White Matter Hyperintensities: Initial Assessments

Hiperintensidades da Substância Branca: Avaliações Iniciais

Eliasz Engelhardt[1], Felipe Kenji Sudo[2], Gilberto Sousa Alves[3], Denise Madeira Moreira[4]

ABSTRACT

The white matter hyperintensities (WMH, leucoaraiosis) represent the most common kind of ischemic vascular lesion of the white matter due to small vessel diseases, and occurs frequently in the elderly. Consequent to the neuroimaging identification arouse the need for their assessment. The group of Fazekas proposed a systematized semi-quantitative visual scale to score such lesions where two parameters were considered, extent and localization. The original scale was further modified, to a simplified version. Although other more complex scales have appeared, researchers remarked that the relatively simple Fazekas scale, in comparison to the complex ones and to volumetric measures, appeared to be sufficient when analyzing relationships between clinical parameters and WMH load in a clinical setting.

Keywords: hyperintensities, leucoaraiosis, visual scale

RESUMO

As hiperintensidades da substância branca (HSB, leucoaraiose) representam o tipo de lesão isquêmica mais comum da substância branca decorrente de doenças de pequenos vasos e ocorre frequentemente em idosos. Consequente à identificação por neuroimagem surgiu a necessidade de sua avaliação. O grupo de Fazekas propos uma escala visual semiquantitativa sistematizada para pontuar tais lesões, onde foram considerados dois parâmetros, extensão e localização. A escala original foi modificada para constituir uma versão mais simplificada. Embora outras escalas mais complexas tenham aparecido, pesquisadores comentaram que a relativamente simples escala de Fazekas, em comparação às mais complexas e a método volumétrico, mostrou-se suficiente quando é analisada a relação entre parâmetros clínicos e a carga de HSB em um cenário clínico.

Palavras-chave: hiperintensidades, leucoaraiose, escala visual

1.Neurology Institute Deolindo Couto – Institute of Psychiatry – Federal University of Rio de Janeiro – Rio de Janeiro – RJ – Brazil; 2.D'Or Institute for Research and Education - Rio de Janeiro – RJ – Brazil; 3.Medicine I Department – Federal University of Maranhão – S. Luiz – MA – Brazil; Post Graduation in Psychiatry and Mental Health, PROPSAM, Federal University of Rio de Janeiro - Rio de Janeiro – RJ – Brazil; 4.Neurology Institute Deolindo Couto – Neuroradiology Unit - Federal University of Rio de Janeiro – Rio de Janeiro – RJ – Brazil.

Correspondence: e-mail: eliasz@centroin.net.br

Conflict of interests: none.

Financial support: none.

INTRODUCTION

The white matter, which makes up around one half of the human brain volume, is a very frequent target of "small vessel diseases" (SVD), which appear under varied kinds of ischemic and hemorrhagic lesions[1,2,3]. The ischemic types comprise the "white matter hyperintensities" (WMH), and the "lacunar infarcts" (lacunes), as visualized on magnetic resonance imaging (MRI)[2,3]. The WMH represent the most common kind of lesion of the white matter, and occurs frequently in adults over 65 years old with a prevalence rate of ~60 - 80% in the general population[4]. Such lesions are even more extensive in those with vascular or Alzheimer's disease type of dementia, where they may reach ~90% when compared with cognitively normal older adults, suggesting its role in dementia pathogenesis and neurocognitive dysfunction[5].

Such WMH, as coined by Zimmerman and collaborators, were initially observed on T2 sequence of MRI (1.5 T), introduced in the beginning of 1980s as an increase of the signal of this change of the white matter[6,7]. A better visualization of WMH was achieved on FLAIR (fluid-attenuated inversion recovery) sequences that appeared (1985)[6,8], becoming soon one of the preferred sequences for this purpose[9]. Such white matter changes can also be detected on CT generated images, introduced in the late 1970s, as decreased densities of the white matter ("white matter hypodensities"), although with a lesser sensitivity in comparison to MRI[6,10]. Concurrently, the term "leucoaraiosis" (rarefaction of the white matter [*leuko*=white and *araios*=rarefaction]), "meaning a diminution of the density of representation of the white matter", was coined by Hachinski and collaborators (1986), to designate these white matter changes, which appear as hypointense (hypodense) on CT and hyperintense on MR[11,12].

