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## Prosopagnosia without topographagnosia and object agnosia associated with a lesion confined to the right occipitotemporal region

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**Abstract** A patient is presented who developed prosopagnosia with a recent occipitotemporal infarct in the distribution of the right posterior cerebral artery. He did not have topographical agnosia or object agnosia. He regained the ability to recognize faces of familiar persons, whereas he remained unable to identify faces of persons whom he met after the disease onset. This case demonstrates that prosopagnosia may occur as a deficit of matching a perceived face

to a memory store of the face, and that the failure to recognize unfamiliar faces may be due to the inability to form memory stores of new faces. These deficits can occur in association with a lesion confined to the right occipitotemporal region.

**Key words** Prosopagnosia  
Visual memory · Right  
occipitotemporal lesion · Magnetic  
resonance imaging · Positron  
emission tomography

### Introduction

The inability to recognize familiar faces is a specific agnosia (prosopagnosia; Bodamer in 1947 [7]; Hécaen and Angelergues in 1962 [18]). At least two major questions remain unsettled concerning the pathogenetic mechanisms of prosopagnosia. First, it is uncertain whether prosopagnosia is (1) a partial symptom of a general impairment of visuospatial perception, (2) a defect of a presumed specific perceptual process for faces, or (3) a deficit of matching a correctly perceived face to a memory store of the face. This is largely because the previously reported patients almost always presented with other visuospatial disturbances such as topographical agnosia or object agnosia, in addition to both retrograde and anterograde prosopagnosia. Second, there is a discrepancy between autopsy and clinical studies which have not determined whether bilateral lesions or a right occipitotemporal lesion alone are required for prosopagnosia to occur. The pathology reports available have suggested that symmetrical lesions in the medial occipital region are generally necessary for prosopagnosia to occur [1, 3, 11, 16, 18, 19, 25, 32, 33, 45], because all the autopsy cases except

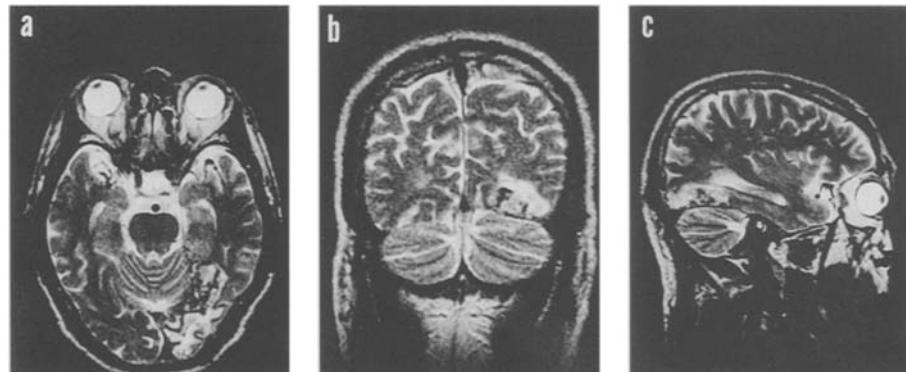
three [18, 23, 44] displayed bilateral occipitotemporal lesions [12]. In most of these reports, autopsy studies were performed long after the onset of prosopagnosia. However, there have also been reports of patients with prosopagnosia having a right occipitotemporal lesion alone. These have been on the basis of autopsy findings shortly after the development of the symptoms [23], or of clinical and radiological evidence [10, 13, 17, 21, 24, 28, 30, 40, 44].

We report the case of a patient with an acute onset of prosopagnosia who did not develop topographagnosia or object agnosia. Three months after onset, his ability to recognize faces of familiar persons recovered, but he remained unable to recognize those whom he first met after the onset. Computed tomography, magnetic resonance imaging (MRI) and positron emission tomography (PET) displayed anatomical and functional abnormalities confined to the right occipitotemporal region.

