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Update article

Semiology of neglect: An update

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ABSTRACT

Hemispatial neglect is a common disabling condition following brain damage to the right hemisphere. Generally, it involves behavioral bias directed ipsilaterally to the damaged hemisphere and loss of spatial awareness for the contralesional side. In this syndrome, several clinical subtypes were identified. The objective of this article is to provide a nosological analysis of the recent data from the literature on the different subtypes of neglect (visual, auditory, somatosensory, motor, egocentric, allocentric and representational neglect), associated ipsilesional and contralesional productive manifestations and their anatomical lesion correlates. These different anatomical-clinical subtypes can be associated or dissociated. They reflect the heterogeneity of this unilateral neglect syndrome that cannot be approached or interpreted in a single manner. We propose that these subtypes result from different underlying deficits: exogenous attentional deficit (visual, auditory neglect); representational deficit (personal neglect, representational neglect, hyperschematia); shift of the egocentric reference frame (egocentric neglect); attentional deficit between objects and within objects (allocentric neglect), endogenous attentional deficit (representational neglect) and transsaccadic working memory or spatial remapping deficit (ipsilesional productive manifestations). Taking into account the different facets of the unilateral neglect syndrome should promote the development of more targeted cognitive rehabilitation protocols.

1. Introduction

Unilateral spatial neglect (USN) belongs to the array of spatial cognition disorders. It is more frequent, severe and persistent after a lesion of the right hemisphere [1]. This syndrome is characterized by a behavioral bias consisting in a spontaneous deviation of the head and eyes toward the ipsilesional side (leftward). However, contrarily to other vestibular or parietal behavioral biases (optic ataxia), this USN-related behavioral bias is associated to a defective awareness of space located in the contralesional side (Fig. 1) [2–7]. USN is not caused by a sensory, motor or mental deficit [5,6,8]. USN is a heterogeneous syndrome involving different clinical subtypes that cannot be explained solely by a single exogenous and/or endogenous spatial orientation deficit [9].

The objective of this work is to describe all USN clinical types reported in the literature, and their anatomical lesion correlates according to sensory or motor modalities, location in space (personal space, near and far extrapersonal space, and imaginary space), relevant spatial reference frames (egocentric or allocentric), as well as defective (negative) or productive (positive) nature of the symptoms [10–12].

2. Sensory neglect

Visual neglect (VN) is the most frequent type of USN. It is defined as the incapacity to detect or respond to stimuli presented

Abbreviations: AG, angular gyrus; AF, arcuate fasciculus; CC, corpus callosum; CR, coronal radiations; IC, internal capsule; IFG, inferior frontal gyrus; IFOF, inferior frontal occipital fasciculus; ILF, inferior longitudinal fasciculus; IPL, inferior parietal lobule; MOG, middle occipital gyrus; MTG, middle temporal gyrus; USN, unilateral spatial neglect; VN, visual neglect; MN, motor neglect; RN, representational neglect; PCG, post central gyrus; PHG, parahippocampal gyrus; PMC, premotor cortex; PPC, posterior parietal cortex; PTC, posterior temporal cortex; SMA, supplementary motor area; pre-SMA, pre-supplementary motor area; SLF, superior longitudinal fasciculus; SMG, supramarginal gyrus; STG, superior temporal gyrus; TPJ, temporoparietal junction; TR, thalamic radiations.

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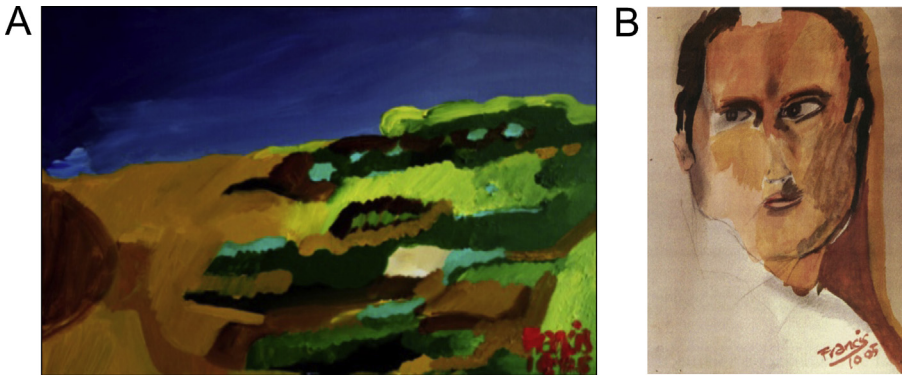

Fig. 1. Oil paintings done by a patient with left USN representing a landscape (extrapersonal space) (A) and a self-portrait (personal space) (B). The picture analysis of the landscape shows neglect on the left side of space as well as a difference in the choice of colors: cold ones (brown, marine blue) on the left side and warm ones (yellow, light green and red) on the right side of the painting (A). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of the article.)

in the contralesional visual field (usually the left) [5]. VN is differentiated from hemianopia, even if sometimes the distinction is difficult, VN can sometimes mimic the clinical picture of pseudo-hemianopia. In a series of 154 left brain-damaged patients and 144 right brain-damaged patients, Sterzi et al. showed that the incidence of hemianopia was greater after a right hemispheric lesion (18%) than a left one (7%) [13]. This difference can be explained by the existence of pseudohemianopia caused by right hemisphere-related VN. The Line bisection test contributes to the differential diagnosis between VN and hemianopia. In case of VN, the bisection bias is directed toward the lesioned hemisphere, whereas it is directed toward the contralesional side in case of hemianopia. When both deficits are associated, the bias is then directed toward the ipsilesional side and is in fact more severe [14,15].

