

Neuroimaging Guidelines in Cognitive Impairment: Lessons from 3 Cases of Meningiomas Presenting as Isolated Dementia

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ABSTRACT

We report three cases of large intracranial meningiomas who presented with dementia alone and no accompanying focal neurological deficits. The meningiomas were detected solely as a result of a policy of routinely scanning the brains of patients presenting with early dementia. Dramatic improvements in the cognitive functions were noted following the excision of the tumours in two of the patients (the third passed away in the perioperative period). The implications of these cases with regard to deciding when to order a brain scan for patients presenting with isolated dementia are discussed in this article and a brief review of the relevant medical literature on this topic is also presented.

Keywords: neuroimaging guidelines, isolated dementia, meningiomas, brain scans

INTRODUCTION

The evaluation and management of dementia is critically dependent upon the first step of clinically recognising the syndrome. Accuracy in this step has increased considerably with the establishment of well-defined diagnostic criteria. The DSM-IV⁽¹⁾, for example, states that for the condition to be diagnosed, the patient must have: (a) memory impairment plus (b) one or more of the following: aphasia, apraxia, agnosia, executive dysfunctioning; (c) that the changes observed represent a significant decline from a previous level of functioning and they significantly impair the social and occupational realms of the patient and, (d) that the diagnosis can only be made when the patient is in a fully conscious and attentive state.

With the recognition of the behavioural abnormalities that constitute dementia, the next step involves the determination of the underlying aetiological process. Some of the causes are characterised as being potentially reversible or arrestable; though reported as constituting 5% to 10% of the total causes in most series⁽²⁾, it is nevertheless important to look out for them in view of the dismal prognosis attached to the irreversible category of causes. This work-up for the reversible causes begins with the clinical examination of the patient and then proceeds onto blood investigations and often at this

stage, the question of neuroimaging of the brain (CT or MRI) arises: should it be ordered especially if neither the history nor the physical examination is in any way suggestive of a structural intracranial lesion?

In this paper, we describe three cases seen in a Memory Clinic over a one-year period who had meningiomas presenting as isolated dementia (that is with no other accompanying neurological symptoms or signs). The meningiomas were diagnosed as a result of a policy of routinely scanning the brains of patients who are assessed to be having early dementia. The dramatic reversibility of the cognitive impairment following the excision of the tumours is also highlighted in two of these three cases (the third patient passed away in the perioperative period).

The encounter with these three cases prompted us to review the medical literature regarding neuroimaging guidelines in patients with dementia for detecting potentially treatable intracranial causes which, apart from brain tumours such as meningiomas, also include subdural haematomas and normal pressure hydrocephalus. In particular, we wanted to see if such guidelines too, proposed routine scanning and if they did not, how our patients (with no obvious focal neurological deficits) measured up against the guidelines. The subject is certainly controversial in the medical literature^(3,4) and our findings and conclusions on these matters are presented in the Discussion section.

CASE HISTORIES

The diagnosis of dementia in our three patients was made in accordance with the DSM-IV criteria which have been succinctly outlined above. To supplement the clinical diagnosis, the patients also underwent psychological assessment, some of the test scores of which are shown in Table I. The local normative scores of these tests, which are indicated in the footnotes to the Table, were established from another recently completed study⁽⁵⁾. Psychological tests alone cannot be used to make a diagnosis of dementia, which is essentially a clinical process; in fact it is well-known that highly educated individuals with early dementia can often attain above-average scores on psychometric testing. With these limitations in mind, the psychological tests can however provide a more objective measurement of the cognitive functions as

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well as facilitate the monitoring of their changes over time.

As will be readily noted below, the three patients presented with varying degrees of cognitive impairment; nevertheless, what was common to all of them was that of a significant cognitive decline compared to their usual, previous level of functioning and a change which was also impinging upon the social and occupational realms of the patients. As the text of DSM-IV states, in making a clinical diagnosis of dementia, the defining features such as amnesia, aphasia, apraxia, agnosia and executive dysfunctioning should be translated into a functional context of the patient's daily activities so as to more clearly detect their presence as well as assess their severity. Thus apraxia may contribute to deficits in cooking, dressing or bathing (assuming the absence of any concomitant motor or sensory deficits) and the affected individual may thus be requiring increasing help in such activities. Similarly, executive dysfunctioning qualitatively impairs the patient's work and planning abilities as well as coping with new tasks.