### Case report

A 62-year-old, right-handed man was admitted for evaluation of his inability to recognize faces. He was educated for 6 years in a

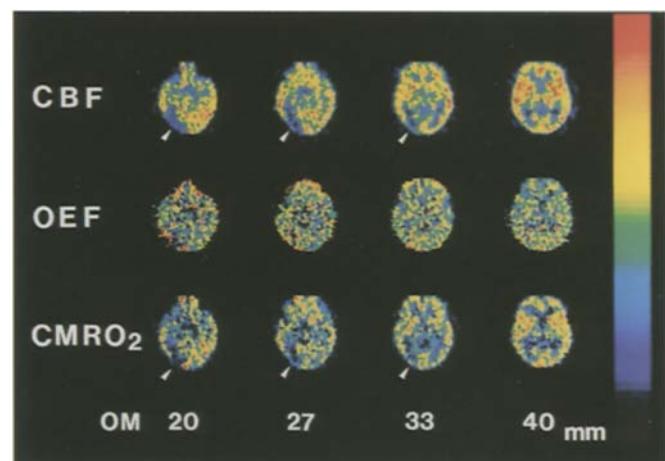
**Fig. 1** T2-weighted MRI (TR/TE 3,000/100 ms, 1.5 T, 5 mm thickness) of (a) axial, (b) coronal (3 cm anterior to the occipital pole), and (c) sagittal (1.5 cm lateral to the midline) sections, demonstrating a hyperintensity area in the cortical and subcortical areas in the distribution of the right posterior cerebral artery. In a and b, the right side of the head is on the right



primary school, worked as a local government employee until aged 48 years, and as a chief executive of a real estate business thereafter. He had been well except for hypertension and a history of acute myocardial infarction. One morning a month before admission, he was watching television when he suddenly noticed that faces on the TV screen appeared blurred and he could not recognize them. The faces appeared reddish. Subsequently, he could no longer recognize the faces of family members, relatives, and photographs of faces of famous people. However, he could find his way to familiar places. As his complaint, which caused him great concern, persisted, he visited our department.

On admission, he was alert, and well oriented for both time and space. He had a Mini-Mental-State examination score of 30 (full score). The Wechsler Adult Intelligence Score (WAIS) was 83 for the verbal tests, and 70 for the performance tests. The low score for the verbal tests may be due to his short education (6 years); in addition, the low performance test score was mainly due to his impaired ability in the picture arrangement tests. However, his score for block design tests was excellent, suggesting unaltered synthesizing and abstract ability in visuospatial functions. He had a left upper quadrantanopia in the right eye, but did not neglect the left half of his visual space. Visual acuity was 0.5 (1.5 with glasses) in the right eye, 0.6 (1.5 with glasses) in the left. His colour perception was normal: he named and matched colours correctly, and had no problem with Ishihara's test for colour blindness. However, he could no longer recognize relatives or other familiar persons by their faces, but could recognize them immediately by voice. He remembered the hospital staff only by voice, but not by face. He could identify a face as a face in real life or on photographs, and describe characteristics of faces. However, he saw faces as partly distorted (metamorphopsia), stating that the right eye of persons was twinkling, and that the right palpebra appeared "long and narrow as a fox's eye". This metamorphopsia was restricted to human faces, and was not seen for faces of animals. He could identify faces of animals (cats, dogs, horses, etc.). When he was presented with a single front-view photograph of an unfamiliar face, he could identify it in several front-view photographs from other unfamiliar faces; his responses were correct in three such tests. However, he always failed to identify it in several oblique-view photographs from other unknown faces.

He did not suffer from topographical agnosia: he could go to clinics of other departments for examinations, find the way back to his room on the ward, and draw a plan of the ward accurately; he could draw a map of Japan, and place the names of cities correctly. He did not have object agnosia: he could identify and name more than 50 objects or pictures of objects (bed, chair, glasses, cars, etc) presented in both canonical (conventional) and non-canonical (unconventional) views, and state the size and shape of presented pictures. He responded correctly to 45 out of 60 pictures of the Boston Naming Test (40–55 for the average Japanese). His language ability was normal as regards speaking, auditory comprehension, reading and writing. He neither displayed apraxias nor agnosias for the auditory and somatosensory modalities.



