

CASE REPORT

Pick's disease and subdural haematoma: A diagnostic red herringAdebayo PB*¹, Taiwo FT², Bakshi F¹, Nur S³¹Department of Medicine, Aga-Khan University, Dar es Salaam, Tanzania²Department of Medicine, Ben Carson Snr School of Medicine, Babcock University, Ilishan-Remo, Nigeria³Department of Radiology, Aga-Khan University, Dar es Salaam, Tanzania***Correspondence: Dr PB Adebayo, Department of Medicine, Aga Khan Hospital, Dar es Salaam, Tanzania. Email: philipab8@yahoo.com; ORCID: <https://orcid.org/0000-0003-0194-6968>****Summary**

Frontotemporal dementia (FTD), otherwise known as Pick's disease, is a clinically heterogeneous group of sporadic and familial neurodegenerative diseases. These conditions are characterized by dementia, behavioural and language dysfunction and loss of executive skills resulting from the degeneration of the frontal and temporal lobes. Although reversible causes of dementia are always sought during the evaluation of patients with progressive cognitive decline, the occurrence of a reversible aetiology may distract from evaluating for neurodegenerative causes of dementia. This report is about a 66-year old man with features of FTD and superimposed chronic subdural haematoma.

Keywords: *Disinhibition, Dementia, Exhibitionism, Frontotemporal disease, Neurodegenerative disease, Subdural haematoma*

Introduction

Frontotemporal dementia (FTD) is a clinically heterogeneous group of sporadic and familial neurodegenerative dementia characterized by behavioural and language dysfunctions and loss of executive skills, resulting from degeneration of the frontal and temporal lobes. FTD accounts for 8-10% of neurodegenerative dementia. ^[1, 2] Available epidemiological data shows a global period prevalence of 0.16-31.04 per 1000 persons and incidence of 0.0-0.3 per 1000 person/year. ^[2] The point prevalence of FTD is estimated at 0.01-4.6 per 1000 persons. ^[3] Aside from anecdotal cases, epidemiological data for FTD in Africa are

scarce. Chronic subdural haemorrhage (CSDH) is a common reversible cause of dementia which affects 1-5.3 per 100,000 individuals annually. The incidence of CSDH is 7.35 cases per 100,000 in adults aged 70-79 years. ^[4] Neurodegenerative dementias and CSDH have advanced age as a common risk factor.

The rule of thumb in the evaluation of individuals with dementia is to search for treatable metabolic causes such as vitamin deficiencies and hypothyroidism and surgical aetiologies such as normal pressure hydrocephalus and CSDH. In rare occasions, these secondary causes of dementia could be red herrings, especially against the background of neurodegenerative aetiology. Therefore, there is

a need for careful evaluation of the temporal profile of patient's symptoms and a careful evaluation of disease phenotypes.

Case Description

A 66-year old man with more than 30 years history of obsessive-compulsive disorder (OCD) presented to Aga Khan Hospital, Dar es Salaam, with declining cognitive function that was difficult to date. The OCD symptoms have been fluctuant while on and off Paroxetine. He developed other behavioural abnormalities such as exhibitionism and disinhibition. He could walk around naked and not be concerned about it. Social misdemeanour such as unprovoked aggression followed by a physical assault of his subject, in addition to repeated agitations, had been noticed by his caregivers. He later developed progressive paucity of speech and urinary incontinence. These symptoms had impaired his activities of daily living.

Three years prior, he developed progressive left hemibody weakness, and he was found to have a right subdural haematoma. History of fall could not be established. He had a burr hole for the CSDH in another facility, but his symptoms did not improve. Instead, the symptoms progressively worsened. He is not known to have hypertension or diabetes mellitus. He smoked cigarettes for most of his adult life but had no family history of dementia.

On examination, his mini-mental state examination (MMSE) was not assessable as he was rather mute and kept to himself. He had a

flexor contracture of the left upper limb. He had gait and eye-opening apraxia in addition to florid frontal release signs. The muscle tone was increased globally, but no other parkinsonian signs were observed.

Magnetic Resonance Imaging (MRI) (Figure 1), showed asymmetric bi-frontal lobe atrophy and temporal lobe atrophy. The posterior cerebral regions were normal for his age. Additionally, a residual right frontoparietal subdural haematoma with no mass effect was also found. He was diagnosed with behavioural variant Frontotemporal Dementia (bvFTD) with concurrent residual subdural haematoma. He is currently on Olanzapine, Fluoxetine, speech and behavioural therapy as well as physical rehabilitation.

