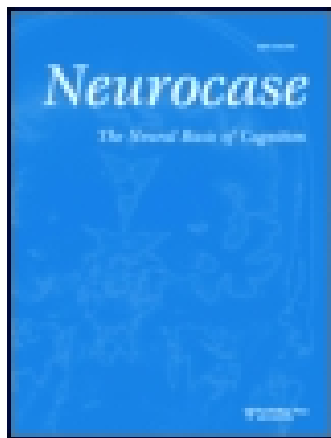


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Visual disorders, the prosopometamorphopsia and prosopagnosia type in the early days after the onset of brain hemorrhagic stroke – a case report

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Presented case report illustrates symptoms of prosopometamorphopsia (PM) and prosopagnosia, observed in the early days after the onset of a hemorrhagic stroke resulting from a complication of endovascular treatment of intracranial aneurysms and the use of anticoagulation therapy. PM is a visual disorder in which faces are perceived as distorted. The female patient described in the present study reported that faces she looked at seemed younger or older than in reality or as if they were dirty, swollen, or with a grimace. She also experienced symptoms of prosopagnosia, which is difficulty of recognizing familiar faces of people (e.g., of her husband and daughter). In the interview 6 months after the first examination, the patient reported spontaneous withdrawal of the visual disturbances.

Keywords: prosopometamorphopsia; metamorphopsia; prosopagnosia; aneurysm; stroke

Face recognition is one of the core capabilities of social cognition, which occurs not only in humans but also in animals (Martinez & Matsuzawa, 2009; Neiworth, Hassett, & Sylvester, 2007). Its extraordinary significance demonstrates the fact that the circuits crucial for face recognition are distinguished among the neuronal structures responsible for the receipt and processing of visual stimuli in general (generally not specialized in the reception of specific stimuli). This was demonstrated in numerous studies carried out with functional magnetic resonance imaging (fMRI) (Gobbini & Haxby, 2007), positron emission tomography (PET) (Ganel, Valyear, Goshen-Gottstein, & Goodale, 2005; Gorno-Tempini et al., 1998; Nakamura et al., 2000), and other techniques. The important areas associated with this function are the posterior-lateral part of the fusiform gyrus (fusiform face area, FFA) (Kanwisher, McDermott, & Chun, 1997) and the adjoining area situated more dorsolaterally, closer to the pole of the occipital lobe (occipital face area, OFA) (Pitcher, Walsh, & Duchaine, 2011; Pitcher, Walsh, Yovel, & Duchaine, 2007; Puce, Allison, Asgari, Gore, & McCarthy, 1996). Naturally, they do not operate in isolation from the other structures of the visual perception system, but their relatively greater activation appears to be a necessary condition for the proper conduct of the complex process of face recognition. It was also found that FFA activation occurs in both the visual stimulus presentation (face images) and the stimulated imagination of the same images, e.g., while giving the name of a known person (Uchida et al., 2000).

Prosopometamorphopsia (PM) was described for the first time in 1947 by Bodamer as a disorder in which the

patient sees faces that look different than in reality, for instance, swollen, grotesque (Heo et al., 2004), extended, flattened (Nijboer, Ruis, van der Worp, & De Haan, 2008), with bulging eyes, or as if they were older, uglier (Bodamer, 1947; Nijboer et al., 2008), with a hostile expression (Ffytche & Howard, 1999), or conversely, as if they were nicer, more friendly (Seron et al., 1995). Patients with PM, as long as their disorder does not co-occur with typical signs of face agnosia, do not reveal any difficulties in the identification of familiar faces (Bodamer, 1947; Nijboer et al., 2008). Relevant case studies described in the literature confirm the distinctiveness of these two disorders (Heo et al., 2004). PM can affect the perception of the entire face (Bodamer, 1947; Heo et al., 2004; Seron & et al., 1995) or its components, mainly the middle (Cho et al., 2011; Miwa & Kondo, 2007; Trojano, Conson, Salzano, Manzo, & Grossi, 2009). PM may be caused by structural brain damage (Nijboer et al., 2008; Seron et al., 1995), epilepsy (Heo et al., 2004), migraine (Trojano et al., 2009), or eye diseases such as macular degeneration, glaucoma, or cataracts (Ffytche & Howard, 1999). Damage to the brain that causes problems described previously most commonly includes the right (or bilateral) occipital cortex, often with additional deterioration of areas of the temporal and parietal lobe, areas at the rear of the splenium, and the posterior part of the dentate gyrus (Nijboer et al., 2008). PM usually spontaneously retreats lasting from several hours to several weeks after the stroke incident (Heo et al., 2004; Nijboer et al., 2008). Another disorder leading to difficulties in the correct reception of faces is face agnosia (prosopagnosia) – a rare disorder