**Fig. 2** PET images of cerebral blood flow (CBF), oxygen extraction fraction (OEF), and cerebral metabolic rate of oxygen (CMRO<sub>2</sub>) at the horizontal planes 20–40 mm above and parallel to the orbitomeatal line (Headome IV, Shimadzu, Japan, with the full width at half-maximum of 4.5 mm). The CBF and CMRO<sub>2</sub> were determined using C<sup>15</sup>O<sub>2</sub> and <sup>15</sup>O<sub>2</sub>, respectively. The CBF and CMRO<sub>2</sub> are decreased in the right occipitotemporal areas (arrowheads). The right side of the head is on the left

When he was discharged 2 months after admission, his symptoms had partially recovered, and he could recognize faces of familiar persons whom he met before his illness. He could also recognize all photographs of families, relatives and old friends, and 90% of the photographs presented of famous people such as politicians, actors and professional wrestlers. He could discriminate genders and races by face. He could label emotional expressions on people and photographs as laughing, crying, or prim and proper. However, he remained unable to recognize faces of persons whom he met after the onset of illness. He identified these people only by their voice, height and hair. He still had difficulty in evaluating age by face. The cranial nerves, muscular strength, motor coordination, deep tendon reflexes and sensation were all normal throughout the course.

T2-weighted MRI (Fig. 1) demonstrated an area of hyperintensity in the cortical and subcortical areas in the right occipitotemporal areas, indicating an infarct in the territory of the posterior cerebral artery. The high-intensity area included the fusiform gyrus, lingual gyrus, tapetum, inferior longitudinal fascicle and a part of the optic radiation. In other areas, only a few lacunes were demonstrated. PET studies, using C<sup>15</sup>CO<sub>2</sub> and <sup>15</sup>O<sub>2</sub> inhalation methods, demonstrated a reduction in regional cerebral blood flow and cerebral metabolic rate of oxygen in the infarct areas indicated

on MRI (Fig. 2). There were no other areas of the brain with impaired blood flow or metabolism.

## Discussion

The main points to be addressed concerning the prosopagnosia of our patient are: (1) that it was relatively isolated; (2) that recognition of faces known to him before the illness recovered, whereas recognition of faces encountered after the illness remained impaired; and (3) that neuroimaging studies indicated morphological and functional changes in the occipitotemporal areas on the right side only.

The difficulty of our patient in recognizing faces could not be attributable to a less specific neuropsychiatric disturbance, because he had neither topographical agnosia nor object agnosia. Most patients with object agnosia are without prosopagnosia, and in some patients like ours, prosopagnosia is not associated with object agnosia [6, 12, 20], suggesting that the process of face recognition is distinct from that of object recognition. In addition to the preserved object recognition, our patient was able to identify familiar faces after his illness had partially recovered, and to interpret facial expressions from the beginning of his illness. The dissociations between deficits of familiar and unfamiliar face recognition [4, 9, 26], and between impairments in face identification and expression analysis [9, 37] have been described in previous reports. Unlike our patient, some patients with prosopagnosia were able to recognize unfamiliar faces [2, 5, 35, 42]. Such dissociations may be well explained by the functional model put forward by Bruce and Young [8], which assumes that recognition of familiar and unfamiliar faces uses different routes, and that face identification and expression analysis are processed independently. The inability of our patient to recognize familiar faces in the early phase of the illness may have occurred as a deficit of matching a perceived face to a memory store of the face that had been constructed; the residual deficit for unfamiliar faces appeared to be due to his inability to form memory stores of new faces. His failure to match unfamiliar faces photographed at different angles suggests that his inability to integrate abstract and structural visual codes from view-centred (pictorial) information of unfamiliar faces prevented further processing in the visual cognitive system.

