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Pure allocentric hemineglect and Bálint-Holmes syndrome as presentation of Posterior Cortical Atrophy (PCA): a case study.

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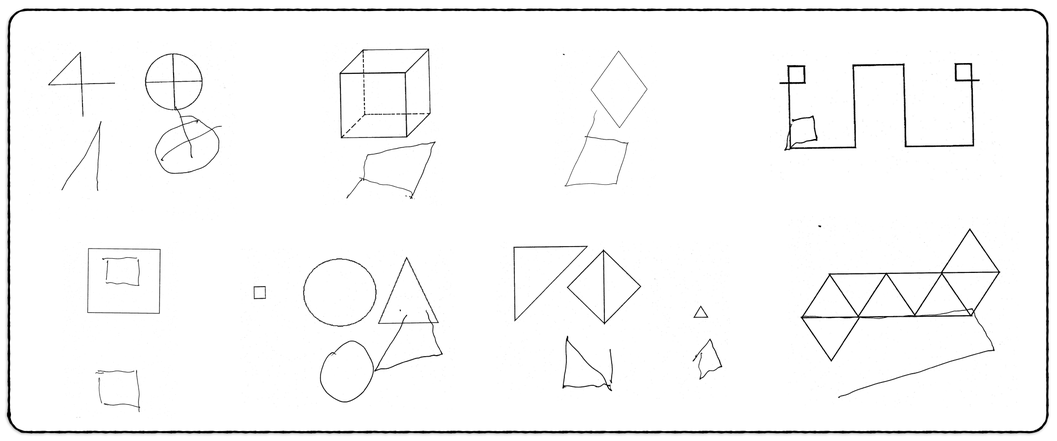
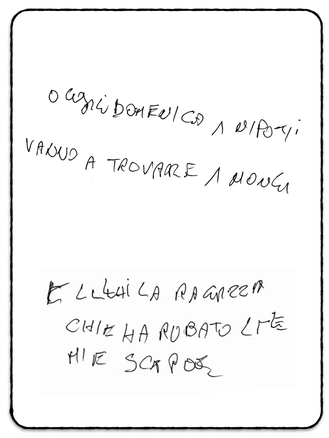
Background: Posterior cortical atrophy (PCA) is a rare and often presenile dementing illness characterized by progressive decline in visuo-spatial and visuo-perceptual abilities.
The neurodegeneration affecting parietal, and occipito-temporal cortices is attributable to Alzheimer's disease in most patients, however alternative underlying diseases as corticobasal degeneration, Lewy bodies disease, and prion disease have been reported.
The spectrum of clinical signs of PCA reflect dysfunction of the dorsal (occipito-parietal) or the ventral (occipito-temporal) visual streams. Lesions of the dorsal system produce progressive impairment of visuo-spatial functions, features of Bálint-Holmes syndrome (simultanagnosia, oculomotor apraxia and optic ataxia), of Gerstmann syndrome (dysgraphia, dyscalculia, finger agnosia and left-right disorientation) and dressing apraxia, while lesions of the ventral system produce progressive impairment of visuo-perceptive functions (identification of objects, symbols, words, or faces).
Not all the patients with PCA have atrophy on conventional neuroimaging and, owing to the typically presenile onset and the unusual symptoms presentation, patients often face considerable delays in diagnosis [1].

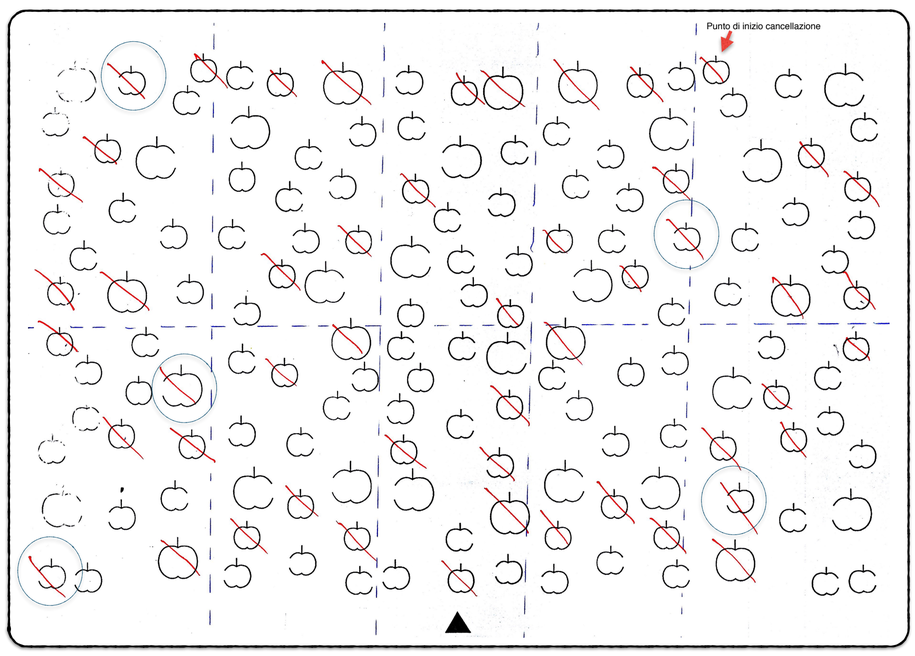
Case report: A 56 year old right-handed man, recently referred to us by an ophthalmologist, presented with 3 years history of progressive visuo-spatial dysfunction initially misattributed to ophtalmologic disease (maculopathy). His first symptoms were difficulty in driving, dressing himself and a peculiar inability to perceive more than an object at a time.
Neurological examination and a first CT scan were unremarkable.