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involving difficulty or total inability to recognize well-known faces (Barton, 2008; Darby & Walsh, 2005), which may or may not be accompanied by other neuropsychological deficits (Sorger, Goebel, Schiltz, & Rossion, 2007). Prosopagnosia usually results from the damage to the FFA and OFA areas (Dalrymple et al., 2011; Haxby, Hoffman, & Gobbini, 2000). It may also be of developmental or congenital origin (Dinkelacker et al., 2011; Grueter et al., 2007). According to the literature, there are various types of difficulties in proper face perception, ranging from the difficulties in distinguishing familiar and unfamiliar faces or matching a face to a specific person to a total inability to distinguish faces from other stimuli (Busigny, Graf, Mayer, & Rossion, 2010; Knutson, DeTucci, & Grafman, 2011; Sacks, 1998) and, what is important, some patients may not be aware of experienced deficits (Young, Haan, & Newcombe, 1990). The present study describes a female patient exhibiting peculiar abnormalities in facial perception consequent to intracerebral hematoma, which was the effect of complication after embolization of intracranial aneurysm surgery and anticoagulation treatment. In the early days after the onset of the hemorrhagic stroke, the patient has reported difficulties in recognizing faces of close, well-known people. Considering the symptoms and location of the brain damage, she was initially diagnosed with face agnosia. In the later stage of diagnostics, the symptoms of PM were revealed and, with time, definitely came into the fore.

Case report

Medical history

Female patient, a bank manager, aged 34, holding a university degree in economics, was admitted to the neurosurgery department for endovascular treatment of non-ruptured aneurysm of the right internal carotid artery (ICA). Aneurysm, which did not give any symptoms, was accidentally discovered in the computed tomography angiography (CTA). The test was performed prophylactically because of the patient's potential genetic load, such as autosomal dominant polycystic kidney disease (ADPKD) associated with hypertension and cardiovascular disease occurrence in the immediate family. On admission to the hospital, the patient was in a very good condition and did not report any symptoms. Moreover, no abnormalities were revealed in a neurological examination. Performed three-dimensional digital subtraction angiography (3D DSA) confirmed the presence of the aneurysm of the right ICA connecting the left rear of the dome measuring $5.1 \times 4.8 \times 4.5$ mm and neck 4.5 mm (Figure 1). The patient was qualified for endovascular treatment.

Vascular prosthesis (flow-diverter, 4.25×16 mm, pipeline, ev3 Inc., Irvine, CA, USA) was implanted into

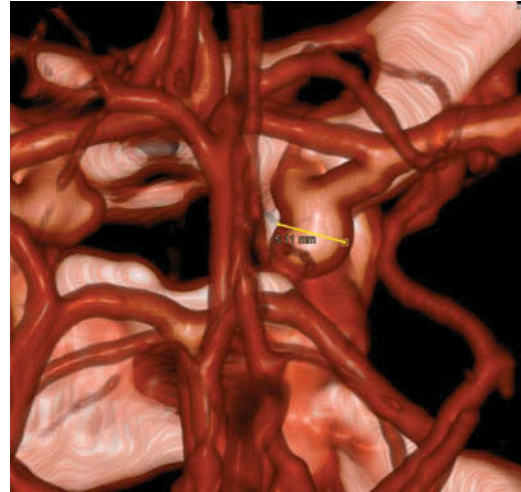


Figure 1. 3D DSA presenting the aneurysm ($5.1 \times 4.8 \times 4.5$ mm) of right internal carotid artery before embolization. [To view this figure in color, please see the online version of this Journal.]