Anatomical lesion correlates of VN are the temporoparietal junction (TPJ), which includes the inferior parietal lobule (IPL) (Brodmann area (BA) 39 or supramarginal gyrus (SMG) and BA 40 or angular gyrus (AG)) [16–20] and the superior temporal gyrus (STG) [22,23], as well as the promotor cortex (BA 6, 7 and 41) [8,21], the basal ganglia and the thalamus [23–25]. Recent studies showed that VN can result from disconnections of the intrahemispheric white matter pathways linking the parietal areas to the frontal ones, involved in spatial selective attention [26,27]: the 2nd and 3rd branches of the superior longitudinal fasciculus (SLF II and III), the anterior segment of the arcuate fasciculus (AF) [28–33], the inferior longitudinal fasciculus (ILF) [20] and the inferior occipitofrontal fasciculus (IFOF) [34]. Furthermore, damage to the interhemispheric connections, especially in the posterior part of the corpus callosum (forceps major), also contributes to VN chronicity (Table 1) [35–38].

Auditory neglect is defined as inattention to sounds or verbal stimuli stemming from the hemispace contralateral to the lesion. When several interlocutors are present, the subject talks to the person located farthest to the right. This deficit can be associated with sound localization impairments, i.e. sound sources are perceived as coming from the lesioned side [39,40]. Bellmann et al. reported four observations in favor of a double-dissociation between these two deficits: during a standard dichotic listening task, two patients presented with a left ear extinction but no auditory spatial mislocalization, whereas the other two patients showed the reverse dissociation. In the first two patients, the auditory spatial attention deficit was due to a subcortical lesion in the basal ganglia whereas the other two patients presented with auditory spatial representation deficit due to a cortical lesion involving the prefrontal cortex, STG and IPL [41]. These two deficits are the consequences of damage to two distinct auditory pathways: auditory extinction is the consequence of an affection to the ventral pathway involved in sound recognition (“what”) (anterolateral part of the STG and IFG), whereas the auditory spatial localization deficit is caused by an affection of the dorsal pathway involved in sound localization (“where”) (posterior part of the STG, posterior and inferior parts of the PG) [42].

In somatosensory neglect patients ignore tactile, thermal or painful stimuli applied to the contralesional body side. Patients can also make stimuli localization errors or mistakes in evaluating the spatial position of their limbs [5]. Somatosensory neglect must be differentiated from primary somatosensory deficits, as evidenced in right brain-damaged patients by the regression of the deficit after the vestibular stimulation test (consisting in irrigating cold water in the left external auditory canal), whereas the same vestibular stimulation performed on the opposite side has no impact in left brain-damaged patients [43]. These results cannot be explained by the regression of non-spatial or lateral deficits of arousal. They suggest the involvement of a spatial factor related to the right hemispheric egocentric representations of the body affected by vestibular stimulation (see review in [44,45]). Similarly to VN, somatosensory neglect can mimic the clinical picture of pseudo-hemianesthesia [5], evidencing the predominance of somatosensory deficits after damage to the right hemisphere (37% of right brain-damaged patients vs. only 25% of left brain-damaged patients) in the study by Sterzi et al. [13].

This spatial neglect can also concern proprioception. Vallar et al. [43,46] conducted an experiment in right brain-damaged patients with USN that required subjects to evaluate the orientation of their upper limbs, which were set passively into different positions, in the vertical and horizontal planes. These patients presented a perceptual deficit of position sense for their contralesional limb, whether it was positioned in their contralesional or ipsilesional side of space, in the absence of primary sensory deficit to the right hemibody. This bodily spatial deficit was decreased by horizontal optokinetic stimulation directed on the left side, and aggravated by stimulation toward the opposite side, thus suggesting the possibility that one could, via sensory manipulation, influence the processes contributing to the construction of full-body spatial representation.

Vestibular information is the most contributive to the elaboration of egocentric spatial representations, and its manipulation leads to the most spectacular effects on NSU regression. Vestibular afferents are integrated at the level of the right parietal insular cortex, which is frequently damaged in middle cerebral artery (MCA) stroke [43,45]. This lesion can explain the “vestibular” symptoms of USN [16], i.e. behavioral bias directed toward the lesion, asymmetry of vestibular-ocular responses and associated disruptions of spatial reference frames [7,16].

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