SCORING THE WMH

Consequent to the MRI findings arouse the need to assess the magnitude and the localization of such WMH, and the researchers also recognized the association of the load of such lesions with cognition in normal aging, and pathological states. Zimmerman and collaborators (1986)[7] have proposed an initial scoring method, aiming mainly the periventricular region. Next, Fazekas and collaborators proposed a systematized scoring scale of such lesions, which became to be known as the "Fazekas scale", where he considered two parameters, extent and localization (1987)[10]. The original Fazekas scale (oF) comprises a semi-quantitative visual assessment of these lesions in two locations: [a] "periventricular hyperintensities" (PVH), following the contour of the lateral ventricles with variable breadth and degrees of irregularity, and [b] "deep white matter hyperintensities" (DWMH), extending from the paraventricular region through the centrum semiovale, and also the deep white matter, with a 4-point grading of each region (total range 0 to 6) (1987)[10] (Box) (Figure).

Much later, the LADIS study group introduced a modified Fazekas scale (mF), "taking into account only deep and subcortical white matter lesions" (DSWM), with a 4-point severity scale (total range 0 to 3) (Box) (Figure). Their initial interest was to correlate the leucoaraiosis load with disability development (cognitive, functional, motor, autonomic) over a time period, and was used successfully by this kind of research (2001-2015)[9,13,14].

Nevertheless, it was felt by numerous researchers that, despite the practicality of the simplification, the original Fazekas' rating presented some advantages, offering more detailed information considering the localization of the lesions, considering that there are overt differences between the PVH and the DWMH, regarding the affected underlying white matter structures[12,15,16,17,18]. However, it should be pondered, that in the subjects with advanced WMH the PVH and DWMH often adjoin each other. Thus, a stringent distinction of these two regions appears to be disputable in advanced stages[12].

The two visual scales that appeared later were more complex, and also distinguished the differential localization of the lesions, the "Scheltens scale" (range 0 to 84), which discriminates the PV and the DWM regions, and additionally the basal ganglia and infratentorial structures (1993)[19], and the "Wahlund (ARWMC) scale" (range 0 to 30), which differentiate also right and left hemisphere lesions, besides those of the basal ganglia (2001)[20].

Worth full to observe, that the LADIS group, which besides the modified Fazekas scale used also complex ones (Scheltens, ARWMC) and volumetric measurement, remarked that the relatively simple modified Fazekas scale appeared to be sufficient when analyzing relationships between clinical parameters and WMH load in a clinical setting [21,22,23].

Box. Characterisitcs of the Fazekas scales[9,10].

ORIGINAL FAZEKAS		MODIFIED FAZEKAS
PVH	**DWMH**	**DSWM**
0=absent	0=absent	- - -
1=caps or pencil-thin lining	1=punctate foci	1=[mild] punctate lesions with a maximum diameter of a single lesion below 10 mm and areas of 'grouped' lesions smaller than 20 mm in any diameter
2=smooth halo	2=beginning confluence of foci	2=[moderate] single lesions between 10-20 mm in any diameter, areas of 'grouped' lesions more than 20 mm in any diameter, no more than 'connecting bridges' between individual lesions
3=irregular hyperintensities extending into the deep white matter	3=large confluent areas	3=[severe] single lesions or confluent areas of hyperintensities of 20 mm or more in any diameter
total score= [(0 – 3) + (0 – 3)] = 0 - 6		**total score = 0 - 3**