the ICA – distal part was located between the departure of anterior choroidal artery (AChA) and the posterior communicating artery (PCoA), the proximal part in the initial section of segment C4. 3D angiographic control performed 15 min after prosthesis implantation showed partial expansion of the distal end, so that the stent was given 4×10 mm HyperGlide (ev3 Inc.) balloon to achieve full prosthesis expansion. Control angiography showed normal flow in the stent and the stagnation of flow in the aneurysm sac. There were no complications during and immediately after the test. It was recommended to give aspirin (ASA) and clopidogrel. The next day after the surgery, the patient developed severe headaches and blurred vision in the form of left-sided hemianopia, accompanied, as declared by the patient, by the general deterioration of vision. Computed tomography showed the presence of intracerebral hematoma measuring 55×36 mm, the zone of edema in the right temporo-occipital area (Figure 2). ASA was discontinued and the patient was treated conservatively with Mannitol to reduce swelling. Neurological examination did not reveal any other neurological deficits.

In the following days, the patient's condition continued to improve with gradually decreased headache and the retreat of hemianopia. A control CT (10 days post-hemorrhage) showed reduction in the hematoma (28×25 mm) and the zone of edema (Figure 3). One week after the hemorrhage, a complete withdrawal of hemianopia was noted which was confirmed by perimetry method.

Neuropsychological assessment

One day after hemorrhage, patient remained in basic mental and physical contact without clearly perceptible behavioral deficits. Due to very severe headaches,

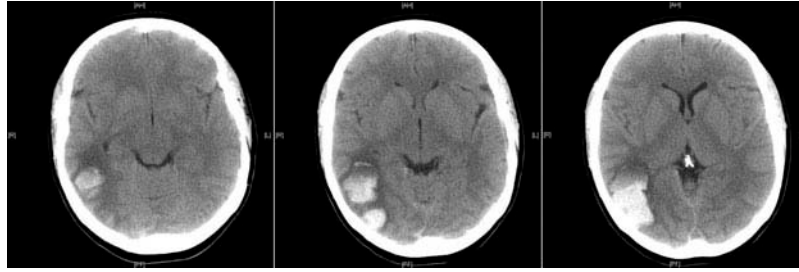


Figure 2. CT taken right after the hemorrhage onset (55 × 36 mm). Left side of the image = right side of the brain.



Figure 3. Control CT taken 10 days post-hemorrhage, showing significantly reduced hematoma (28 × 25 mm).

neuropsychological examination was limited to screening tests only. In the following days, the general condition of the patient was improving. During a routine psychological interview on the 4th day after onset, she began to report difficulties in recognizing people. She admitted that she had noticed them before, but she had thought that they were due to exhaustion and a severe headache and that they might be related to vision problems in the left visual field (symptoms of hemianopsia). In a detailed interview, it was found that quite soon after the surgery, she had realized that she could hardly recognize the faces of nurses and doctors who were taking care of her. Therefore, she identified the hospital staff only by their voice, body posture, or the manner of movement. She claimed that their faces seemed to be similar to each other, but dissimilar to the images she was storing in her memory. This was true for both medical personnel and close family members. The patient had difficulty in recognizing her own daughter and her husband being present among others in the corridor. She was able to recognize most people only by their clothes, body shapes, hairstyles, and, most importantly, voices.

In the next 3 days, the abnormalities in the identification of faces were gradually decreasing. However, she started to see faces in distorted form. They appeared to be swollen, sometimes elongated, disproportionate, or dirty.

