PostScript

LETTERS

"Who came with you?" A diagnostic observation in patients with memory problems?

The importance of obtaining collateral history when assessing patients attending the neurology clinic complaining of memory difficulties is well known.^{1,2} Patients developing amnesia in the context of Alzheimer's disease may underplay their difficulties because of cognitive anosognosia, whereas those with purely subjective memory complaints (the "worried well") may overemphasise difficulties. Memory complaint, preferably corroborated by an informant, is one of the suggested diagnostic criteria of mild cognitive impairment (MCI).³ Misdiagnosis of memory complaints may occur when no collateral history is available.⁴

For these reasons, all patients referred to my cognitive function clinic are sent, as part of their clinic appointment letter, a request asking them to bring a relative, friend, or carer from whom additional clinical information may be obtained; this is printed in bold type and in a separate paragraph. Despite this, some patients attend the clinic alone. A study was undertaken to measure the diagnostic value of this observation.

As part of an audit of referrals over a 2 year period (September 2002 to August 2004 inclusive), attendance or non-attendance of a relative or friend at each consultation was noted. Diagnosis of dementia was based on DSM-IV criteria, established by clinical interview, neuropsychological assessment and structural neuroimaging. Diagnosis of dementia subtype (Alzheimer's disease, vascular dementia, dementia with Lewy bodies, frontotemporal dementia) and of MCI followed widely accepted diagnostic criteria. All patients had minimum follow up of 6 months.

Of 183 new referrals seen, 150 (82%; 95% confidence interval (CI) 76 to 88%) followed the written instruction in the clinic appointment letter and attended with a relative, friend or carer; the remaining 33 (18%; 95% CI 4 to 31%) attended alone. In this cohort, 90 patients were diagnosed with dementia and 93 were not demented; three had MCI. Of the 150 patients accompanied to the clinic, 90 (60%; 95% CI 52 to 68%) had dementia; of the 60 not demented, one had MCI. None of the 33 patients attending alone had dementia, although two had MCI.

Hence, if attending the clinic with a relative, friend, or carer (that is, following the instructions given in the appointment letter) were considered a diagnostic test for dementia, it would have a sensitivity of 100% (95% CI 96 to 100%, Wilson method), specificity of 35% (95% CI 26 to 46%), and positive and negative predictive values of 60% (95% CI 52 to 67%) and 100% (95% CI 90 to 100%) respectively. Positive likelihood ratio was 1.55 (95% CI 1.33 to 1.80, log method), judged unimportant, but negative likelihood ratio (0) was large.

Although not absolute, as those unaccompanied patients with MCI might yet evolve to dementia, the period of follow up for some patients is brief, and clinically established diagnoses may require revision (for example, when neuropathological data become available), these findings nevertheless support the belief that attending the neurology clinic alone despite written instructions to the contrary is a robust sign of the absence of dementia.⁵

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References

- 1 Tierney MC, Szalai JP, Snow WG, *et al*. The prediction of Alzheimer disease: the role of patient and informant perceptions of cognitive deficits. *Arch Neurol* 1996;53:423-7.
- 2 Carr DB, Gray S, Baty J, *et al*. The value of informant versus individual's complaints of memory impairment in early dementia. *Neurology* 2000;55:1724-6.
- 3 Petersen RC. Mild cognitive impairment as a diagnostic entity. *J Intern Med* 2004;256:183-94.
- 4 Larner AJ. Getting it wrong: the clinical misdiagnosis of Alzheimer's disease. *Int J Clin Pract* 2004;58:1092-4.
- 5 Matthews WB. *Practical neurology*, 3rd ed. Oxford: Blackwell Science, 1975:111.

Laryngeal abductor paralysis can be a solitary manifestation of multiple system atrophy

Laryngeal abductor paralysis (LAP) and stridor are well known features that occur in over one third of patients with multiple system atrophy (MSA).¹ The pathogenesis of LAP is thought to be crico-arytenoid abductor muscle denervation,² although there is a lack of consistent evidence of motor cell loss in the nucleus ambiguus.^{3,4} More recently, dystonia of the laryngeal adductor muscle has also been proposed.⁴ LAP/stridor usually occurs in the advanced stages of the disease,⁶ and is considered to be a poor prognostic feature.^{1,2} In contrast, some MSA cases have shown LAP initially, with most of these reported by otolaryngology departments.⁵⁻⁷ However, there are no systematic surveys as to the extent to which MSA patients initially present with LAP. We describe the result of a survey of 200 MSA inpatients conducted in a neurology department.

We reviewed the case records of 200 consecutive "probable" MSA patients who met the inclusion and exclusion criteria.⁸ They were 119 men and 81 women, mean age 60 years; 29 had the Parkinsonian form (MSA-P) and 171 the cerebellar form

(MSA-C). Among these, eight patients (4%) (four MSA-P, four MSA-C) were shown to have stridor as the initial manifestation (table 1). Stridor was the solitary manifestation in six of the patients, though it was combined with minimal laryngeal signs in two of these six patients (inspiratory gasp in one; hoarseness in one) and REM (rapid eye movement)-sleep related behavioural disorder ("night terror") in one. In the remaining two patients stridor occurred together with bladder dysfunction or gait ataxia. In the former six patients, stridor was followed by bladder dysfunction in four, constipation in three, tremor/akinesia in one, ataxia in one, and postural hypotension in one. The average interval between the development of stridor and these later symptoms and signs was 3.3 years (range 1 to 6). The average interval between stridor and hospital admission was 5.4 years (1-10). In all eight patients, laryngoscopy confirmed that the stridor was caused by LAP. The grade of LAP at the first admission to our hospital (table 1), according to Isozaki's laryngoscopy classification,⁹ was moderate (abductor paresis during waking; paradoxical adduction during sleep) in three and severe (complete paralysis) in five. Among those patients, continuous positive airway pressure was introduced in three, laser incision of the vocal fold was carried out in one, and subsequent tracheostomy was necessary in five.

In the cases presented, it proved true that LAP/stridor can be a solitary manifestation of MSA. The interval between LAP/stridor and hospital admission was rather long (on average 5.4 years), suggesting that the progression of LAP was not very rapid in those patients. Although the initial presentation of LAP/stridor was not common (it occurred in only 4% of all MSA patients), it is clinically relevant because patients with LAP/stridor but without obvious neurological symptoms may see general physicians or otolaryngologists first. Laryngeal stridor also occurs because of local inflammation or tumours, or from distant causes that affect the vagal nerves, such as upper thoracic or nasopharyngeal carcinoma. If such conditions have been excluded, central neurological causes should be considered. Co-morbid bladder dysfunction (particularly urinary incontinence and post-voiding residual volume of more than 100 ml), postural and postprandial syncope, parkinsonism, and ataxia are all red flags suggestive of MSA.¹⁰ In our eight patients, bladder dysfunction was an early sign and was chronologically correlated with LAP/stridor; this finding is in line with a previous report.⁹ These atypical features for a local laryngeal pathology suggest that further studies of the brain are necessary to confirm the diagnosis of MSA.

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