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Unilateral left prosopometamorphopsia: A neuropsychological case study

Luigi Trojano[a,b,*], Massimiliano Conson[a], Sara Salzano[a], Valentino Manzo[c], Dario Grossi[a]

[a] Neuropsychology Laboratory, Dept. of Psychology, Second University of Naples, Via Vivaldi 43, 81100 Caserta, Italy
[b] Salvatore Maugeri Foundation, Scientific Institute of Telese Terme, IRCCS, Italy
[c] Dept. of Neurology, A.O.R.N. Cardarelli, Via Cardarelli 45, Naples, Italy

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ABSTRACT

We describe a patient who suddenly developed prosopometamorphopsia after a childbirth; she claimed that the left half of well-known and unfamiliar faces looked distorted. Brain MR was normal, whereas SPECT showed hypoperfusion of the left infero-lateral occipital cortex. No visual recognition defects for objects or faces were present. In three matching tasks with half-faces (Experiment 1), chimeric faces (Experiment 2), or chimeric objects (Experiment 3), the patient was impaired only when she matched pairs of chimeric faces differing in their left half; the same results were obtained after 1 year. This is the first behavioural demonstration of selective chronic metamorphopsia for the left side of faces, and provides new insights for models of face processing.

Metamorphopsia encompasses a wide spectrum of visual perceptual distortions (Critchley, 1953), such as alteration of perceived object size (micropsia and macropsia) or, rarely, altered perception of faces (prosopometamorphopsia; ffytche & Howard, 1999). Prosopometamorphopsia (PMO) can involve the whole face, as in the classical description by Bodamer (1947), but also only one side of face, usually after a right hemisphere damage (Brust & Behrens, 1977; Ebata, Ogawa, Tanaka, Mizuno, & Yoshida, 1991; Young, de Haan, Newcombe, & Hay, 1990; for a review see, Miwa & Kondo, 2007). For instance, Brust and Behrens (1977) observed a patient with a right posterior temporal damage who had episodes of visual illusions during which the right half of faces seemed to melt, "like clocks in a Dalì painting". An example of stable unilateral face distortion (the right side of a face appearing smaller than the left) has been reported by Ebata et al. (1991) in a patient with a small haematoma in the right retrosplenial region. In such cases, visual distortion did not involve perception of objects other than faces, and tasks assessing apperceptive or associative face processing did not show relevant impairments. Unilateral PMO after a left hemisphere lesion has been described only in four patients who however also showed metamorphopsia for objects or body parts (Imai, Nohira, Miyata, Okabe, & Hamaguchi, 1995; patient M.Z., Nijboer, Ruis, van der Worp, & De Haan, 2008; Satoh, Suzuki, Miyamura, Katoh, & Kuzuhara, 1997; Shiga, Makino, Ueda, & Nakajima, 1996). For instance, patient M.Z. described by Nijboer et al. (2008) had a left temporo-occipital lesion and unilateral contralesional metamorphopsia; however, in an object confrontation task, she reported that objects presented on her right side appeared distorted as well.

From the above overview, it should appear that only right hemisphere lesions can determine selective unilateral PMO, consistent with the idea of a right specialization for face processing. However, the analysis of selective PMO has always been based upon patients' subjective complaints, thus preventing any attempt to comprehend its mechanisms. Here we describe a neuropsychological study on a patient with selective unilateral PMO. Our aim was to explore the cognitive basis of this rare disorder and its implications for cognitive models of face processing.

1. Case report

D.G., a 24-year-old right-handed (no familiar history of left-handedness; score on Edinburgh Handedness Inventory = 90; Oldfield, 1971) housewife with 8 years of formal education, suddenly developed severe migraine, confusional state, and blurred vision, mainly in her right visual hemifield, after child delivery. Consciousness returned normal and migraine subsided in a few hours, but the visual disturbances persisted longer.

Repeated EEG recordings showed theta-wave slowing over posterior regions of the left hemisphere. Pattern reversal and flash VEPs were within normal limits 6 months after the stoke. Repeated MR did not disclose pathological areas. Brain SPECT (performed 30 min after the injection of 740 MBq of Tc-99m HMPAO) showed reduced blood flow in the inferior and lateral cortex of the left occipital lobe (Fig. 1).

* Corresponding author at: Dept. of Psychology, Second University of Naples, Via Vivaldi 43, 81100 Caserta, Italy. Tel.: +39 0823 274774; fax: +39 0823 274774.
E-mail address: luigi.trojano@unina2.it (L. Trojano).

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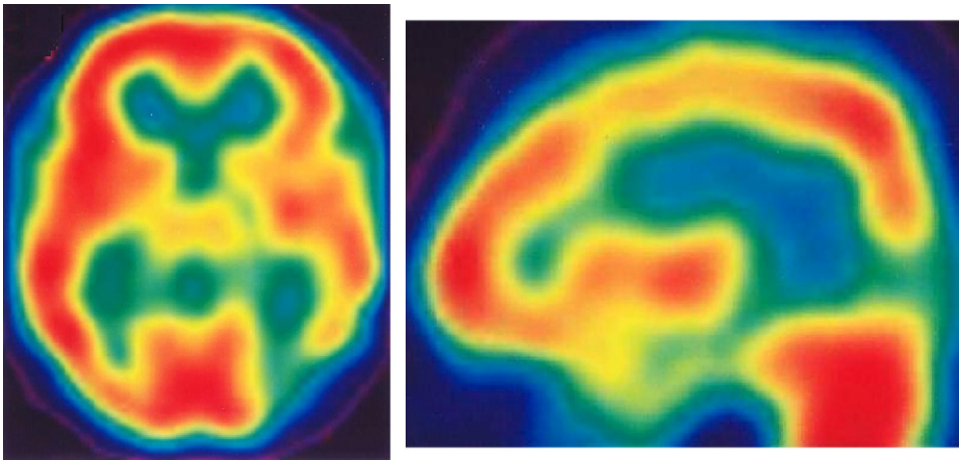

Fig. 1. Axial (on the left) and left paramedian sagittal (on the right) brain SPECT scans showing reduced blood flow in the inferior and lateral cortex of the left occipital lobe (regional blood flow reduced by 25.8% with respect to contralateral cortex).