Test assessment

Neuropsychological evaluation started 3 days after the patient reported difficulties in recognizing familiar people

(7 days post-hemorrhage). During the subsequent diagnostic meetings and follow-up stay in hospital, the patient remained in a very good logical and physical contact. She was fully oriented mentally and aware of the experienced symptoms.

She gave informed consent for neuropsychological testing, recording the examination and interview, and using these data for research purposes.

Visual perception assessment

To verify the hypothesis of the presence of prosopagnosia, we started the assessment with a selection of tests and clinical trials evaluating the basic level of visual perception and visual-spatial functions (see Table 1).

The scores of the psychometric tests and non-standardized clinical trials ruled out the presence of visual perceptual disorders concerning stimuli other than faces.

Assessment of mnemonic functions

In addition, we assessed mnemonic functions, such as learning new information (verbal and nonverbal material presented in the visual and auditory modalities) as well as long-term memory, to control the impact of these functions on the reported disorders. The results of the tests are summarized in Table 2.

Results of memory tests suggest that basic mnemonic functions probably did not have influence on symptoms reported by the patient. Based on a detailed interview and

Table 1. Psychometrics tests and non-standardized clinical tasks of visual perception.

Psychometrics tests	Raw scores	Number of tasks	
CORVIST	1. Symbol Acuity Test	36	
	2. Shape Discrimination Test	8	
	3. Size Discrimination Test	2	
	4. Shape Detection Test	8	
	5. Hue Discrimination Test	4	
	6. Scattered Dot Counting Test	4	
	7. Fragmented Numbers Test	8	
	8. Word Reading Test	16	
	9. Face Perception Test	8	
	10. Crowding Test	4	
VOSP	Shape Detection Screening Test	Percentiles 100	
	1. Incomplete Letters	19	51.9
	2. Silhouettes	23	50
	3. Object Decision	18	39.5
	4. Progressive Silhouettes	11	42.2
	5. Dot Counting	10	100
	6. Position Discrimination	19	22.1
	7. Number Location	10	100
8. Cube Analysis	10	100	
Rey Complex Figure – Copy	36	>16	
Block Design (WAIS-R)	44	98	
Boston Naming Test	15/15	–	
Non-standardized clinical tasks ^a		Number of tasks	
Overlapping shapes	3	3	
Masked shapes	3	3	

Note: CORVIST – Cortical Vision Screening Test; VOSP – Visual Object and Space Perception Battery; WAIS-R – Wechsler Adult Intelligence Scale – Revised; ^aSet of clinical tasks to assess cognitive functions (Lucki, 1995).

Table 2. Memory standardized tests.

Memory tests	Raw scores	Norms
RAVLT	Trials 1–5	Mean (SD) 53.6 (8,3) ^a
	Recall after 20 min	11.1 (2,8) ^a
BVRT (Ver. C; Met. D)	Number of correct responses	8.04 (1.19) ^a
	Number of errors	2.52 (1.70) ^a
RCF – recall after 3 min	13.5	Percentile rank 1 ^b
Memory tests for faces		Mean (SD)
CMTF	60	58.9 (8.3) ^c
RBMT-3 – Face Recognition subtest	13	Percentile rank 37 ^d

Note: RAVLT – Rey Auditory Verbal Learning Test; BVRT – Benton Visual Retention Test; RCF – Rey Complex Figure; CMTF – Cambridge Memory Test for Faces; RBMT – Rivermead Behavioural Memory Test-3.

Sources: ^aStrauss, Sherman, and Spreen (2006); ^bMeyers and Meyers (1995); ^cDuchaine and Nakayama (2006); ^dWilson et al. (2010).

Information subtest from Wechsler Adult Intelligence Scale-Revised (WAIS-R), we excluded long-term memory disorders. In tests that assess memorizing new information (both verbal and non-verbal including faces), the patient displayed difficulties in memorizing only in Rey Complex Figure Test in 3-minute delay recall (<1 percentile). This score may indicate that the patient experienced difficulties in memorizing complex abstract visual material.

