A new case of progressive aphasia due to Pick’s disease


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Introduction

Ever since Mesulam (1982) delineated progressive aphasia has its nosological status been debated. It is presently viewed as one subtype of frontotemporal lobar degeneration. We report a clinicopathological observation with typical Pick’s disease at post mortem.

Neuropathology

Gross pathological findings were consistent with the earlier PET findings in the distribution of atrophy. Histopathological examination revealed extensive neuronal loss and astrocytic gliosis in the temporal and frontal cortex. Numerous tau-positive intracytoplasmatic inclusions (Pick bodies) were detectable within the granule cells of the dentate gyrus and in layers II, V and VI of the frontal, parietal, temporal, insular and cingulate gyrus. Moreover, several ballooned neurons (Pick cells) were present in the deeper cortical layers.

Results of aphasia testing using the AAT (Aachen Aphasia Test) from 1996 to 1999. In 2000, the patient’s language skills had deteriorated so much as to preclude formal testing.

Histopathology from temporal cortex shows ballooned cells (a, HE stain) with basophilic inclusions (b). They stained positive for tau with the AT8-antibody (c).

Pick cells were abundant in the temporal cortex.

Lateral view of the left hemisphere after formaldehyde fixation (the right hemisphere was immediately frozen) and coronal cuts showing cortical as well as striatal and white matter degeneration.

Conclusion

This case adds to the evidence that progressive aphasia is non-specific with respect to aetiology. Typical Pick’s disease may be one of several underlying processes, that in other cases have been identified as Alzheimer’s disease, “dementias lacking distinctive histopathology”, and as tau-negative, ubiquitin-positive inclusions. At present, differentiation is only possible with histology.

Since treatment will eventually depend upon the individual pathology, our observation stresses the need for non-invasive techniques of aetiologic diagnosis in progressive aphasia and other frontotemporal lobar degeneration syndromes.

References


M.-M. Mesulam. Slowly progressive aphasia without generalized dementia.

M.-M. Mesulam. Primary progressive aphasia.


A. Kertesz and D. G. Munoz. Primary progressive aphasia: A review of the neurobiology of a common presentation of Pick complex.

