SHORT REPORT

Olfactory impairment in motor neuron disease: a pilot study

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Abstract

The ability to identify smells was tested in nine males and six females with motor neuron disease (MND) of varying severity, using the University of Pennsylvania Smell Identification Test (UPSIT). The olfactory impairment found in MND patients compared with age and sex matched controls is statistically significant at the 0.005 level. The relationship with Parkinson's disease, with Alzheimer's dementia and the possible aetiological implications of this new aspect of the MND are discussed.

Anatomical and electrophysiological evidence suggests involvement of sensory pathways in Parkinson's disease, Alzheimer's dementia and motor neuron disease (MND).¹⁻⁴ Olfactory dysfunction was shown to occur in Parkinson's disease and in Alzheimer's dementia both in their sporadic form,⁵⁻⁸ and recently in the Parkinson-dementia complex (PDC) in Guam;⁹ MND, very common in Guam and often associated with PDC, has not been investigated. Olfactory function was tested in 15 patients with MND with age and sex matched normal controls. Fifteen patients with MND diagnosed by several neurologists volunteered for the test (nine males and six females, aged 52 to 77 years).

Eight had moderate to severe bulbar involvement, eight were in wheelchairs. Ten controls were colleagues and friends of the examiner; four controls were orthopaedic inpatients for pathology in the lower limbs (fracture of leg, hip replacement, patella, hallux valgus). They were matched for sex and age—five controls were up to one year older and four controls were up to one year younger than the respective patients. Smoking is considered to reduce the ability to smell significantly, but no attempt was made to match for smoking habits. More controls than patients had a history of smoking and for a longer period of time than the patients, eight patients and two controls had never smoked; one patient and two controls were still smoking. Six patients had smoked in the past for a period of two to 49 years, average 27.8 years. The eleven controls had smoked in the past for 10 to 44 years, average 35.2 years.

The University of Pennsylvania Smell Identification Test (UPSIT) commercially

available, was used. This is a standardised microencapsulated odour test consisting of four booklets containing 10 odourants apiece, one odourant per page.^{10,11} The stimuli are embedded in 10 to 50 μ m diameter microcapsules fixed and positioned on each page. A multiple-choice question with four response alternatives for each item is located above each "scratch and sniff" odourised strip. The subject is required to answer one of the four alternatives, even if no smell is perceived (that is, the test is forced-choice). The test¹⁰ has been evaluated on 961 females and 649 males of various ages: the percentile score represents the proportion of subjects correctly identifying each odourant in each five year age group, sex standardised. The UPSIT is reliable (short-term test-retest $r = 0.95$) and sensitive to a variety of olfactory deficits, including those associated with ageing.¹¹

Figure Number of correctly identified smells and age of subjects tested.

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Results

The correctly identified smells according to age of patients and controls are shown in the figure.

Nine patients showed an UPSIT percentile between zero and 30; the score of all but one control was above the fiftieth percentile, eight of these controls scoring between 73–100%. No patient scored above the seventieth percentile. The mean (SD) score on the UPSIT in the MND patients, 25.2 (7.25) was significantly lower than that for the controls 35.15 (4.97). The effects of age as a variable were also investigated as the UPSIT score is known to decrease with age. The correlation of the raw UPSIT score with age was 0.37 ($p < 0.05$). The mean age of the patients was 68.0 years and of the controls 68.3 years (not significantly different) and the group difference remained significant at the 0.005 level even when age effects were covaried out of the analysis. No sex differences in UPSIT scores were found for either the patient or the control group. The patient's group age standardised UPSIT score was significantly lower than the value found in the UPSIT manual standardisation sample.

Discussion

In recent years olfactory deficit was noted in several dementia-related diseases⁵⁻⁸ including Huntington's chorea, Korsakoff psychosis, DAT and PD; in Parkinson's disease the olfactory impairment is equally present in demented and in non-demented patients. No olfactory impairment was found in multiple sclerosis (MS) another chronic neurological disease somewhat similar in geographic distribution to that of MND (Elian and Dean, unpublished data).

The cause and the significance of the impairment of olfactory function in MND is not yet understood. Bulbar involvement and intercostal muscle weakness in MND may cause impaired inspiration; as a result less of the smelling substances may become available at receptor level. None of the 15 patients complained of respiratory dysfunction and seven patients had no bulbar signs. In this pilot study respiratory function was not monitored. Olfactory evoked responses, an objec-

tive measure of olfactory pathway function¹² would eliminate the effect of inspirational impairment, but no appliance is commercially available.

MND—and perhaps PD and DAT—may cause destruction of the olfactory pathways aetiologically unrelated to the disease process; or, environmental agents aetiologically related to MND may directly impair the olfactory pathways. Individuals who subsequently develop PD, DAT or MND may have a vulnerable olfactory system damaged from birth or through previous illness. Alternatively the dysfunction in these diseases may facilitate the entry of an environmental agent which damages the olfactory system.

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