Facial gnosis assessment

Facial gnosis was assessed with the use of a carefully selected set of images depicting close people (husband, daughter, friends) and a set of 74 photographs of famous people divided into four groups: politicians, actors, musicians, and athletes. The photographs showed faces only, did not include any extra-facial cues (e.g., hair, glasses) and were neutral in emotional tone (no expression of

Table 3. Face recognition – non-standardized clinical trials.

Famous people	Subject Raw scores	Controls (<i>n</i> = 4) Raw scores			
		I.	II.	III.	IV.
Whole faces <i>n</i> = 74	55 (74%)	60 (81%)	58 (78%)	63 (85%)	67 (90%)

emotions). The criterion for choosing the people was researchers' subjective discretion. The high-definition color photographs were presented on a 17-inch display with no time limit; however, the face recognition time was assessed. The patient was asked about the name of each person; if she experienced difficulties in giving it, she was asked to give some characteristics, which would lead to correct identification (e.g., an office held). After the examination, the familiarity of the presented people was verified: last names were given and the patient was asked to say whether she knew how the people presented in the examination looked. The patient's scores were compared with control group.

Table 3 summarizes recognition tasks scores in relation to control group.

The clinical trials did not clearly confirm the presence of prosopagnosia. The patient recognized close people correctly and the faces of public people only slightly worse than control group did. Interestingly, she provided numerous comments on facial distortions that she saw. She said that faces looked more or less favorable, clean or dirty, younger or older, or particular face elements appeared changed. In her own words, the observed people looked "a bit similar to each other, like someone we spotted on the street who resembles someone we know." Looking at the pictures of her child she said:

I can see her eyes and smile quite different than usual, everything seems to be puffy, swollen, her smile is different than the one I remember, as if she was unnaturally baring his teeth, as if the smile would like to tell something, a grimace.

In the photo of her daughter's class "some children are dissimilar, the girl smiles and looks like a monkey." Looking at her husband's picture she said: "he looks strange like one of the Jetsons from the cartoon or as if he was a midget." The faces of people seen at the hospital "look fresh or non-fresh, the fresh person seems to be so radiant, his clean face is smooth, rejuvenated, and suddenly it turns gray and lacks expression." "If someone has a little round face it seems to me that it is puffy and soon it will burst, as if this face was swelling up."

The patient pointed to a couple of factors, which significantly influenced the perception of face. The first factor she mentioned was the degree to which a face was familiar. More familiar faces were perceived as more distorted: "The people I know better are distorted more, it's as if in a

movie that I know well completely different actors were appearing." Large distortions occurred also when she looked at her own face in a photograph: "In this photograph I don't look myself, my face is sort of smaller, rounder, the part above the nose has kind of contracted, all is smaller, the part up to the hair is flattened and stretched at the eye level"

The second factor, which modified the occurrence of distortions, was emotions on the observed face. As she reported:

the woman who was with me in the same room ... yesterday I watched her showing different emotions – she was smiling, she was happy, experiencing something, and suddenly she became sad and depressed, and at that very moment her face looked as if she was a completely different person – a stranger who had entered the room.

Facial movements also appear to play a part in face perception:

When someone, like my old friend stands in front of me and says something, I see him at this moment a little differently, the way he is moving or he says anything. The longer I look at him, the more I see that the elements of his face are different, suddenly the picture is a little different, although not very much, but some elements are different, which makes the whole picture look different.

The patient reported that light and shadows played a part in face perception as well. She also indicated that the time for which she was looking at a picture was an important factor affecting the perception – when she was observing faces (in real life and in the pictures) they were distorting:

The longer I look at the face, the more it changes (...) sometimes in the beginning I know who it is, but the longer I look, the more elements merge with each other so much that a very different picture arises, the more I look at it, the more it is changing.

