provided with psychoeducation about the illness and treatment options. All questions regarding her diagnosis and issues were addressed and answered. Her antidepressant was stopped, and she was started on a cholinesterase inhibitor (Donepezil 5 mg OD). Currently her symptoms have not worsened. As an additional benefit, after the removal of her SSRI antidepressant (Escitalopram), her emotional blunting improved and she can better express her emotions.

Outcome and Follow up

The prognosis for subcortical vascular dementia is poor and the treatment is given to avoid further damage, slow down the progression of the illness and help the patient and her family deal with various illness symptoms. On a positive note, she has good support from her husband, and they attend regularly for follow-up. She is currently awaiting the process for the Malaysian Medical Board to obtain per-

manent disability benefits from the national government. Hence it was important for her to have her diagnosis revised and documented clearly, as she would have not been able to procure disability benefits with her previous inaccurate diagnosis.

Discussion

Young Onset Dementia (YOD) is a quite common disorder, affecting patients younger than 65 years old [1]. It commonly presents with behavioural changes, psychiatric manifestations, and cognitive decline [2]. This eventually leads to a deterioration of day-to-day function, not only affecting patients, but also families, employers, friends, and caregivers [3].

Subcortical Vascular Dementia is a small vessel disease dementia with mild memory disturbances [4]. Vascular risk factors, including cerebrovascular disease, history of diabetes, high cholesterol and hypertension, can cause brain damage, predisposing individuals to dementia and cognitive impairment [5]. The psychopathology of vascular dementia is related to interactions between host factors (e.g. older age, lower education), brain changes (e.g. infarcts, atrophy), genetic factors (e.g. specific genetic features, family history), vascular insults (e.g. diabetes, hypertension, myocardial infarction, lipid abnormalities), ischemic-lesion-related (type of cerebral vascular disease, site and size of stroke) and poor lifestyle maintenance (e.g. reduced exercise, fast food, sugary food) [5]. White matter lesions, lacunar infarctions and varying degrees of vascular lesions are seen in Subcortical Vascular Dementia [6,7].

Due to the overlapping criteria for many dementia syndromes, misdiagnosis is common even in tertiary centres [8]. This is because in the early stages of illness, patients see various physicians and undergo various medical examinations, which causes delays in diagnosis [9]. A relevant and similar case was reported by Mendez (2006), who reported a 43-year-old


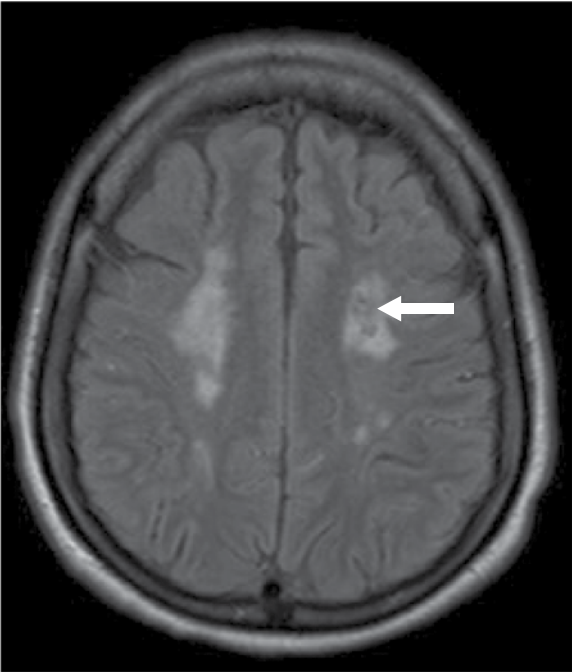
| MRI Brain Image | Description |
|--|---|
|  <p>(top image)</p> | a) Coronal FLAIR demonstrates old internal watershed infarcts involving the left frontal lobe (arrow) and old lacunar infarcts in the right basal ganglia (thick arrow) |
|  <p>(bottom image)</p> | b) Axial FLAIR image showing ischemia of internal watershed areas on both sides with additional infarcts seen on the left side (arrow) |

Figure 1. Magnetic Resonance Imaging (MRI) of the brain performed in February 2019.

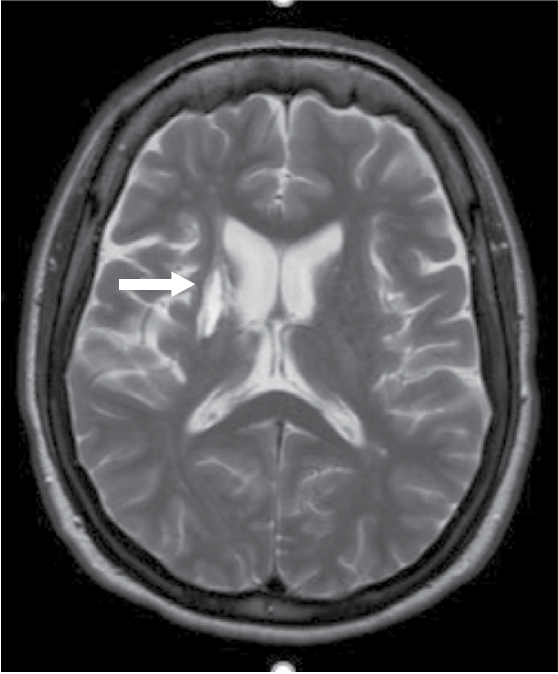

| MRI Brain Image | Description |
|--|---|
|  <p data-bbox="506 942 620 970">(top image)</p> | <p data-bbox="979 250 1295 473">c) Axial T2 brain demonstrates cystic encephalomalacia in right basal ganglia that extends into right corona radiata, as a result of old infarcts within right lenticulostriate artery (perforating branch of right Middle Cerebral artery) territory</p> |
|  <p data-bbox="484 1677 637 1705">(bottom image)</p> | <p data-bbox="979 986 1295 1180">d) Magnetic Resonance Angiography of the Brain showing bilateral occlusion of terminal segment of Internal Carotid arteries bilaterally (arrows) leading to poor flow signals within both Middle Cerebral arteries.</p> |