Discussion

The Lund/Manchester Consensus Statement published in 1998 for easy clinical recognition of FTD has shown good discrimination between FTD and Alzheimer's disease [4] A study of the criteria, based on 34 patients with pathologically diagnosed Fronto-Temporal Lobar Degeneration (FTLD) among a series of 433 individuals, reported good pre-mortem diagnostic accuracy, with a sensitivity of 85% and specificity of 99%. [5] The patient in the index report demonstrated the core diagnostic criteria for clinical diagnosis of FTD which comprises (i) insidious onset and gradual progression, (ii) decline in social interpersonal conduct, (iii) early impairment in regulation of personal conduct, (iv) early emotional blunting and (v) early loss of insight.

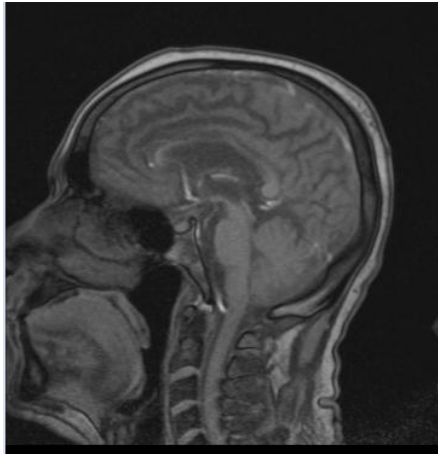


Figure 1 (Image A): Sagittal T1 weighted MRI showing predominant frontal lobe atrophy as well as corpus callosal atrophy anteriorly.

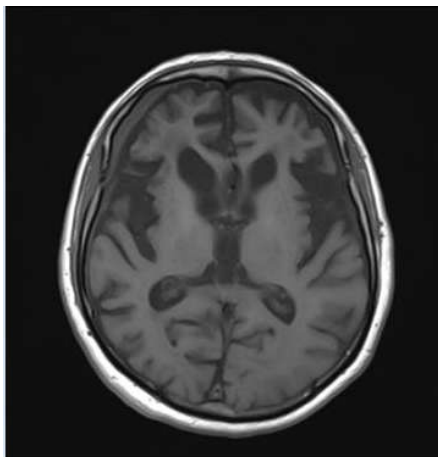


Figure 1 (Image B): Axial T1 weighted MRI showing ventromedial and dorsolateral bi-frontal atrophy with a thin rim of right subdural haematoma without mass effect. This image also showed the prominent Sylvian fissures bilaterally.

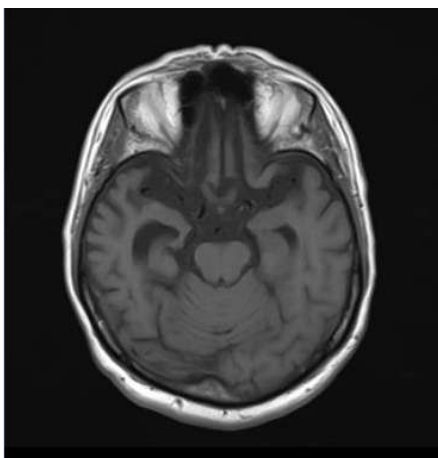


Figure 1 (Image C): Axial T1 weighted MRI showing asymmetric temporal lobe atrophy (right > left).

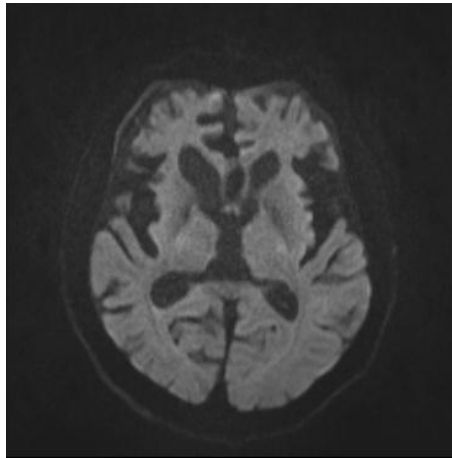


Figure 1 (Image D): Axial DWI image showing no acute infarct or area of restricted diffusion. Features similar to Image B are also noted.