Throughout the hospitalization, the symptoms were gradually changing and retreating: "every day is better, at the beginning I did not recognize faces." The patient reported that she periodically could see faces properly (in the morning), but the difficulties tended to recur and worsen in the afternoon. Consequently, the faces that she once perceived correctly were becoming distorted again. The symptoms were improving every day of

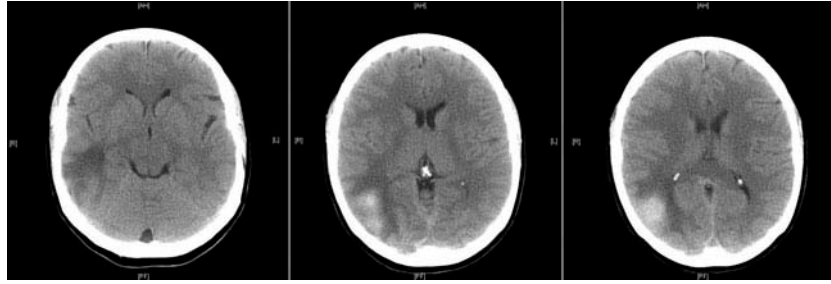


Figure 4. Control CT of intracerebral hematoma just before patient's discharge from the hospital.

hospitalization, but did not retreat completely. After discharge from hospital, 14 days since bleeding started, the patient reported small, periodically emerging difficulties in identifying some of the faces. Most of them were still perceived in a distorted form. The CT scan showed almost complete absorption of the hematoma (Figure 4).

Six months after the diagnosis, a detailed interview was conducted with the patient about the visual disturbances, which had occurred after discharge from the hospital. The patient was describing that she had had difficulties in recognizing familiar people for 2 consecutive weeks. After 1 month, the abnormal facial perception had gradually retreated and eventually disappeared.

Discussion

We are describing a rare case of a patient with prosopagnosia and PM symptoms consequent to intracerebral hematoma in the right temporo-occipital area as a complication after the embolization of aneurysm of the right ICA. Few cases of patients with diagnosed PM have been described to date (Cho et al., 2011; Ebata, Ogawa, Tanaka, Mizuno, & Yoshida, 1991; Hwang et al., 2012; Trojano et al., 2009) and we found only one case in which both syndromes in one patient were described (Seron et al., 1995). Similar to our description, in the beginning, researchers observed in a patient with prosopagnosia symptoms that retreated dynamically and the patient started to complain about PM symptoms.

There are various descriptions of PM symptoms in the literature. Firstly, perception distortions can relate to the entire face (Hecaen & Angelergues, 1962; Heo et al., 2004; Heutink, Brouwer, Kums, Young, & Bouma, 2012; Seron et al., 1995), which we observed in our patient as well. They can be limited to some part of the face including one half of the face, most often the left half, which is related to the lateralization of damage (Cho et al., 2011; Miwa & Kondo, 2007; Nijboer et al., 2008; Trojano et al., 2009), or concern particular parts of the face such as the nose and mouth (Hwang et al., 2012).

Secondly, qualitative changes in face perception (such as change in face proportions) are pointed to, e.g., the lengthened and narrowed nose (Hwang et al., 2012).

Further, what draws attention in Seron et al.'s study (1995) and in our patient is a subjective evaluation of the seen face, the evaluation being less favorable (older, dirty, gray, swollen) or more favorable (younger, smooth skin, clean, radiant). In turn, Heuting et al.'s (2012) patient reported that her grandchildren looked "as if they were overweight and had an extremely tanned skin."

Thirdly, it is worth noting that in our patient we could observe great symptom variability related to dynamic healing processes. The patient reported that symptom intensity not only fluctuated during her hospitalization but also was changing dynamically while she was watching a face for a longer time ("the longer I watch, the more the face changes, some elements change and the overall face is different"). What distinguishes the present case is that the variability pertained also to the pattern of distortions itself, e.g., the patient reported that on one occasion a face looked swollen, and on another occasion it looked changed and flattened. Other authors describe stability of symptoms and this same pattern of distortions (Cho et al., 2011; Ebata et al., 1991; Hwang et al., 2012). For example, Hwang et al. (2012) described PM restricted to the nose and mouth, while the patient described by Ebata et al. (1991) saw the right half of the face as diminished.

