CASE REPORT

Frontotemporal dementia: neuroanatomical correlates of an atypical presentation

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SUMMARY

Frontotemporal dementia (FTD) is a heterogeneous group of disorders characterised by frontal and temporal lobes atrophy. Three different clinical subtypes are recognised: behavioural variant, progressive non-fluent aphasia and semantic dementia. Neuroanatomical associations in a diffuse neurodegenerative disease such as FTD should be interpreted carefully; however, each FTD subtype has provided a clinical model that has contributed immensely to our understanding of clinical/neuroanatomical relationships. This case report and recent studies suggest that neuroanatomical findings concerning face-processing mechanisms in FTD can identify the brain regions that are critical for face processing. As seen in this case, right fusiform gyrus atrophy seems to be implied in the aetiology of prosopagnosia.

BACKGROUND

Frontotemporal dementia (FTD) is a clinically and pathologically diverse group of neurodegenerative diseases associated with focal frontal and temporal lobe atrophy.[1]

The Neary criteria recognise three clinical subtypes: behavioural variant, progressive non-fluent aphasia and semantic dementia.

We present a case of an atypical clinical presentation of FTD: prosopagnosia associated with behavioural changes.

CASE PRESENTATION

A 62-year-old man presented with behaviour and personality changes associated with progressive difficulty in recognising familiar faces, such as those of coworkers and famous people. He did not have any weakness, parkinsonism, bulbar symptoms or alien limb phenomenon. There was no family history of any significant illness and there was no history of any trauma or psychiatric illness.

INVESTIGATIONS

MRI showed marked right temporal anterior atrophy, with particular involvement of the right anterior fusiform and parahippocampal gyrus, best depicted in fluid attenuated inversion recovery (figure 1) and T2 (figure 2). Frontal and parietal lobes were relatively spared. MRI perfusion showed bilateral temporal hypoperfusion (figure 3).

Neuropsychological assessment showed frontal lobe dysfunction, verbal perseverance, mild memory impairment but visual and space perceptions were normal.

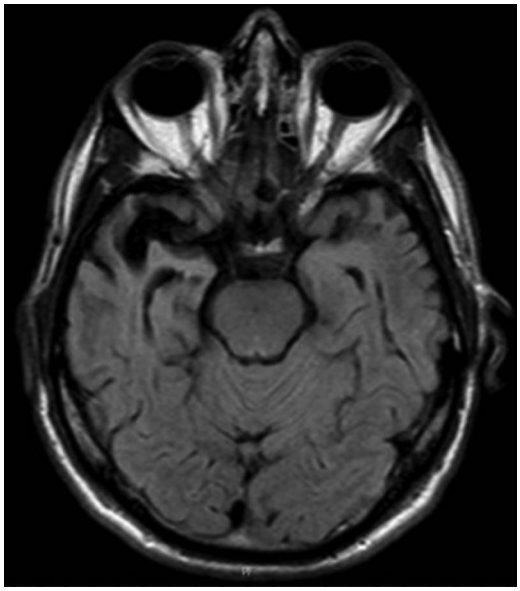

Figure 1 Axial fluid-attenuated inversion recovery: bilateral temporal atrophy with marked predominant right temporal atrophy.

Perceptual analysis of faces was assessed using the Benton facial recognition test. Prosopagnosia was assessed using famous faces test. Results were compatible with associative prosopagnosia.

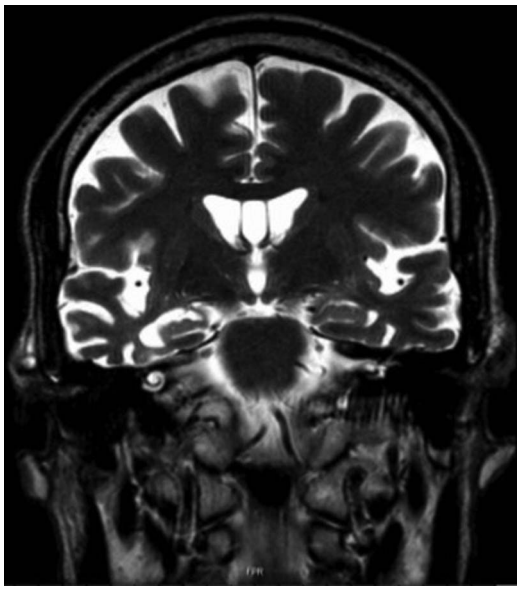

Figure 2 Coronal T2 weighted image: marked right temporal atrophy with widening of choroid fissure and temporal horn of lateral ventricle.

