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Towards an earlier diagnosis of Alzheimer's disease presenting with visuospatial disorders (posterior cortical atrophy)

Vers un diagnostic plus précoce de la maladie d'Alzheimer révélée par des troubles visuospatiaux (atrophie corticale postérieure)

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ABSTRACT

Progressive visual complaints related to visuospatial disorders, and less often to visuoperceptual disorders, may be the presenting and isolated manifestation of a focal degeneration in the posterior cortical areas, called posterior cortical atrophy (PCA). PCA is a clinical syndrome corresponding to a focal variant of Alzheimer's disease in 80% of cases. The predominant dysfunction in the occipitoparietal pathways results in predominant visuospatial disorders, manifesting primarily as dorsal simultanagnosia, alone or associated with other symptoms of Balint's syndrome. PCA is rare and affects young patients who are fully aware of their deficits. Diagnosis of PCA is often delayed, due to insidious onset and development of symptoms, and to poor awareness of the condition in the medical community. An earlier diagnosis requires both better knowledge of PCA among ophthalmologists and neurologists and better recognition of visual complaints, leading to simple bedside tasks that can tackle the syndrome.

RÉSUMÉ

Des plaintes visuelles progressives en lien avec des troubles visuospatiaux (perception, attention et exploration de l'espace), ou plus rarement avec des troubles visuoperceptifs, peuvent être les premières et parfois les seules manifestations d'une forme focale de pathologie dégénérative corticale, intéressant les régions postérieures des hémisphères et définissant le cadre syndromique de l'atrophie corticale postérieure (ACP). La maladie d'Alzheimer rend compte d'environ 80 % des cas d'ACP. L'atteinte prédominante de la voie

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visuelle occipito-pariétale explique la prédominance des troubles visuospatiaux, qui se manifestent avant tout par une simultagnosie dorsale, isolée ou s'intégrant dans le cadre d'un syndrome de Balint. L'ACP est un syndrome rare, affectant des patients jeunes, qui demeurent longtemps conscients de leurs déficits. Le diagnostic est souvent porté tardivement, du fait du développement insidieux des symptômes et de leur méconnaissance de la part des ophtalmologistes et des neurologues. Un diagnostic plus précoce nécessite une meilleure connaissance du syndrome et un repérage des plaintes visuelles, amenant à la réalisation d'un examen clinique simple qui confirme l'existence d'une dysfonction cérébrale.

1. Introduction

Visuospatial disorders cover the deficits in perception, in attention and in exploration of surrounding space. The variety of the deficits, usually intricated, include elementary deficits (disorders in perception of line orientation, spatial localisation, stereoscopic vision, movement), disorders in spatial attention (unilateral spatial neglect, simultanagnosia), deficits in manual and ocular exploration (optic ataxia and apraxia, respectively), which associated with simultanagnosia, define Balint's syndrome. They affect the ''spatial thought'' and complex gestures realization (constructive apraxia, gesture apraxia). These visuospatial disorders schematically stem from a dysfunction of the occipitoparietal pathway (or dorsal, the ''where'' and the ''how'' pathway) of the cortical processing of visual information (Biotti et al., 2012). They are defined as a counterpoint to the visuoperceptual disorders, which concern the analysis and recognition of forms and colours that account for the various forms of visual agnosia and result from damage to the occipitotemporal pathway (or ventral, the ''what'' pathway).

The visuospatial and visuoperceptual deficits participate in the dementia syndrome of Alzheimer's disease (AD). Often difficult to individualise at this stage, they contribute to the topographical disorientation, the misidentification of people, to the disorders of the recognition and handling of objects, as well to hallucinations. The visuospatial, or more rarely visuoperceptual deficits, are the primary manifestations, isolated or largely predominant, of rare forms of a degenerative pathology, defining the framework of posterior cortical atrophy (PCA).

Posterior cortical atrophy is a clinical syndrome, identified by Benson et al., 1988, that is characterised by the progressive development of vision disorders of cerebral origin, and related to a focal dysfunction of the posterior areas of the cerebral cortex, of degenerative origin. Cortical atrophy of the posterior areas of the hemispheres is often visible on magnetic resonance imaging (MRI) or CT scan. The posterior cortical atrophy can also be identified by its functional consequences, such as a regional decreased blood flow (single photon emission computed emission tomography – SPECT), or decreased glucose metabolism (positon emission tomography – PET) using fluorodesoxyglucose in these same areas.

Regarding its aetiology, post-mortem studies have shown that the underlying cortical lesions are those of AD in 80% of cases (Tang-Wai et al., 2004; Renner et al., 2004; Alladi et al., 2007). Although PCA is most often related to AD, due to the elementary lesions, it can be distinguished from the typical form by the absence of memory or language problems, at least initially, and by the distribution of the lesions that predominate in the associative posterior cortex, and spare the internal temporal areas. Other pathologies can be found in the setting of PCA, such as Lewy bodies disease, cortico-basal degeneration subcortical gliosis or Creutzfeldt-Jakob's disease.

Owing to its rarity and to its misleading presentation with visual complaints of insidious development, the diagnosis of PCA is often difficult and delayed (3.8 years for Tang-Wai et al., 2004; 2.7 years for Seguin et al., 2011). The early diagnosis of posterior cortical atrophy is a major challenge, notably because PCA represents a focal pathology, which occurs in young subjects not yet demented and who are conscious of their difficulties.

2. Diagnosis and epidemiology of PCA

Up till now, there are no validated diagnosis criteria. The studies in the literature have used relatively close but sufficiently different criteria to make any comparison difficult. They have hence adopted more or less restrictive criteria regarding the degree of purity of the visual disorders versus other cognitive and particularly memory and dysexecutive disorders, with the acceptance or not of the presence of a homonymous hemianopsia (HH) or of motor problems (parkinsonism, dystonia) and the requirement or not of the presence of cortical atrophy on imaging (Mendez et al., 2002; Renner et al., 2004; Tang-Wai et al., 2004; McMonagle et al., 2006).

Crutch et al. (2012) have proposed diagnostic criteria that include core elements and suggestive elements. The core elements are:

- the insidious onset and progressive development;
- visual disorders without ophthalmological explanation;
- with relative sparing of the episodic memory and awareness of the deficit;
- with evidence on examination of visual agnosia, simultanagnosia, optic ataxia, apraxia, praxic disorders or topographical disorientation;