Results and discussion: At the neuropsychological assessment the patient showed no defect in visual recognition and naming of colors, objects, symbols and single words while he had severe difficulties in line length and orientation judgement, angle width judgement, point position identification, mental rotation, and in all the constructional tasks. He also had simultaneoagnosia (inability to perceive more than a single object at a time), optic ataxia (inaccuracy of visually guided arm movements) and dressing apraxia.
Language examination showed the presence of dysgraphia with no dyslexia, dyscalculia or any problem with spoken language.
The patient performed all the cancellation tasks for hemineglect without showing any space asymmetry but a test aimed to differentiate allocentric (object-based) and egocentric (space-based) forms of neglect [2] revealed the presence of left pure allocentric neglect. The patients had no sing of personal neglect or visual or tactile extinction on double stimulation.
All the other cognitive functions (memory, information processing, praxis, and executive functioning), assessed using tasks that minimize visual demands, resulted fully intact.
He was alerted, oriented, collaborative and completely aware of the disturbances.
The patient had difficulties in visuo-spatial abilities, a complete Bálint-Holmes syndrome and some features of Gerstmann syndrome (dysgraphia and finger agnosia) but no difficulties in visuo-perceptive abilities. This dissociation suggests a relatively intact ventral visual stream ("what") and disruption of the dorsal stream ("where").
Cerebra MRI examination with volumetric scans showed inferior parietal lobule atrophy more pronounced on the right hemisphere while hippocampus appeared bilaterally preserved. Cerebral [^{18}F]FDG-PET showed bilateral temporo-occipital and parietal hypometabolism, more pronounced on the right hemisphere.
Our patient fulfilled the criteria for PCA [3].
A cerebrospinal fluid analysis for neurodegenerative diseases screening was performed: a preliminary biochemical datum is a slight elevation of total tau; other determinations are in course.