Unfortunately, the finding of a right subdural haematoma in this case, even though worth treating, was a red-herring, as the underlying disease progressed despite the surgical intervention. Even though a clear history of fall was not established in this case, it could be assumed that the CSDH could be secondary to brain atrophy. Brain atrophy expands the subdural space, which increases the frequency of CSDH occurrence. The latest MRI in the index patient confirmed asymmetric bi-frontotemporal atrophy (right more than left), which is unexpected for a disorder solely attributable to his subdural haematoma.

Aside from clinical features in keeping with frontal lobe disorder, the patient also had typical imaging findings in keeping with FTD. Repetitive behaviours such as OCDs are seen in frontal lobe degenerations and could be a presenting feature. [6] Although OCD has been managed in the patient as idiopathic OCD, the accompanying cognitive and behavioural dysfunction with impaired activities of daily living suggests that the OCD could be part of the FTD symptom complex. Hemiparesis has also been described in patients with FTD, [7] the

rationale being the involvement of the descending motor track. [7] The masquerades of long-standing OCD and recent onset subdural haemorrhage delayed the diagnosis of FTD in the index case. The progression of the illness, with accompanying disinhibition, anti-social behaviour and executive dysfunction, points to a frontal lobe dysfunction, in addition to the brain MRI findings.

Epidemiological and outcome data in patients with combined subdural haemorrhage and neurodegenerative dementias are quite scarce. Although it is established that CSDH may also lead to the development of dementia, the mechanisms for this phenomenon and the exacerbation of pre-existing dementia are currently being investigated. One aspect that has gained interest of late is the role of poor dura lymphatic drainage in patients with CSDH. It is postulated that dura lymphatic stasis accompanying subdural haematoma leads to the accumulation of toxic metabolites from neuroparenchymal tissue. [8] It is hoped that future research will provide data on outcomes in patients with combined degenerative and non-

degenerative dementia while unravelling the neurobiological underpinnings.

Conclusion

This case highlights the possibility of a coexisting symptomatic and neurodegenerative aetiologies of dementia. This likelihood should always be kept in view while evaluating patients with progressive cognitive decline even when a reversible aetiology is evident. A careful review of the temporal profile of the patient's complaints as well as neuropsychological monitoring is advocated. Employing the use of existing clinical diagnostic criteria is equally pertinent. No definitive treatment exists for FTD, as the mainstay of management is mainly symptomatic. Nevertheless, it is essential to recognize this entity early because focused care and modulation of the caregiver's expectation are critical in ultimately bridging the treatment gap for the dementias, especially in the Sub-Saharan African settings.

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References

1. Bang J, Spina S, Miller BL. Frontotemporal dementia. *Lancet*. 2015; 386: 1672-1682.
2. Akinyemi RO, Owolabi MO, Makanjuola VA, Ogunseyinde AO, Ogunniyi A. Frontotemporal dementia in a Nigerian woman: case report and brief review of the literature. *Afr J Med Med Sci* 2009; 38: 71-75.
3. Neary D, Snowden JS, Gustafson L, Passant U, Stuss D, Black S, Freedman M, Kertesz A, Robert PH, Albert M, Boone K, Miller BL, Cummings J, Benson DF. Frontotemporal lobar degeneration: a consensus on clinical diagnostic criteria. *Neurology* 1998 ; 51: 1546-1554.
4. Foelholm R, Waltimo O. Epidemiology of chronic subdural haematoma. *Acta Neurochir (Wien)*. 1975; 32(3-4): 247-250.
5. Hogan DB, Jetté N, Fiest KM, Roberts JL, Pearson D, Smith EE, *et al*. The Prevalence and Incidence of Frontotemporal Dementia: a Systematic Review. *Can J Neurol Sci* 2016; 43(S1): S96-S109.
6. Miller BL, Ikonte C, Ponton M, Levy M, Boone K, Darby A, *et al*. A study of the Lund-Manchester research criteria for frontotemporal dementia: clinical and single photon emission CT correlations. *Neurology* 1997; 48: 937-942.
7. Ames D, Cummings JL, Wirshing WC, Quinn B, Mahler M. Repetitive and compulsive behavior in frontal lobe degenerations. *J Neuropsychiatry Clin Neurosci* 1994; 6: 100-113.
8. Schmidtke K, Hiersemenzel LP. Progressive hemiparesis in frontal lobe degeneration. *Eur Neurol* 1997; 38: 105-112.
9. Sahyouni R, Goshtasbi K, Mahmoodi A, Tran DK, Chen JW. Chronic Subdural Hematoma: A Perspective on Subdural Membranes and Dementia. *World Neurosurg* 2017; 108: 954-958.



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