Facing up to a problem with recognition

Introduction

In 1891 Sigmund Freud introduced the term agnosia, meaning an absence of knowledge, to describe deficits of higher sensory processing which cause impaired recognition. These disorders are of great interest to neuropsychologists who try to characterize brain–behaviour interrelationships. Agnosias may be perplexing for both patients and clinicians, prompting an initial and understandable consideration of a primary sensory deficit (McCormick and Larner, 2018).

Agnosias may occur in any sensory modality but those affecting the visual domain are the most commonly encountered in clinical practice (Farah, 1995). Visual agnosias may result in impaired recognition of objects or of the written word, the latter manifesting as an acquired inability to read (alexia). More circumscribed visual agnosias may also occur, for example restricted to colours.

This article reports a patient who presented with a sudden onset of inability to recognize familiar faces.

Discussion

Prosopagnosia is a rare, circumscribed form of visual agnosia characterized by an inability to recognize previously known human faces or equivalent stimuli (Farah, 1995; Mayer and Rossion, 2007; Rivolta, 2014; Corrow et al, 2016; Larner, 2016). The defect is not necessarily limited solely to faces; it may encompass other categories such as animals ('zooagnosia'; Assal et al, 1984; Larner, 2016).

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Prosopagnosia is associated with lesions in the right inferior occipito-temporal region of the brain, particularly involving the lingual and fusiform gyri and subjacent white matter. This cerebral localization means that the disorder of facial recognition may be associated with other neurological features such as visual field defect (left upper quadrantanopia or homonymous hemianopia, although for the diagnosis of prosopagnosia to be made this should not be sufficient to produce a perceptual deficit) and/or achromatopsia, neither of which was evident in this patient. However, she did complain of visual distortions ('long noses') suggestive of metamorphopsia, which has on occasion been associated with prosopagnosia (e.g. Seron et al, 1995).

The aetiology of prosopagnosia is broad. It most often results from stroke (infarct or haemorrhage) but has also been reported on occasion in association with carbon monoxide poisoning, temporal lobectomy, encephalitis, neoplasm, trauma, and with Parkinson's disease and Alzheimer's disease (Mayer and Rossion, 2007). A form of frontotemporal dementia with focal right temporal atrophy may present with progressive prosopagnosia (Evans et al, 1995). A developmental or congenital form of prosopagnosia has also been described (e.g. Larner et al, 2003) and these cases have been increasingly studied in recent years to try to understand the underlying neuropsychological deficit responsible for the syndrome. To the authors' knowledge there is only one previous report of prosopagnosia in association with cerebral amyloid angiopathy (Hainline et al, 2017), and this appears to have been an inflammatory variant of this condition.

Prosopagnosia is sometimes characterized as 'face blindness' but this terminology may be a misnomer since patients are not blind

CASE REPORT

A 63-year-old right-handed woman presented with a new onset of visual problems, complaining of an inability to recognize faces. This had developed suddenly, about 3 months before her presentation to the neurology clinic. She was unable to discern family members or familiar TV characters by their face alone, although she reported that she could recognize them based on other cues such as the sound of their voices, general body habitus, movement and clothing. She said that faces appeared grey and blank with odd 'long noses'. Because of the visual symptoms she had attended an ophthalmology outpatient clinic, but no defect was found in visual acuity or on field testing or ophthalmoscopy. Her reading ability was unimpaired.

Her past medical history included a transient ischaemic attack and hypercholesterolaemia, treated with aspirin and atorvastatin respectively. She was an ex-smoker (25 pack years). There was no family history of similar visual problems or stroke.

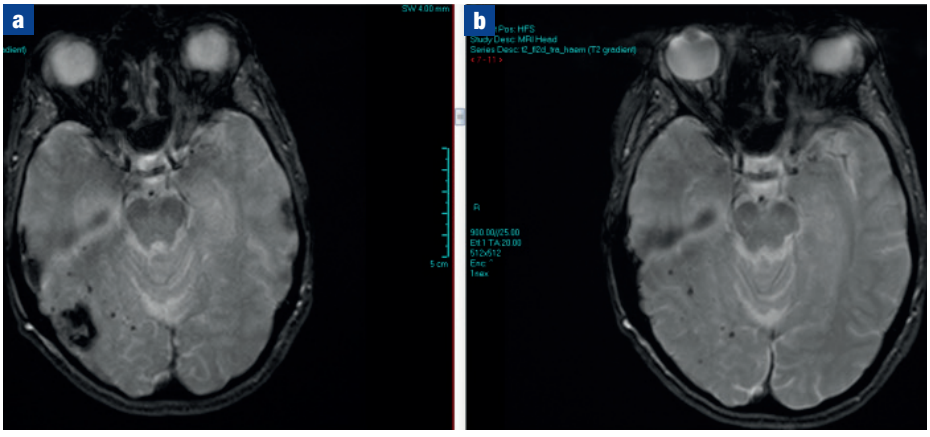
Neurological examination disclosed no focal abnormalities, in particular there was no visual field defect or impairment of colour vision.

Magnetic resonance imaging of the brain was performed. On standard sequences (not shown) there were some patchy punctate high signal lesions in the cortical white matter typical of small vessel ischaemia. However, on susceptibility weighted imaging, a technique of particular value in detecting blood products which distort the local magnetic field (Cheng et al, 2013), diagnostic changes were seen (*Figure 1*). There was a circumscribed area of signal change in the right inferior occipito-temporal region, indicative of a previous haemorrhage. In addition, there were a number of small dot-like areas of signal change, indicative of microbleeds.

A diagnosis of acute-onset prosopagnosia as a consequence of a right inferior occipito-temporal region cerebral haemorrhage was made. The combination of lobar haemorrhage and microbleeds indicated an underlying pathological diagnosis of cerebral amyloid angiopathy.

At follow up the patient continued to complain of difficulty discriminating faces in detail.

Figure 1. Axial susceptibility-weighted magnetic resonance brain imaging showing **(a)** a circumscribed area of signal change in the right inferior occipito-temporal region indicative of a previous haemorrhage, with **(a, b)** a number of separate, dot-like areas of signal change indicative of microbleeds, the combination suggesting an underlying pathological diagnosis of cerebral amyloid angiopathy.



LEARNING POINTS

- A deficit in the ability to recognize familiar faces in the absence of significant impairment of visual acuity or visual fields has been termed prosopagnosia.
- Prosopagnosia is rare, but the symptoms are characteristic.
- Prosopagnosia most commonly results from stroke (infarct or haemorrhage) affecting the right inferior occipito-temporal region.
- A developmental or congenital form of prosopagnosia is also described.
- Attempted rehabilitation has little to offer, but patients can often use other visual and non-visual cues to achieve recognition.

to all facial features. They may be able to identify emotional expression and eye gaze direction (Larner et al, 2003), and features such as hair or a moustache may be used to aid recognition. Patients with acquired prosopagnosia retain insight into their deficit in face recognition (Livingston and Shah, 2018).

Attempted rehabilitation in acquired prosopagnosia has achieved, at best, only modest results (Mayer and Rossion, 2007; Corrow et al, 2016). **BJHM**

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