TEST	SUBTEST	P.G.	P.C.	P.E.	CUT-OFF	ESITO	DATI NORMATIVI
VALUTAZIONE MEMORIA A BREVE E A LUNGO TERMINE							
Digit Span	AVANTI	6	6,04	4	< 4.26	NORMA	Monaco et al., 2013
Test delle 15 parole di Rey	RIEVOCAZIONE IMMEDIATA	43/75	43,7	4	< 28.53	NORMA	Carlesimo et al., 1996
	RIEVOCAZIONE DIFFERITA (15')	9/15	9,2	4	< 4.69	NORMA	Carlesimo et al., 1996
VALUTAZIONE MEMORIA SEMANTICA							
Batteria per le Agnosie	ASSOCIAZIONE SEMANTICA INTRACATEGORIALE	20/20	na	na	< 17.92	NORMA	Barletta-Rodolfi, 1996
	TEST SEMANTICO BREVE (LAIACONA)	239/240	na	na	< 234.09	NORMA	Barletta-Rodolfi, 1996
VALUTAZIONE DI MEMORIA DI LAVORO E INFORMATION PROCESSING							
Digit Span	INDIETRO	4	4,10	4	< 2.65	NORMA	Monaco et al., 2013
	RAPPORTO	na	0,67/1	2	< 0.48	NORMA	Monaco et al., 2013
PASAT	PASAT 3s	48/60	53,77	na	< 28.4	NORMA	Amato et al., 2006
VALUTAZIONE DI ATTENZIONE E FUNZIONI ESECUTIVE							
Multiple Features Target Cancellation (MFTC)	PUNTI	0/13	na	na	na	na	Marra et al., 2012
	FALSI ALLARMI (ERRORI)	0/67	0	na	> 2.77	**DEFICITARIO**	Marra et al., 2012
	TEMPO DI ESECUZIONE	420 sec.	413,97	na	> 135.73	**DEFICITARIO**	Marra et al., 2012
	ACCURATEZZA	0/1	na	na	<0.869	**DEFICITARIO**	Marra et al., 2012
Matrici Attentive (Visual Search)		30/60	25,5	0	< 31	**DEFICITARIO**	Spinnler e Tognoni, 1987
Test di Stroop	EFFETTO INTERFERENZA ERRORI	0	0	4	≥ 4.24	NORMA	Caffarra et al., 2002
	EFFETTO INTERFERENZA TEMPO	38,5	35,5	1	≥ 36.92	LIMITI DI NORMA	Caffarra et al., 2002
Fluenze Verbali	FONEMICA	48	53,91	4	< 17.77	NORMA	Costa et al., 2014
	SEMANTICA	33	37,34	3	< 28.34	NORMA	Costa et al., 2014
	ALTERNATA (TASK SWITCHING)	38	43,28	4	< 12.70	NORMA	Costa et al., 2014
	RAPPORTO	0,938	0,968	4	< 0.38	NORMA	Costa et al., 2014
Test delle Analogie Semplici		19/20	20	na	< 15.1	NORMA	Carlesimo et al., 1996
VALUTAZIONE DEL LINGUAGGIO							
Esame Neuropsicologico per l'Afasia	RIPETIZIONE DI PAROLE	10/10	10	na	< 8.8	NORMA	Capasso e Miceli, 2001
	RIPETIZIONE DI NON-PAROLE	5/5	5	na	< 2.0	NORMA	Capasso e Miceli, 2001
	RIPETIZIONE DI FRASI	3/3	3	na	< 3.0	NORMA	Capasso e Miceli, 2001
	DENOMINAZIONE DI NOMI	10/10	10	na	< 8.2	NORMA	Capasso e Miceli, 2001
	DENOMINAZIONE DI VERBI	10/10	10	na	< 6.1	NORMA	Capasso e Miceli, 2001
	COMPRENSIONE UDITIVA DI PAROLE	20/20	20	na	< 18.4	NORMA	Capasso e Miceli, 2001
	COMPRENSIONE UDITIVA DI FRASI	14/14	14	na	< 11.6	NORMA	Capasso e Miceli, 2001
	LETTURA DI PAROLE	10/10	10	na	< 6.4	NORMA	Capasso e Miceli, 2001
	LETTURA DI NON PAROLE	4/5	4	na	< 4.0	NORMA	Capasso e Miceli, 2001
	SCRITTURA DI PAROLE	6/10	5.4	na	< 6.3	**DEFICITARIO**	Capasso e Miceli, 2001
	ADDIZIONI	3/3	3	na	< 2.2	NORMA	Capasso e Miceli, 2001
	SOTTRAZIONI	2/3	1.8	na	< 1	NORMA	Capasso e Miceli, 2001
	MOLTIPLICAZIONI	3/4	2.4	na	< 1.4	NORMA	Capasso e Miceli, 2001
VALUTAZIONE DELLE ABILITÀ VISUO-PERCETTIVE							
Denominazione su stimolazione Visiva		48/48	48	4	≤ 41.48	NORMA	Catricalà et al., 2013