Although his metamorphopsia for the eye region may have played some role in his prosopagnosia in the early period [5], it does not seem to have contributed to the prosopagnosia in the later period. This is because after partial recovery he could identify familiar faces, when he still complained of distortion of eye regions, although it was less remarkable than in the earlier period. Our patient also complained that the faces appeared red-tinted, and that the right eye appeared excessively bright. Similar dyschromatopsias have been described in patients with

disturbed perception of colours [28, 35]. The frequent association of prosopagnosia in patients with achromatopsias (12 out of 14 patients reviewed by Meadows in 1974 [29]) may suggest a close proximity of their pathogenetic loci. His low scores for picture arrangement in the WAIS tests suggest an impaired ability to discriminate essential parts from non-essential details. However, this cannot account for his prosopagnosia, because he could describe the characteristics of faces presented to him.

Experimental studies in monkeys indicated that visual information is relayed successively from the striate to the prestriate cortex, OB and OA (according to the terminology of Bonin and Bailey), and the posterior temporal areas, TEO and TE, and finally to the medial temporal lobe memory system, hippocampus, and the adjacent related cortex [31, 39]. This system has widespread reciprocal connections with the association cortex, where memories are recognized, consolidated and stored; as time passes, the memories stored in association cortex become independent of the medial temporal lobe [31, 39]. The visual field is re-represented in the striate and prestriate cortex and the posterior temporal areas, which in turn perform submodality processing. For face recognition, the face-sensitive cells have been demonstrated in the monkey [46]. Provided that facial information in the human is processed in a similar way, the isolated face recognition difficulty in our patient may be attributable to selective damage to one of the modalities in the prestriate area. The retrograde face memories may be executed independently from the medial temporal lobe memory system, if the striate and prestriate cortex and their connections are intact; the anterograde face memories may be selectively impaired if connections from the prestriate cortex to the medial temporal lobe are damaged. Thus, the dissociation between anterograde and retrograde face memories in our patient can be explained by the subcortical white matter lesion in the occipitotemporal areas, which may be located more anteriorly than in patients with both anterograde and retrograde prosopagnosia [41].

While most of the previous autopsy cases with prosopagnosia had bilateral cerebral lesions, some cases did not have left occipitotemporal lesions [18, 23, 33], suggesting that bilateral involvement of the visual system may not be a prerequisite for prosopagnosia [23]. This contention was further supported by many other reports based upon clinical and radiological findings [10, 13, 17, 21, 24, 28, 30, 40, 44]. In our patient, we could clearly rule out lesions and functional impairments in the left occipitotemporal areas, and in areas other than the right occipitotemporal areas. Although the mechanisms for prosopagnosia with a right occipitotemporal lesion remain to be clarified, previous researchers suggested a frequent co-occurrence of prosopagnosia and other agnosias for space which are associated with right posterior hemisphere lesions [22, 28]. The left-hemisphere template could be based on separate elements of the face, and have a

favoured position for verbal evocation, whereas the right-hemisphere template might be more global [12]. It has been suggested that while association or semantic categorization were impaired with left hemisphere damage, the visual identification and discrimination of complex patterns [14] and perceptual categorization [43] were deficient with right hemisphere lesions. Studies on normal subjects [15, 27] and split-brain patients [38] have demonstrated different strategies of facial recognition between the left and right brain. Thus, there may be various pathogenetic mechanisms of prosopagnosia, depending upon the location and size of the occipitotemporal lesions. In addition, there may be individual variation in the dependency of facial recognition on the right and left occip-

itotemporal areas complicated by different cognitive strategies. It is, therefore, possible that a small proportion of individuals whose left occipitotemporal area cannot assume the function of the corresponding areas on the right side may suffer from prosopagnosia. The findings in our patient indicate that relatively pure prosopagnosia and anterograde prosopagnosia with preserved retrograde facial recognition can occur with a lesion confined to the right occipitotemporal area.

