

MULTIMODAL (MRI, fMRI, SPECT) DATA INTEGRATION IN A CASE OF PRIMARY PROGRESSIVE APHEMIA

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INTRODUCTION

Primary Progressive Aphemia is a rare neurodegenerative disease characterised by progressive dysarthria and dysprosodia, with sparing of other language functions in early phase. Progressive cognitive deterioration, including other language functions, occurs with the worsening of the disease. Primary Progressive Aphemia seems to be associated with focal brain atrophy, mainly of left frontal operculum. A case of Primary Progressive Aphemia is presented, in whom structural (MRI) and functional (fMRI, SPECT) examinations clearly demonstrate prevalence of right hemisphere dysfunction.

CASE REPORT

A 59 years right-handed woman came to our attention one year ago, showing slight difficulty to articulate words, especially when she spoke quickly. That symptom showed a progressive course until now. After extensive language evaluation (BADA)(1), . . . 4/80 mistakes in lexical visual decision task were found. No other neuropsychological or neurological deficits were found; in particular, there was no bucco-facial apraxia, sometimes reported in this syndrome.

IMAGING

Methods.

By a Siemens 1.5 T Magnetom Vision scanner, T1-weighted MPRAGE sequence (128*256*256) was acquired; cerebral cortex was segmented, extracted, and rendered.

Then, 60 EPI volumes (128*128*18) were acquired during covert phonemic fluency task (5 activ+5 rest*6 cycles); registration, smoothing (6 mm FWHM) and statistical analysis of functional images were performed.

In another day, 99mTc-HMPAO SPECT study was performed by a brain-dedicated, high-resolution camera (CERASPECT, Digital Scintigraphics). Perfusional images (128*128*64) were reconstructed by the Coniugate Gradient method. Moreover, improved SPECT images were generated on the basis of structural MR data, thus obtaining high resolution functional images where the anatomical features of MR scan are preserved, without significantly affecting counts.

Finally, fMRI activation voxels were registered with MR-improved SPECT, producing a single multidimensional functional image. All imaging analysis was performed by SPM99, unless MR-improved SPECT, which was reconstructed by a custom-made software.

Results.

The MRI cortical rendering clearly shows that atrophy mainly involves the right frontal lobe, especially the inferior frontal gyrus, that appears to be cranially retracted, and the Sylvian fissure, so as to uncover the insula region.

The MR-improved SPECT shows that hypoperfusion involved wider brain regions than atrophy, especially in the right hemisphere, where the inferior and the medium frontal lobe, the superior parietal lobule and the inferior part of the thalamus appeared to be affected by hypoperfusion.

The most interesting finding is the location of phonemic fluency activation. In fact, the right inferior frontal gyrus showed at the same time the larger cluster of activation and the most relevant atrophy and hypoperfusion.

As a matter of fact, the patient can remember that she began to write with the left hand, but she was corrected in early school years.

CONCLUSION

In clinical setting, the integration of multimodal neuromaging data carries information not provided from each technique. In fact, in this uncommon case of Progressive Primary Aphemia, the main involvement of the right hemisphere is demonstrated by MR anatomo-functional and SPECT perfusional imaging, whereas the fusion of the three methods produces an unique image showing the residual function of the right frontal operculum, at the same time the main area involved by both atrophy and hypoperfusion. By this novel MR-improved SPECT reconstruction method, tracer uptake deficits are easily identified whether due to atrophy or to hypoperfusion.