Street Completition Test		1/14	0	0	< 2.25	**DEFICITARIO**	Spinnler e Tognoni, 1987
Batteria per le Agnosie	TEST DI EFRON	20/20	na	na	< 16.51	NORMA	Barletta-Rodolfi, 1996
	TEST X-O-N DI WARRINGTON E TAYLOR	30/33	na	na	< 29.03	NORMA	Barletta-Rodolfi, 1996
Batteria di Screening Neuropsicologico dei Colori	DENOMINAZIONE DI COLORI SU INDICAZIONE	29/30	na	na	< 24	NORMA	Della Sala et al., 1996
	INDICAZIONE DI COLORI SU DENOMINAZIONE	30/30	na	na	< 26	NORMA	Della Sala et al., 1996
	MEMORIA VERBALE DEI COLORI DI OGGETTI	30/30	na	na	< 21	NORMA	Della Sala et al., 1996
VALUTAZIONE DELLE ABILITÀ VISUO-SPAZIALI							
Battery for Visuo-spatial Abilities (TERADIC) (Prove visuo-spaziali percettive)	GIUDIZIO DI LUNGHEZZA DI LINEE	8/20	7,79	0	≤ 11.75	**DEFICITARIO**	Trojano et al., 2015
	GIUDIZIO DI ORIENTAMENTO DI LINEE	4/10	2,85	0	≤ 3.06	**DEFICITARIO**	Trojano et al., 2015
	GIUDIZIO DI AMPIEZZA DI ANGOLI	2/10	1,54	1	≤ 1.32	LIMITI DI NORMA	Trojano et al., 2015
	GIUDIZIO DI POSIZIONE DI PUNTI	0/12	0	0	≤ 6.08	**DEFICITARIO**	Trojano et al., 2015
Battery for Visuo-spatial Abilities (TERADIC) (Prove visuo-spaziali rappresentazionali)	ROTAZIONE MENTALE	1/10	-0,82	0	≤ 1.70	**DEFICITARIO**	Trojano et al., 2015
	IDENTIFICAZIONE DI FIGURE COMPLESSE	0/10	0	0	≤ 5.65	**DEFICITARIO**	Trojano et al., 2015
	IDENTIFICAZIONE DI FIGURE NASCOSTE	0/10	0	0	≤ 2.33	**DEFICITARIO**	Trojano et al., 2015
	COSTRUZIONE MENTALE	0/20	0	0	≤ 7.48	**DEFICITARIO**	Trojano et al., 2015
Apples Cancellation Test	ACCURATEZZA (n° TARGET)	46/50	na	na	<45	NORMA	Mancuso et al., 2015
	NEGLECT PER LO SPAZIO	0	na	na	$\frac{< -2}{> +2}$	NORMA	Mancuso et al., 2015
	NEGLECT PER L'OGGETTO	5	na	na	$\frac{< -1}{> +1}$	**DEFICITARIO**	Mancuso et al., 2015
	TEMPO DI ESECUZIONE	231	na	na	>182	**DEFICITARIO**	Mancuso et al., 2015
Barrage di Linee (BIT)	ACCURATEZZA	25/36	na	na	≤ 34	**DEFICITARIO**	Spinazzola et al., 2010
	RAPPORTO OMISSIONI SX/TOT	0.64	na	na	≥ 0,75	NORMA	Spinazzola et al., 2010
VALUTAZIONE DEI DISTURBI DELLA RAPPRESENTAZIONE DEL CORPO							
Test per l'Agnosia Digitale		4/24	na	0	< 14	**DEFICITARIO**	Spinnler e Tognoni, 1987
Test per il disorientamento dx/sx	ORIENTAMENTO VERSO IL PROPRIO CORPO	12/12	na	na	na	NORMA	Benton, 1983
	ORIENTAMENTO VERSO L'ESAMINATORE	8/8	na	na	na	NORMA	Benton, 1983
	PUNTEGGIO TOTALE	20/20	4	4	< 17	NORMA	Benton, 1983
Valutazione dell'Emisomatoagnosia		18/18	na	na	< 18	NORMA	Bisiach et al., 1985
VALUTAZIONE PRASSIE							
Pantomima dell'uso di oggetti		20/20	na	na	< 18	NORMA	De Renzi et al. 1980
Test dell'Aprassia ideo-motoria		19/20	na	3	na	NORMA	Spinnler e Tognoni, 1987
Test dell'Aprassia bucco-facciale		20/20	na	4	na	NORMA	Spinnler e Tognoni, 1987
VALUTAZIONE DISTURBI VISUO-COSTRUTTIVI							
Test dell'Aprassia costruttiva		1/14	0,5	0	<8	**DEFICITARIO**	Spinnler e Tognoni, 1987
Disegno con i cubi (WAIS-IV)		2	na	na	na	**DEFICITARIO**	WAIS-IV