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## References

1. Arseni C, Botez MI (1958) Consideraciones sobre un caso de agnosia de las fisionomias. *Rev Neuropsiquiatr Lima* 21: 583-593
2. Assal G (1969) Régression des troubles de la reconnaissance des physionomies et de la mémoire topographique chez un malade opéré d'un hématome intracérébral pariéto-temporal droit. *Rev Neurol (Paris)* 121: 184-185
3. Benson DF, Segarra J, Albert ML (1974) Visual agnosia-prosopagnosia: a clinicopathologic correlation. *Arch Neurol* 30: 307-310
4. Benton AL (1980) The neuropsychology of facial recognition. *Am Psychol* 35: 176-186
5. Benton AL, Van Allen MW (1972) Prosopagnosia and facial discrimination. *J Neurol Sci* 15: 167-172
6. Blanc-Garin J (1984) Perception des visages et reconnaissance de la physionomie dans l'agnosie des visages. *Année Psychol* 84: 573-598
7. Bodamer J (1947) Die Prosop-Agnosie (die Agnosie des Physiognomieerkennens). *Arch Psychiat Nervenkr* 179: 6-53
8. Bruce V, Young A (1986) Understanding face recognition. *Br J Psychol* 77: 305-327
9. Bruyer R, Laterre C, Seron X, Feyereisen P, Strypestein E, Pierrard E, Rectem D (1983) A case of prosopagnosia with some preserved covert remembrance of familiar faces. *Brain Cogn* 2: 257-284
10. Christen L, Landis T, Regard M (1985) Left hemispheric functional compensation in prosopagnosia? A tachistoscopic study with unilaterally lesioned patients. *Hum Neurobiol* 4: 9-14
11. Cohn R, Neumann MA, Wood DH (1977) Prosopagnosia: a clinicopathological study. *Ann Neurol* 1: 177-182
12. Damasio AR, Damasio H, Ban Hoesen GW (1982) Prosopagnosia: anatomic basis and behavioral mechanisms. *Neurology* 32: 331-341
13. De Renzi E (1986) Prosopagnosia in two patients with CT evidence of damage confined to the right hemisphere. *Neuropsychologia* 24: 385-389
14. De Renzi E, Scotti G, Spinnler H (1969) Perceptual and associative disorders of visual recognition: relationship to the side of cerebral lesion. *Neurology* 19: 634-642
15. Ellis HD, Shepherd JW, Davies GM (1979) Identification of familiar and unfamiliar faces from internal and external features: some implications for theories of face recognition. *Perception* 8: 431-439
16. Gloning I, Gloning K, Jellinger K, Quatember R (1970) A case of "prosopagnosia" with necropsy findings. *Neuropsychologia* 8: 199-204
17. Habib M, Sirigu A (1987) Pure topographical disorientation: a definition and anatomical basis. *Cortex* 23: 73-85
18. Hécaen H, Angelergues R (1962) Agnosia for faces (prosopagnosia). *Arch Neurol* 7: 92-100
19. Heidenhain A (1927) Beitrag zur Kenntnis der Seelenblindheit. *Monatsschr Psychiatr Neurol* 66: 61-116
20. Jeeves M A (1984) The historical roots and recurring issues of neurobiological studies of face perception. *Hum Neurobiol* 3: 191-196
21. Landis T, Cummings JL, Christen L, Bogen JE, Imhof HG (1986) Are unilateral right posterior cerebral lesions sufficient to cause prosopagnosia? Clinical and radiological finding in six additional patients. *Cortex* 22: 243-252
22. Landis T, Cummings JL, Benson DF, Palmer EP (1986) Loss of topographic familiarity: an environmental agnosia. *Arch Neurol* 43: 132-136
23. Landis T, Regard M, Bliegle A, Kleihues P (1988) Prosopagnosia and agnosia for noncanonical views. An autopsied case. *Brain* 111: 1287-1297
24. Lhermitte F, Pillon B (1975) La prosopagnosie: Rôle de l'hémisphère droit dans la perception visuelle (à propos d'un cas consécutif à une lobectomie occipitale droite). *Rev Neurol (Paris)* 131: 791-812
25. Lhermitte F, Chain F, Escourrolle R, Ducarne B, Pillon B (1972) Étude anatomoclinique d'un cas de prosopagnosie. *Rev Neurol (Paris)* 126: 329-346
26. Malone DR, Morris HH, Kay MC, Levin HS (1982) Prosopagnosia: a double dissociation between the recognition of familiar and unfamiliar faces. *J Neurol Neurosurg Psychiatry* 45: 820-822
27. Marzi CA, Berlucchi G (1977) Right field superiority for recognition of famous faces in normals. *Neuropsychologia* 15: 751-756
28. Meadows JC (1974) The anatomical basis of prosopagnosia. *J Neurol Neurosurg Psychiatry* 37: 489-501
29. Meadows JC (1974) Disturbed perception of colours associated with localized cerebral lesions. *Brain* 97: 615-632
30. Michel F, Poncet M, Signoret JL (1989) Les lésions responsables de la prosopagnosie sont-elles toujours bilatérales? *Rev Neurol (Paris)* 146: 764-770
31. Mishkin M (1982) A memory system in the monkey. *Philos Trans R Soc Lond [Biol]* 298: 83-95
32. Nardelli E, Buonanno F, Coccia G, Fiaschi A, Terzian H, Rizzuto N (1982) Prosopagnosia. report of four cases. *Eur Neurol* 21: 289-297