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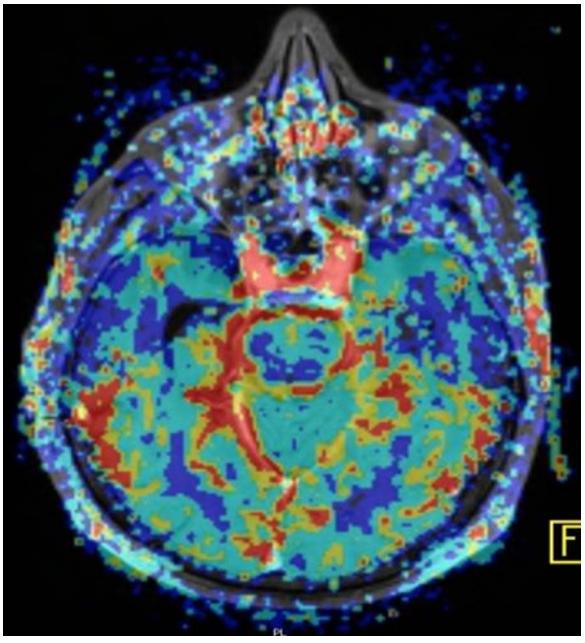

Figure 3 Cerebral blood flow: bilateral temporal hypoperfusion.

Single-photon emission CT cerebral perfusion disclosed reduced perfusion of anterior temporal lobes, predominantly of the right lobe and of the frontal lobes (figure 4). Cerebrospinal fluid was compatible with a diagnostic of FTD.

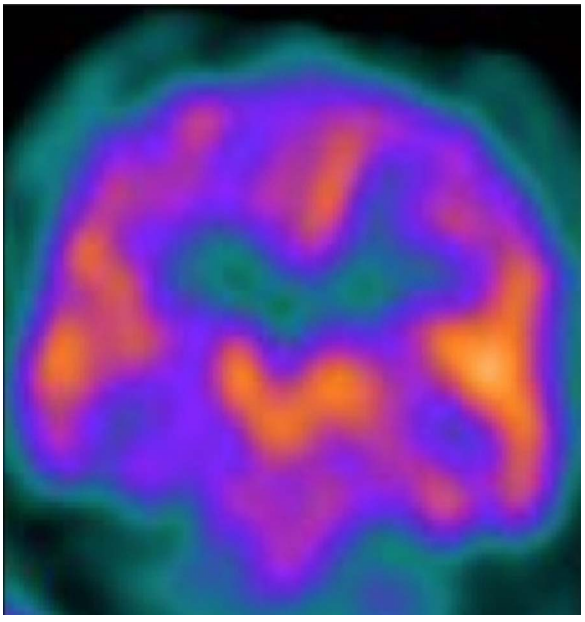

Figure 4 Single-photon emission CT (hexamethyl propylene amine oxine-Tc99 m): bilateral frontotemporal hypoperfusion with predominant right temporal hypoperfusion.

DIFFERENTIAL DIAGNOSIS

The patient presented early prosopagnosia, at the same time that he presented marked behavioural changes. According to the International Consensus Diagnostic criteria[2] the diagnosis of probable (right-sided) behavioural variant FTD was carried out. However, symptoms association seen in this case report is demonstrative of the overlapping and difficulty of FTD-subtype classification. As reported in recent studies[3–5] and in this case report, right fusiform gyrus atrophy seems to be implied in the aetiology of prosopagnosia.

OUTCOME AND FOLLOW-UP

With the disease progression the speech impairment evolved to anomia and the prosopagnosia became more severe, with only sons and wife being recognised, but interestingly the patient never presented visual agnosia.

DISCUSSION

Neuroanatomical associations in a diffuse neurodegenerative disease such as FTD should be interpreted carefully; however, this case report and recent studies suggest that neuroanatomical findings concerning face-processing mechanisms in FTD can identify the brain regions that are critical for face processing. The imaging findings in this case report corroborate previous evidence[3–5] that prosopagnosia appears to be associated with grey matter atrophy in the right fusiform gyrus, occurring in the context of bilateral temporal lobe atrophy.

Learning points

- Frontotemporal dementia (FTD) can be divided in three clinical syndromes: frontalvariant, progressive non-fluent aphasia and semantic dementia.
- Some overlap exists between the FTD-subtype classification.
- Right fusiform gyrus atrophy seems to be implied in the aetiology of prosopagnosia.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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