Case 1

This was a 73-year-old Chinese housewife with no significant past medical illness of note. She was fairly well till about a year prior to her visit to the Memory Clinic when her family noticed her to be having progressively worsening difficulties with her memory

(eg the names of her children and other close relatives). Accompanying this were also speech-related difficulties - she was beginning to have more and more problems with the formation of sentences; comprehension of speech was however intact. About 6 months later, there were also calculation deficits and handling of money became a problem, and at around this time the patient also became incontinent of urine and occasionally even of bowels. There were no complaints of visual problems or headaches. Given her cognitive changes, she was no longer able to do any shopping or cooking.

Physical examination was unremarkable. There were no signs of focal neurological deficits and the reflexes were brisk bilaterally. Fundoscopy showed no papilloedema; she could not co-operate with the visual field testing. There was no evidence of urinary tract infection. Psychological testing showed the following (the scores are given in Table I): on the cognitive screening test such as the Elderly Cognitive Assessment Questionnaire (ECAQ)⁽⁶⁾, there was marked impairment suggestive of dementia. Significant deficits were also elicited upon testing the domains of memory (Word-list tests)⁽⁷⁾ and there were also language-related problems noticeable (especially in the Animal-category naming test⁽⁸⁾, which essentially assesses frontal lobe integrity). Visuo-spatial functions as assessed by the drawing of various geometric shapes⁽⁹⁾ appeared adequate. Depression was not detectable.

Table I - Psychological Assessment

Tests	Case 1		Case 2		Case 3
	pre-op	post-op	pre-op	post-op	pre-op
ECAQ	3	9	9	9	7
Word List - immediate recall	7	15	20	25	11
- delayed recall	1	6	7	8	3
- recognition	0	9	0	10	4
Animal Category Naming Test	4	9	12	16	-
2-D Reproduction	0	0	1	0	1

Notes:

ECAQ:

A 10-question cognitive screening instrument with a score of 5 and below suggestive of significant cognitive impairment.

Word List:

A 10-word list given to test verbal memory over 3 learning trials; the test assesses immediate recall after each trial, as well as delayed recall (10 minutes after the 3rd learning trial) and recognition memory (from a 20-word list where the original 10 words are interspersed with another 10 distractor words); the maximum test scores for immediate, delayed and recognition memories are 30, 10 and 10 respectively and the local corresponding median scores are 17, 5 and 9.

Animal Category Naming Test:

This tests an individual's ability to generate names of animals within one minute; local median is 14 names.

2-D Reproduction:

This assesses the ability to copy various geometric figures; errors are scored on this test and the local median is 1.

In the work-up for reversible/arrestable causes of dementia, the dramatic abnormality was a huge tumour in the left fronto-basal region with obstruction at the level of the foramen of Munro and enlargement of the right lateral ventricle. This was soon thereafter excised (biopsy result - meningioma). Post-operatively she made good recovery and with the aid of physical rehabilitation, she was functionally independent at the time of discharge, almost another month later.

When reviewed in the Memory Clinic a year after the operation, significant reversals of the cognitive deficits were noticeable. Her memory, calculation and sentence-forming abilities were back to normal and her functional state too (in both the basic as well as instrumental activities-of-daily-living) had fully recovered. She had also regained bladder continence. The second psychological profile is also shown in Table I, objectively verifying the clinical improvement.

Case 2

This was a 66-year-old former Chinese school-teacher with no significant past medical illness of note. Her main presentation was in the form of progressively worsening short-term memory over the previous one and a half years with frequent misplacing of items at home, forgetting her shopping list etc. Long-term memory was deemed to be intact. Over the preceding 6 months, there were frequent repetitions from the patient. She was also beginning to have more difficulties with her housework and the family noticed that she was no longer planning the variety of dishes, as she would normally do, for mealtimes. Two weeks prior to visiting the Memory Clinic, one episode of urinary incontinence was noted by the family. Apart from this, there were no other cognitive or visual symptoms or headaches and from the functional point of view, in both basic as well as instrumental activities-of-daily-living, she was independent.