33. Pevzner S, Bornstein B, Loewenthal M (1962) Prosopagnosia. *J Neurol Neurosurg Psychiatry* 25:336–338
34. Pillon B, Bakchine S, Lhermitte F (1987) Alexia without agraphia in the left-handed patient with a right occipital lesion. *Arch Neurol* 44:1257–1262
35. Rondot P, Tzavaras A, Garcin R (1967) Sur un cas de prosopagnosie persistante depuis quinze ans. *Rev Neurol (Paris)* 117:424–428
36. Schwartz M, Smith ML (1980) Visual asymmetries with chimeric faces. *Neuropsychologia* 18:103–106
37. Shuttleworth EC Jr, Syring V, Allen N (1982) Further observations on the nature of prosopagnosia. *Brain Cogn* 1:307–322
38. Sperry RW (1982) Some effects of disconnecting the cerebral hemispheres. *Science* 217:1223–1226
39. Squire LR (1987) Memory: neural organization and behavior. In: Mountcastle VB (ed) *MD Handbook of physiology*, section 1, vol 5, part 1. American Physiological Society, Bethesda, pp 295–371
40. Torii H, Tamai A (1985) The problem of prosopagnosia: report of three cases with occlusion of the right posterior cerebral artery. *J Neurol* 232 [Suppl 140]
41. Tranel D, Damasio AR (1985) Knowledge without awareness: an autonomic index of facial recognition by prosopagnosia. *Science* 228:1453–1454
42. Tzavaras A, Merienne L, Masure MC (1973) Prosopagnosie, amnésie et troubles de langage par lésion temporaire gauche chez un sujet gaucher. *Encéphale* 62:383–394
43. Warrington EK, Taylor AM (1978) Two categorical stages of object recognition. *Perception* 7:695–705
44. Whiteley AM, Warrington EK (1977) Prosopagnosia: a clinical, psychological, and anatomical study of three patients. *J Neurol Neurosurg Psychiatry* 40:395–403
45. Wilbrand H (1892) Ein Fall von Seelenblindheit und Hemianopsie mit Sectionsbefund. *Dtsch Z Nervenheilkd* 2:361–387
46. Young MP, Yamane S (1992) Sparse population coding of faces in the inferotemporal cortex. *Science* 256:1327–1331