What is interesting, our patient reported that the degree of familiarity, among other things, influences face perception (similar observations were reported by Heuting et al., 2012; Seron et al., 1995). Furthermore, other factors such as movement, light, and the emotional expression of the face influenced the symptoms in our patient as well, but they were not described by other authors.

The influence of the location of lesion on the manifold picture of the disorder is not fully clear. Two types of lesion, which can cause PM, are pointed to: lesion affecting a relatively large temporoparietal and temporo-occipital areas (in most cases) or small retrosplenium lesions (Miwa & Kondo, 2007). Majority of studies describe patients with a right fusiform gyrus lesion (Cho et al., 2011; Heutink et al., 2012; Hwang et al., 2012; Miwa & Kondo, 2007; Nass, Sinha, & Solomon, 1985; Nijboer et al., 2008; Seron et al., 1995), similar to the case that we have described. In the literature, PM as a result of a left hemisphere lesion located in this area has also been

described (Cho et al., 2011; Mendez, 1992 as cited in Miwa & Kondo, 2007; Trojano et al., 2009). Many studies, mainly concerning prosopagnosia, confirm the importance of temporo-occipital area (especially fusiform gyrus) in processing and storage of specific stimuli such as faces. Damage to this structure can therefore cause disturbances in the perception of visual information, specifically limited to faces, which can result in prosopagnosia or PM. To date, there is a lack of studies, which elucidate the influence of brain lesion location on the diversity of PM symptoms. Further research is needed to more clearly understand the nature of this disorder.

What seems to be interesting is the occurrence of two symptoms related to face perception in our patient: prosopagnosia and PM. A similar case has been described only by Seron et al. (1995). Other authors described PM symptoms with no prosopagnosia symptoms. When considering the mechanism underpinning the two syndromes, we take into account two hypotheses. The first one, they are independent symptoms, which can co-occur. The second one, they are one and the same symptom differing in intensity, which results in a different clinical picture (a continuum of symptoms).

When analysing the case of our patient, according to the first hypothesis, we can assume that prosopagnosia symptoms, which dominated in an early stage, then retreated, which enabled an observation of PM, or this disorder occurred as a new symptom. The second hypothesis, proposed in Seron et al.'s paper (1995), would claim that intensive PM symptoms take the form of apperceptive prosopagnosia and, in diminishing gradually, result only in perceived face distortions while enabling face identification.

In the case that we have described, the second hypothesis seems to be more apt. It is supported by the fact that at a stage when prosopagnosia was not diagnosed, some difficulties in face recognition, related to distortions, occurred periodically. The next argument is linked to a change in perceiving face while looking at it. The patient reported that she recognized the person in photograph, but the longer she looked at him/her, the less the person resembled himself/herself, which was caused by arising distortions.

Because of there being few publications on the subject, especially few studies of cases combining prosopagnosia and PM, proposed mechanisms explaining the co-occurrence of these symptoms and their relationship remain only hypotheses, which emphasizes the need for further research in this domain.

As far as the treatment of patients suffering from both PM and prosopagnosia is concerned, it is absolutely crucial to gain understanding of their feelings and inform them about the nature of their subjective experiences. As noted earlier, the described patient was deeply concerned about the symptoms she was displaying. In particular, she was apprehensive that the symptoms could negatively affect her further professional and social life. She was also afraid of

being diagnosed with a mental illness. Thanks to the clarification of the specificity of the experienced problems and information about the possibility of spontaneous improvement, the level of stress experienced by the patient was significantly reduced. Psychoeducation and emotional support are – so far – the only form of psychotherapy available for patients with deficits of this type.

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