Physical examination was essentially normal. In particular there were no focal deficits and fundoscopy showed no papilloedema. Unfortunately, the visual fields were not clinically assessed. There was no evidence of urinary tract infection. Some of her psychological test scores are shown in Table I. On the ECAQ, the patient scored 9 which was not suggestive of significant cognitive deficits. (Abnormality, however, was noted on another cognitive screening instrument, the Blessed Dementia Scale⁽¹⁰⁾, which assesses the behavioural aspects of patients with dementia.) On testing the patient's verbal memory - despite her complaints of forgetfulness - only recognition memory was found to be impaired; both immediate and delayed recall appeared relatively alright. There was mild difficulty with the animal category naming test (especially in view of the patient's own previous educational attainments) but otherwise no significant dysphasia was recorded with the modified Boston naming tests⁽¹¹⁾. Visuo-spatial testing showed that the patient had difficulties with the construction of a cube. There were no clinical features of a major depressive syndrome.

In the work-up for reversible/arrestable causes, the MRI brain scan showed this patient to be having

a large right temporo-parietal tumour with contralateral shift of the midline structures. This was excised soon thereafter (biopsy result - meningioma) and the patient made excellent post-operative recovery being discharged a week after the surgery. A year later when she was again reviewed in the Memory Clinic, the re-evaluation scores showed very good improvement (Table I). The family members confirmed that all of the patient's previous forgetfulness had disappeared and that there were no further episodes of urinary incontinence.

Case 3

This was a 84-year-old Chinese housewife with a past history of hypertension, diabetes mellitus and ischaemic heart disease, who presented with the problem of progressively worsening short-term memory loss as well as on and off urinary incontinence over a period of six months prior to her admission to our Department. She was also noted to have perseveration in her speech and there were times when she would talk nonsense. She was also described to have become more apathetic and more self-centred and she needed, because of her worsening confusion, increasing assistance in daily activities such as bathing and dressing over this same period of time. There were no complaints of headaches.

Physical examination was essentially unremarkable; specifically there were no focal neurological deficits, visual field abnormalities or papilloedema. There was no evidence of urinary tract infection. The patient was lucid and attentive throughout the psychological testing and it showed the following: on the brief cognitive screening test of ECAQ, the patient scored 7, which was not suggestive of significant cognitive impairment. Despite the complaints of forgetfulness, she only had mild difficulties in all the immediate, delayed and recognition memory components of the Word List Test. The patient's main problems were in her language and visuo-spatial abilities; thus there were impairments in the modified Boston naming test and she had difficulties in drawing a cube as well as in other visuo-spatial tests such as the Block Design and Object Assembly subtests of the Revised Wechsler Adult Intelligence Scale⁽¹²⁾. Depression was not clinically detected.

Like the above two cases, the subsequent work-up for reversible/arrestable causes showed an abnormality only in the CT scan of the brain: a large tumour was noted in the right temporal fossa. This was successfully excised (biopsy result - meningioma). Unfortunately, just as the patient was making rapid progress, she developed an acute myocardial infarction on the third post-operative day and soon thereafter she sustained a major cerebro-vascular accident and subsequently demised.

DISCUSSION

The above three cases were seen in the course of the first year of a newly developed Memory Clinic (essentially on early dementia assessment and

management service) in our hospital. They are being highlighted firstly, for their relatively silent (in terms of focal neurological signs) presentation despite the large size of the underlying tumours and secondly, for the dramatic post-operative reversal of their cognitive impairment in two of the cases.

This recovery of cerebral functions also establishes the fact that the meningiomas were actually the cause of the cognitive impairment and not just incidental findings; the possibility of incidental findings needs to be thought of as epidemiological studies generally indicate the incidence of primary intracranial tumours to be peaking at age 55 to 65^(13,14), and if post-mortem findings of asymptomatic tumours are also to be included, the age-specific incidence rate increases with advancing age above 65⁽¹⁵⁾. Moreover, meningiomas are the most common of primary brain tumours in the elderly, representing over 40% of all tumours⁽¹⁶⁾. Since multiple, concomitant illnesses are a characteristic feature of the elderly patient, the possible co-existence of Alzheimer's disease with an incidental cerebral meningioma can certainly occur. However, it must be appreciated that in the setting of patients with dementia and cerebral tumours (especially large ones), the only way of knowing the true role of the tumour is by its excision.

