Isolated Prosopagnosia in Multiple Sclerosis: A Case Study

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Background

Prosopagnosia, commonly known as face blindness, is a rare neurocognitive symptom characterized by the **inability to recognize familiar faces**. It is usually due to **right or bilateral temporo-occipital damage**, particularly in the **fusiform face area (FFA)**. Isolated prosopagnosia is rare in **Multiple Sclerosis (MS) patients** who **usually present combined cognitive symptomatology**.

Hereby, we describe a **clinical case** of a woman affected by **Relapsing Remitting (RR) MS** and presenting as **isolated prosopagnosia**.

Case Report

I.L. female, right –handed, 34 years old, 18-years of education

- ***Medical History***

  Positive family history for MS (i.e. mother and aunt)

  Three episode in remote medical history:

  - **Acute visual loss** in the right eye with full recovery (2020)
  - **Acute visual loss** in the right eye and **NORB** diagnosis (2021)
  - **Acute loss of sensitivity** in the right side and **RR-MS** diagnosis (2022)
  - Start of **Natalizumab** treatment (2022)

  Acute onset of cognitive impairments characterized by **short-term memory** problems; deficits of **face recognition**; deficits of **topographical memory**

- ***Neurological Examination***

  Mild right painful hypoesthesia, EDSS = 1.0

- ***1,5T Brain MR imaging***

  Multiple contrast-enhancing lesions predominantly in the **right hemisphere**, located in the **superior temporal gyrus, the dorsolateral prefrontal area**, and in the **left temporo-polar, occipital, and semioval areas** (**Figure 1**)

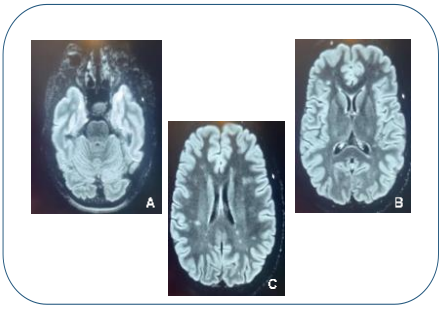

Figure 1. T2-FLAIR MRI sequences showing multiple lesions located in the left temporal pole (A), right dorsolateral prefrontal region (B) and semioval areas (C)

Neuropsychological Evaluation

- MS Quality of Life (MSQoL-29) Physical Health score = 64.48/100
- MS-QoL Mental Health score = 42.41/100
- Fatigue Severity Scale (FSS) score = 59/63

- Rao's Brief Repeatable Battery (BRB) showed normal performances in each tasks (**Table 1**)

	Raw Score	Adjusted Score	Outcome	Cut-Off
Rao's Brief Repeatable Battery				
SRT - LTS	36.14	-	Normal	< 23.3
SRT - CLTR	22.36	-	Normal	< 15.5
SPART	18.9	-	Normal	< 12.7
SDMT	43.23	-	Normal	< 37.8
PASAT3	16.49	-	Normal*	< 28.4
PASAT2	21.78	-	Normal	< 17.1
SRT – Delayed Recall	6.87	-	Normal	< 4.9
SPART – Delayed Recall	6.28	-	Normal	< 3.6
WLG	17.87	-	Normal	< 17

Table 1. Performances at single Rao' s BRB cognitive tasks

- In-depth neuropsychological assessments showed **semantic fluency** deficit and severe **face and emotion recognition** impairments (**Table 2**)

	Raw Score	Adjusted Score	Equivalent Score	Cut-Off
Learning & Memory				
Babcock's Story				
Immediate Recall	4.6	3.52	-	-
Delayed Recall	7.7	31.9	-	-
Total elements	12.3	9.88	2	≤ 4.75
Famous People Recognition Task – 12 Items				
- Naming	3	0	0	≤ 53
- Identification	25	-	-	-
- Semantic Score	2.08	-	-	≥ 0.50
Attention & Executive Functions				
Trail Making Test - Part A	46	61.1	2	> 93
Trail Making Test - Part B	133	190.2	1	> 282
Trail Making Test – B-A	87	129,15	1	>186
Stroop – Time	14	25.3	3	≥ 36.91
Stroop – Errors	0	0	4	≥ 4.24
Language				
Verbal Fluency (FAS)	34	26.7	2	≤ 17.77
Semantic Fluency	32	21.5	0	≤ 28.34
Alternate Fluency	26	13,34	1	≤ 12.7
Picture Naming Task (From CaGi)	47	47.9	4	≤ 41,48
Social Cognition				
Ekman Test-Global Score	40	34,24	0	<37,46
Face Test	14	25,45	2	<22.68
Visuo-Perceptual Motor Function				
Benton Unfamiliar Faces	41	-	-	< 41
Famous People Recognition Task (FA-REC)				
- Familiarity score	36	35.08	0	< 47.23
- Semantic score	68	62.44	0	< 69.41
- False alarms	4	4.78	1	> 8.41

Table 2. Performances at the in-depth neuropsychological evaluation

Discussion

Our case reports a peculiar clinical presentation of a relapse in a **RR-SM patient with positive family history of MS** due to a **selective damage of neural networks critical for semantics and face processing.** Our findings support the need of **in-depth neuropsychological assessment** in MS patients presenting with selective cognitive impairments for a better phenotyping and planning of **tailored and timely intervention including personalized cognitive rehabilitation strategy**

References

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