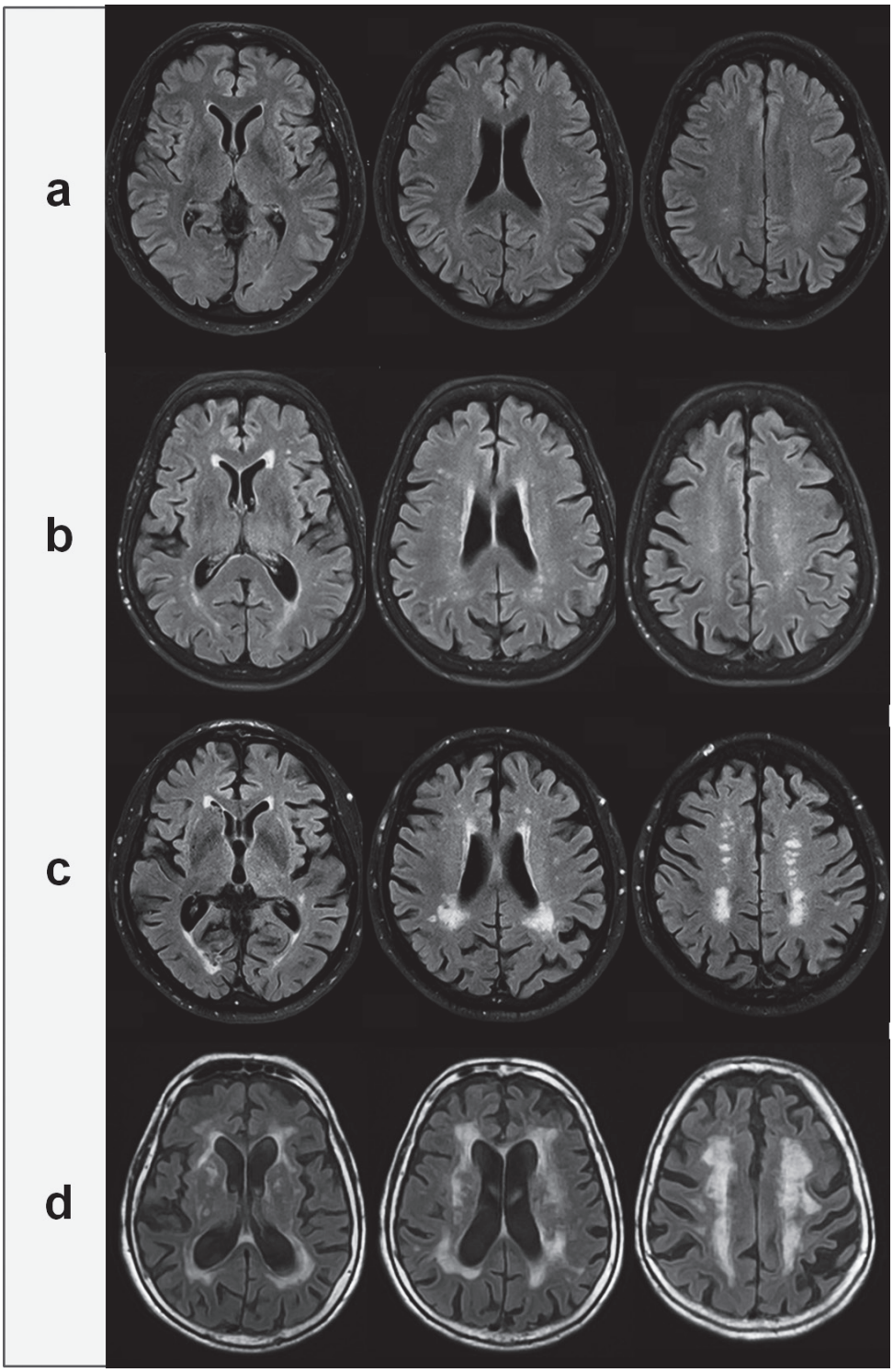

Figure. Fazekas scoring scales. [oF=original Fazekas, mF=modified Fazekas].
MRI – FLAIR sequence: axial sections at basal ganglia, lateral ventricles and supraventricular levels for visualization of WMH [PVH and DWMH].
Grades of severity:
a=absent [oF=0+0, mF=0], b=mild [oF=1+1, mF=1], c=moderate [oF=2+2, mF=2], d=severe [oF=3+3, mF=3]

COMMENTARIES

The advent of the contemporaneous neuroimaging machines, which appeared in the late 1970s (CT), and early 1980s (MRI) permitted for the first time to analyze the brain structure, in normal and pathological states (tumors, vascular diseases, among others), in a direct manner[6]. In this way, the WMH (leucoaraiosis), a result of SVD were identified, followed by the relatively simple visual scoring scale, proposed by Fazekas and collaborators, adopted by a large number of researchers of the clinical neuroimaging field, in its original form or in a modified version[9,10]. It was followed by more complex visual scoring scales, and also by volumetric measures[9,24]. However, it was remarked that the Fazekas scale, even in its modified form, appeared to be sufficient to analyze, in a clinical setting, the relationships between the WMH load and the varied disabilities that developed over a period of time[21,22,23].

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