Figure 1. (Continued)

woman with 3-year personality change who was initially diagnosed with Frontotemporal Dementia. After a year of continuous decline, a positron emission tomography (PET) study and genetic testing was done revealing a diagnosis of presenilin-1 mutation for autosomal dominant Alzheimer's disease [8], leading to more appropriate clinical management.

To avoid such misdiagnoses, clinicians need to conduct adequate clinical histories, assess thoroughly levels of functioning including cognitive deficits, behavioural changes, and neuropsychiatric features, and also evaluate dementia risk factors and family history of dementia. Magnetic Resonance Imaging is helpful in such situations to evaluate for presence of white matter changes [8,10]. In complicated cases like this, Good Medical Practice Guidelines (GMPG) suggest that it is the responsibility of the treating physician to seek a second opinion from other specialties for the benefit of the patient [11]. In this case, interprofessional and multidisciplinary consultation between various subspecialties was essential in order to provide the patient with a correct diagnosis and treatment she needed.

Treatment for subcortical vascular dementia involves a two-pronged approach, namely dementia treatment per se and stroke prevention. Various studies have shown that cholinesterase inhibitors ameliorate cognitive dysfunction in those suffering from vascular dementia, as was done in this case [7]. In terms of stroke prevention, primary intervention targeted at high-risk groups should be done to reduce the incidence of stroke [10]. For example, in J's case, her modifiable risks were diabetes mellitus, hypercholesterolemia, hypertension and lack of exercise. Motivational interviewing techniques and activities targeting weight loss would be beneficial for her and would invariably help her to improve her metabolic parameters. As for secondary prevention, the goal is to reduce the incidence of recurrent stroke, which can cause devastating effects like full dependency on car-

ers, disability and mortality [5]. J suffered from 2 strokes within almost 6 months. Relevant lifestyle changes for her would include a healthy and well-balanced diet, 15 to 30 minutes per day regular exercise for three to five days weekly, and stress management techniques including relaxation exercises. Stroke rehabilitation is another crucial aspect of care and is inherently multidisciplinary and longitudinal, starting from acute hospitalization, progressing to a systematic program of rehabilitation services for those with residual impairments, and continuing after the individual returns to the community, helping them regain, where possible, premorbid productivity [5].

Lastly, it is important also that we, as clinicians, consider the psychosocial and cultural factors that may preclude a diagnosis of dementia. There is stigma towards diagnosing dementia, especially in younger individuals, present equally in patients and in clinicians [12]. This stigma can include multiple facets such as cultural perceptions of illness, as many patients of the Malay ethnic group consume traditional medications that can precipitate hypertension in younger adults [13]. Hence, this combination of stigma and cultural sensitivities can lead unsuspecting clinicians to misdiagnose dementia as depression in younger adults. This can then lead to avoidable morbidity due to biologically inappropriate treatment and increased risk of unnecessary side effects from taking a non-indicated medication.

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Conflict of Interest

The author(s) declared no potential conflicts of interest with respect to the research and authorship of this paper. This paper did not receive any funding source.

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Prikaz slučaja pogrešno dijagnosticirane subkortikalne vaskularne demencije - važnost dobrog poznavanja psihijatrije i pravilnog uzimanja povijesti bolesti

Sažetak - Psihijatrijski simptomi često se mogu previdjeti ako se na njih ne posumnja ili ako se posebno ne istraže. Propuštena psihijatrijska dijagnoza može dovesti do dalekosežnih posljedica u smislu loše kvalitete života i funkcionalnosti pacijenta, što u konačnici utječe na ukupnu kvalitetu pružene zdravstvene zaštite. Prikazana je pacijentica koja se prezentirala simptomima depresije nakon višestrukih moždanih udara te joj je početno dijagnosticiran organski afektivni poremećaj (nakon moždanog udara). Međutim, nakon što je primijećeno da nije došlo do regresije simptoma unatoč uzimanju antidepresiva, daljnjom dijagnostičkom obradom otkrivena je podloga koja više sugerira na subkortikalnu vaskularnu demenciju. Slijedom toga, provedene su detaljne neuropsihološke i neuropsihijatrijske procjene, uključujući NUCOG i daljnja ispitivanja, uključujući MRI snimke mozga prema kojima je sugerirana dijagnoza vaskularne demencije. Ovaj slučaj ilustrira da nedovoljno temeljita procjena i postupak liječenja rezultiraju nepotrebnim morbiditetom, produljuju trajanje bolesti i povećavaju socijalnu i profesionalnu disfunkciju pacijenta. Stoga, nadalje naglašava potrebu uzimanja temeljite povijesti bolesti, provođenja fizikalnog pregleda i relevantne dijagnostičke obrade kako bi se osiguralo isključivanje organske etiologije kod pacijenata s određenim sociodemografskim i kliničkim čimbenicima rizika.

Ključne riječi: demencija; depresija; stanje nakon moždanog udara; neuropsihijatrija; organitet