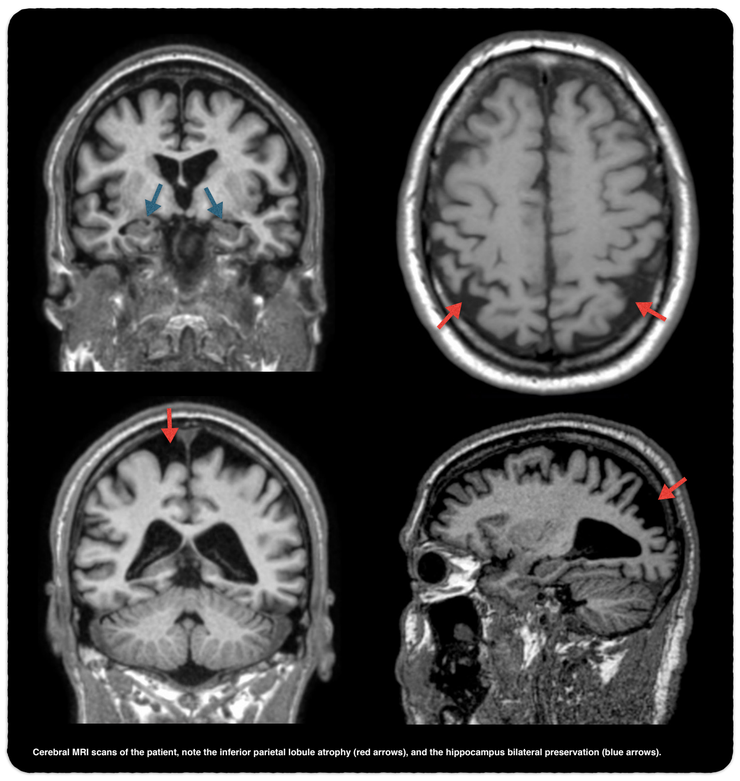


Diagnostic criteria for Posterior Cortical Atrophy (Tang-Wai et al., 2004)

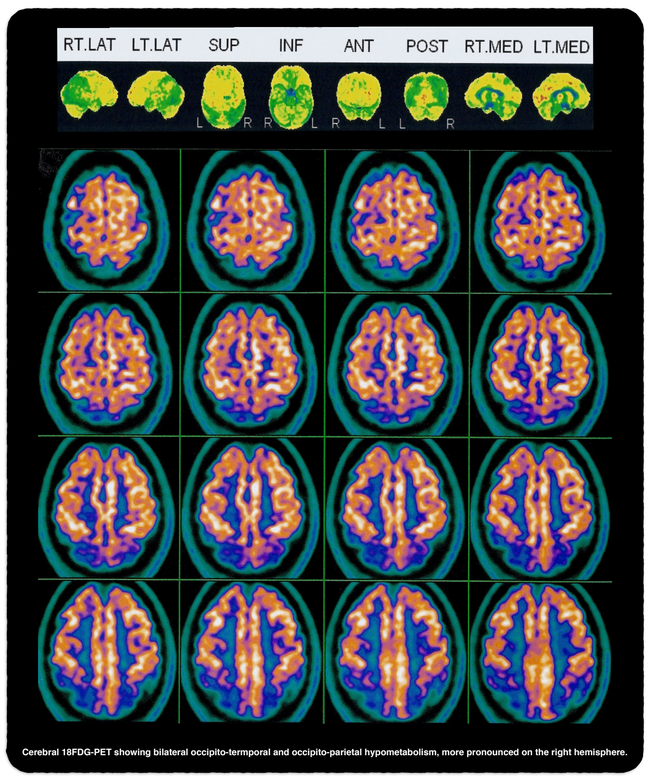
Core features
- Insidious onset and gradual progression
- Presentation of visual complaints in the absence of significant primary ocular disease explaining the symptoms
- Relative preservation of anterograde memory and insight early in the disorder
- Disabling visual impairment throughout the disorder
- Absence of stroke or tumor
- Absence of early parkinsonism and hallucinations
- Any of the following findings:
  - Simultanagnosia with or without optic ataxia or ocular apraxia
  - Constructional dyspraxia
  - Visual field defect
  - Environmental disorientation
  - Any of the elements of Gerstmann syndrome

Supportive features
- Alexia
- Presenile onset
- Ideomotor or dressing apraxia
- Prosopagnosia

Investigations
- Neuropsychological deficits referable to parietal and/or occipital regions
- Focal or asymmetric atrophy in parietal and/or occipital regions on structural imaging
- Focal or asymmetric hypoperfusion/hypometabolism in parietal and/or occipital regions on functional imaging



Cerebral MRI scans of the patient, note the inferior parietal lobule atrophy (red arrows), and the hippocampus bilateral preservation (blue arrows).



Cerebral 18FDG-PET showing bilateral occipito-termporal and occipito-parietal hypometabolism, more pronounced on the right hemisphere.

Conclusion: Among the constellation of visuo-spatial disorders, that are prominent in PCA, object-centered (allocentric) neglect may be the first and unique manifestation of hemineglect.
This highlights the importance of both egocentric and allocentric neglect testing in these patients.
This report underline the importance of an extensive assessment of selective visuo-spatial dysfunctions in clinical practice.

References:
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