The patient came to our observation 6 months after the stroke. At that time she was alert and cooperative, had recovered most of her visuo-perceptual abilities and was autonomous in her daily activities. The patient did not show visual field defects on Goldmann perimetry, but she complained that the left half of people's faces (the part on her right side) appeared "out of shape". D.G. claimed that "the left eye looks elongated towards left ear, the nose appears to be bended towards left cheek and the mouth towards the chin" (Fig. 2), irrespectively of whether she looked at familiar or unknown people, or even at herself in a mirror. Nonetheless, she reported to be able to recognize relatives and famous people by face, and also to visualise familiar faces in her mind without distortions.

A general neuropsychological assessment did not reveal intellectual or language disturbances; the patient could correctly describe a complex scene, draw figures, read and write (words and non-words). A specific battery for visuospatial abilities (BVA, known in Italy as TERADIC; Angelini & Grossi, 1993) did not disclose impairments in visuospatial perceptual skills; in particular, patient's performances on tasks tapping discrimination of line length, line orientation and angle size were within the normal range.

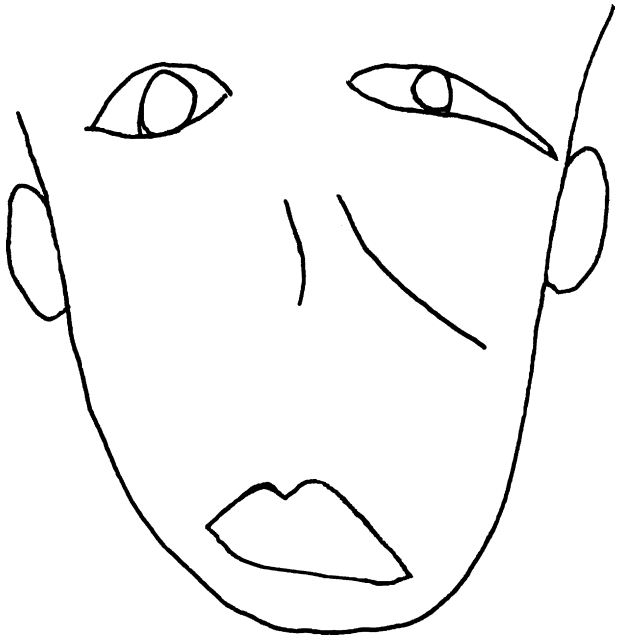

Fig. 2. Patient's drawing of a face to depict her own subjective complaint.

Assessment of face processing was performed by means of the Benton Face Recognition Task (Benton, De Hamsher, Varney, & Spreen, 1992) and by a face recognition task requiring the patient to identify black and white images of 56 famous faces. Five normal right-handed females, matched to D.G. for age (range 22–26) and education, were tested for obtaining reference values for recognition of famous faces; the patient's performance was compared with that of the normal subjects according to Crawford and Garthwaite's (2002) statistical method. Moreover, we assessed visual object recognition by a naming task (Laiacona, Barbarotto, Trivelli, & Capitani, 1993) and the Poppelreuter-Ghent's Overlapping Figures Test (Della Sala, Laiacona, Trivelli, & Spinnler, 1995).

D.G. achieved a normal score on the Benton Face Recognition Task (44/54; cut-off = 38) and did not differ from normal controls in recognizing famous faces (D.G. = 46, controls = 48, S.D. 2.6; $t = -1.053$, two-tailed $p = 0.352$). The patient performed within normal range on the object naming task (raw score = 78/80, cut-off score = 61) and on the Poppelreuter-Ghent's Overlapping Figures Test (raw score = 71; normal range = 51.5–71). These data demonstrated that D.G. did not show overt prosopoagnosic defects or object recognition impairments.

2. Special neuropsychological assessment

A special neuropsychological assessment was performed to investigate the patient's abilities to process information from the two sides of faces and objects. To this scope, D.G. underwent three experiments, requiring to match pairs of half-faces (Experiment 1), of chimeric faces (Experiment 2), and of chimeric objects (Experiment 3).

Five normal right-handed females, matched to D.G. for age (range 21–29) and education, were tested to obtain reference values for the experimental tests. Also in this case we compared the patient's performance with that of the normal subjects according to Crawford and Garthwaite's (2002) statistical method.

2.1. Experiment 1: matching of half-faces

The first experiment was aimed at verifying whether the patient's subjective defect in perceiving the left half of faces could be evidenced in a task where she had to identify left (or right) half-faces. To this purpose, we devised a half-face matching task, in which the patient was required to match a target face with half-faces (right or left) in free-viewing condition without time constraints. This procedure allowed to assess the patient independently