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A pathological fondness for noises with right temporal lobe atrophy

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Key point

Focal right temporal lobe degeneration presents clinically with changes in social and emotional behaviour and awareness, often accompanied by new obsessional interests despite loss of warmth toward people.

A 68 year old, right-handed retired farmer presented with a six-year history of progressive behavioural change. He had developed an intense liking for particular noises, specifically the engine sounds of Spitfires and other veteran aircraft which flew over his house from the local aerodrome. He would hasten outside to listen to them, standing transfixed and even weeping with pleasure. He also derived pleasure from certain car engine notes (V8 and V12), but found birdsong irritating; his liking for music had not noticeably changed. His fixation with engine sounds did not extend to a more general interest in aircraft or vehicles nor other kinds of obsessions. His wife was perplexed by this behaviour, which was out of keeping with his previously reserved personality and accompanied by uncharacteristic emotional coldness toward his family and friends as well as increased irritability, social disinhibition, faux pas, humourlessness and a sweet tooth. He was more attentive to punctuality and routines, and during the past year had sometimes failed to recognise acquaintances. There were no language or memory difficulties. He had mild age-related hearing loss but no other past medical or family history of note. When examined, he appeared somewhat impassive but there were no abnormal neurological findings. Neuropsychological assessment revealed impairments of facial identity and emotion recognition and executive dysfunction.

His brain MRI showed relatively selective atrophy of the right anteromesial temporal lobe (Figure 1).

The syndrome of right temporal lobe atrophy has emerged as a major variant presentation of frontotemporal dementia[1-4]. It encompasses features of both behavioural variant frontotemporal dementia (early disintegration of socio-emotional conduct and awareness) and semantic variant primary progressive aphasia (early prosopagnosia evolving to more generalised semantic deficits). Our patient exemplified this profile and illustrates another, less widely recognised but characteristic feature: development of intense, often highly specific obsessions with inanimate or abstract phenomena such as colours, puzzles, music, numbers, religion or time, despite waning empathy and interest in other people[2-4]. This abnormal hedonic behaviour may be accompanied by altered physiological reactivity[5] but is easily misinterpreted as a primary psychiatric problem, particularly where accompanying cognitive deficits are mild. Intense pleasure in inanimate sounds which we call ‘‘keladophilia’ (Greek, *kelados* [clamour] *philia* [love]) is an unusual auditory hedonic phenotype in

dementia[6, 7]. It has also been described in children with the neurodevelopmental disorder Williams' syndrome[8].

The syndrome of right temporal lobe atrophy is uncommon[10]. It is most often caused by accumulation of TAR-DNA-binding-protein (TDP)-43 type C, though various histopathological and genetic associations have been described[4]. Its pathophysiological mechanisms have not been clarified, however impairments of nonverbal semantic processing (especially knowledge of people and social concepts), theory of mind, homeostatic regulation and reward coding may all act synergistically. The right anterior temporal lobe normally integrates these diverse functions[3, 9], and right temporal lobe structures are involved in generating strong hedonic responses to sound and other sensory stimuli[6]. The syndrome should be suspected where behaviour changes (especially new fixations) develop in late middle life, however diagnosis currently relies on brain imaging.

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Competing interests

None declared.

Contributors

JVTH prepared and wrote the first version of the manuscript. All authors were involved in the critical revision of the manuscript. JJ and LCB were involved in the recruitment and data collection. JDW, YALP and RWP reviewed the manuscript from a neurological perspective. JDW provided the imaging and is the guarantor.

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Patient consent for publication

Consent obtained from next of kin.

Data availability statement

Data are available upon reasonable request to the corresponding author.

Ethical approval information

This study involves human participants and was approved by University College London and National Hospital for Neurology and Neurosurgery Joint Research Ethics Committees Participants gave informed consent to participate in the study before taking part.

Further reading

Belder CRS, Chokesuwattanaskul A, Marshall CR, Hardy CJD, Rohrer JD, Warren JD. The problematic syndrome of right temporal lobe atrophy: Unweaving the phenotypic rainbow. Frontiers in Neurology 2023; 13: 1082828. doi: 10.3389/fneur.2022.1082828.

Ulugut H, Bertoux M, Younes K, Montembeault M, Fumagalli GG, Samanci B, et al. Clinical recognition of frontotemporal dementia with right anterior temporal predominance: A multicenter retrospective cohort study. Alzheimers & Dementia 2024; 20: 5647-61. doi: 10.1002/alz.14076.

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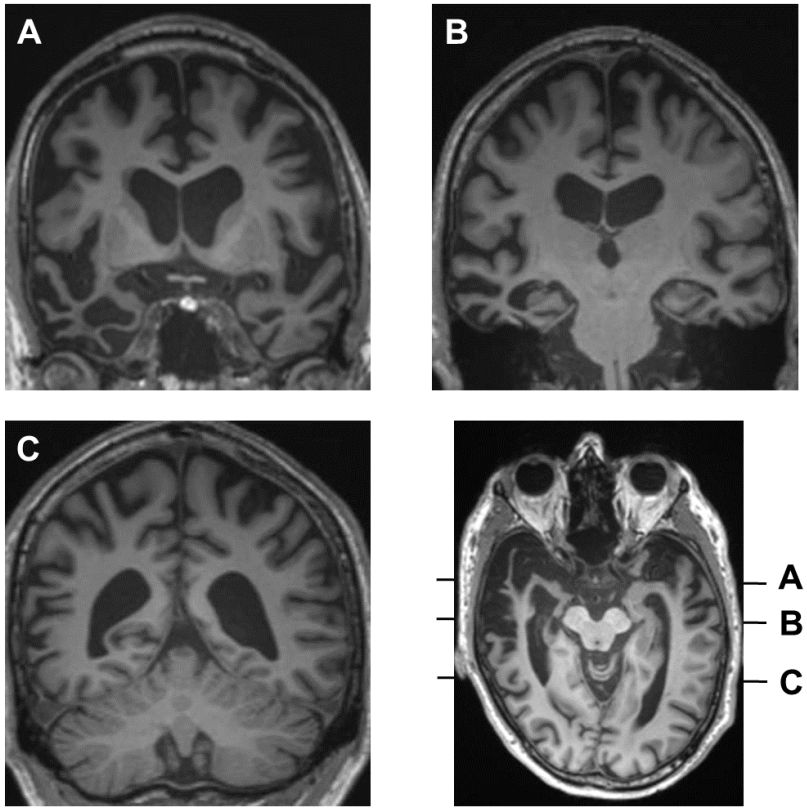

Figure 1. Representative coronal and axial sections of the patient's T1-weighted structural brain MRI. The right hemisphere is projected on the left side of each image. There is marked, relatively selective atrophy involving the right anterior temporal lobe, most severe in temporal polar, inferior temporal, fusiform and entorhinal cortices, amygdala and anterior hippocampus, with much less marked, mirroring atrophy of the left anterior temporal lobe and more diffuse background cerebral atrophy. Coronal sections at the level of the temporal poles (**A**), mid hippocampi (**B**) and temporo-parietal junctions (**C**) (see axial reference section, bottom right) show the striking gradient of atrophy within the right temporal lobe, with severe involvement anteriorly and relative sparing of more posterior areas.