For the clinician therefore, it is important to always consider cerebral neoplasms amongst the list of potentially treatable intracranial causes (which also includes subdural haematomas and normal pressure hydrocephalus). The tumours in our patients were detected solely as a result of a practice of routinely scanning all the ambulatory patients presenting in the earlier stages of possible dementia. Does our experience then justify such a policy of routinely scanning all similar patients?

In attempting to address this problem, investigators abroad had developed clinical prediction rules to guide and define the indications for brain scanning in the diagnosis of dementia^(17,19). In a retrospective comparative study of these guidelines, Martin et al⁽²⁰⁾ found that the clinical rules developed by Dietch⁽¹⁷⁾ had the best sensitivity (87.5%) to pick up surgical lesions. Dietch's rule was as follows: Patients meeting all the criteria (listed below) need not have a brain CT scan:

- dementia present for more than a month
- no history of head trauma in the preceding week
- gradual onset (> 48 hours)
- no history of cerebrovascular accident
- no history of seizures
- no history of urinary incontinence
- no focal signs
- no papilloedema
- no visual field deficits
- no apraxia/ataxia of gait
- no headache

Subsequently, others have suggested that the one month criterion is too short and that it would be wiser to extend the duration of cognitive impairment to

being more than one year for deciding that the CT brain scan is not necessary⁽²¹⁾.

The one criterion from Dietch's that was not consistently applied in all our 3 cases, was that of visual field abnormalities. The visual field assessment by the confrontation approach was only done for one patient and it was unfortunately missed out in another, and the third patient could not co-operate with the assessment. Our own impression of this generally-considered standard neurological examination is that it has significant limitations in some elderly patients especially those presenting with cognitive impairment. Some have difficulties understanding the instructions and are (as in one of our cases) unable to co-operate with the clinical testing. Its true value as one of Dietch's clinical criterion needs an independent future assessment.

All our three patients also presented with urinary incontinence which is listed as one of Dietch's criteria. Before one states that this alone would thus qualify our patients to be receiving a brain scan, a further amplification of the association between urinary incontinence and dementia is necessary. It is a known fact that brain disease can lead to diminution of the cortical inhibitory influence on the micturition reflex and this can result in involuntary detrusor hyperreactivity (DH)⁽²²⁾; however, several studies have also made it clear that DH in dementia has no relationship with any specific cerebral disease^(23,24) and furthermore, DH can also be associated with the normal ageing bladder⁽²²⁾. A wide variety of causes - both within and outside of the lower urinary tract - can therefore be responsible for the presence of urinary incontinence in the setting of cognitive impairment^(25,26). In fact a careful reading of Dietch's original work makes it clear that when the author stated urinary incontinence to be a criterion, it was in the context of detecting an underlying normal pressure hydrocephalus (which has the additional features of gait apraxia - also included in Dietch's list - and dementia). There is no evidence presently that urinary incontinence alone in the setting of dementia is predictive of a structural brain lesion.

In summary, Dietch's criteria may have put our patients under the category of not needing a brain scan, at least at the time of their initial presentations. Though the visual field testing was only done in one patient (and the result obtained was normal), we also have reservations about the true value of this criterion as our (albeit anecdotal) experience to date suggests that amongst the elderly patients with cognitive impairment, a number may not be able to participate meaningfully with its clinical testing.

CONCLUSION

We described three cases of meningiomas presenting with isolated dementia, two of whom recovered dramatically after excision of the tumours. The existing medical literature suggests that Dietch's clinical criteria has the best sensitivity for detecting a structural brain lesion but these criteria may have

certainly put at least one to two of our patients as not requiring a CT brain scan at the time of their initial presentation. Without the brain scans, the meningiomas would not have been detected and the benefits of early excisions may not also have been experienced. Our three cases however do not conclusively prove the invalidity of Dietch's criteria; the value or limitations of these clinical prediction rules require a separate prospectively designed study.

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