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Clinical and radiological analysis of right temporal variant of frontotemporal dementia: A case report

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Abstract

Background: The right temporal variant of semantic dementia (rSD) is an anatomic variant characterized by a radiological pattern of asymmetric anterior temporal atrophy, with predominance of the right lobe. It constitutes a syndrome with primary progressive aphasia with differential cognitive and behavioral symptoms, the key manifestations being prosopagnosia, topographic agnosia, and impairment of socio-emotional functioning.

Case presentation: We describe the case of a 59-year-old man who developed a progressive cognitive decline in a six-year period. The first clinical manifestation was semantic anomia and difficulty recognizing objects; he later developed prosopagnosia, topographic agnosia, and progressive deterioration of episodic memory. These cognitive symptoms were accompanied by behavioral changes. Brain magnetic resonance imaging revealed asymmetric atrophy in the temporal poles, predominantly in the right hippocampal formations.

Discussion: The exposed case illustrates the clinical presentation of the right temporal variant of semantic dementia and how it correlates with specific neuroimaging findings. This correlation highlights the role of the right temporal lobe for developing multimodal concepts. Neuropathological and genetic characterization have been proposed as novel diagnostic approaches that might help distinguish right temporal variant of semantic dementia from other variants.

Conclusions: Although the right temporal variant of semantic dementia presents distinctive clinical manifestations, clinical diagnosis alone may be challenging due to overlapping features with other variants of frontotemporal dementia (FTD), highlighting the limitations faced in settings with little access to neuropathological and genetic evaluations and the need for more comprehensive characterization of clinical diagnostic criteria through longitudinal studies.

Keywords: Semantic dementia, Primary progressive aphasia, Right temporal lobe, Prosopagnosia, Topographic agnosia.

Análisis clínico y radiológico de la variante temporal derecha de la demencia frontotemporal: un reporte de caso

Resumen

Introducción: la variante temporal derecha de la demencia semántica (DS) es una variante anatómica caracterizada por un patrón radiológico de atrofia temporal anterior asimétrica, con predominio del lóbulo temporal derecho. Constituye un síndrome de afasia primaria progresiva con síntomas cognitivos y conductuales diferenciales, siendo los más característicos la prosopagnosia, la agnosia topográfica y el deterioro del funcionamiento socioemocional.

Presentación del caso: describimos el caso de un hombre de 59 años que desarrolló un deterioro cognitivo progresivo en un período de seis años. La primera manifestación clínica fue anomia semántica y dificultad para reconocer objetos; posteriormente desarrolló prosopagnosia, agnosia topográfica y deterioro progresivo de la memoria episódica. Estos síntomas cognitivos estuvieron acompañados de cambios conductuales. La resonancia magnética cerebral reveló atrofia asimétrica en los polos temporales, predominantemente en las formaciones hipocampales derechas.

Discusión: el caso expuesto ilustra la presentación clínica única de la demencia semántica derecha y cómo se correlaciona con hallazgos de neuroimagen específicos. Esta correlación resalta el papel del lóbulo temporal derecho para desarrollar conceptos multimodales. La caracterización neuropatológica y genética se han propuesto como enfoques diagnósticos novedosos que podrían ayudar a distinguir la demencia semántica derecha de otras variantes.

Conclusiones: aunque la demencia semántica derecha presenta manifestaciones clínicas distintivas, el diagnóstico clínico por sí solo puede ser un desafío debido a la superposición de características con otras variantes de demencia frontotemporal (DFT), lo que resalta las limitaciones que se enfrentan en entornos con poco acceso a evaluaciones neuropatológicas y genéticas, y la necesidad de una caracterización más completa de los criterios diagnósticos clínicos a través de estudios longitudinales.

Palabras clave: demencia semántica, afasia primaria progresiva, lóbulo temporal derecho, prosopagnosia, agnosia topográfica.

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Introduction

Frontotemporal lobar degeneration (FTLD) is a neuropathological term used to describe a spectrum of neurodegenerative disorders affecting the frontal and temporal regions, clinically manifesting as frontotemporal dementia (FTD) (1). These disorders exhibit diverse clinical manifestations, delineating three primary syndromes: the behavioral variant of frontotemporal dementia (bvFTD) and two language-related variants—non-fluent or agrammatic primary progressive aphasia (PPA) and semantic dementia (SD) (1,2).

Semantic dementia manifests with a progressive deterioration of language, characterized by anomia, with preservation of phonology and grammar, and impaired comprehension of word meanings and concepts (3). Over time, this impairment extends to other sensory modalities, culminating in a multimodal disorder of semantic memory, while episodic memory remains relatively preserved (4,5).

Typically, semantic dementia is associated with bilateral atrophy of the anterior temporal lobes, predominantly affecting the left hemisphere (6). However, in approximately 30% of cases, this classical pattern is absent, and there is a greater involvement of the right hemisphere, defined as the right temporal variant of semantic dementia, characterized by distinct clinical features (7,8).

Case presentation

We present the case of a 59-year-old right-handed man with 11 years of formal education who worked as a driver and had a medical history of non-Hodgkin lymphoma, ankylosing spondylitis, hypertension, and chronic alcoholism, with no family history of dementia. His family sought outpatient neurology consultation due to a progressive cognitive decline over 6 years, associated with behavioral changes.

The first symptoms emerged at the age of 53, with difficulty naming objects, which gradually progressed to an inability to distinguish everyday items. He frequently confused items such as combs and brooms and was unable to recognize items while shopping. Concurrently, he developed topographic agnosia, struggling to navigate familiar routes despite his experience as a driver, and could only reach destinations when given step-by-step instructions.

By the age of 57, he developed prosopagnosia, recognizing people only by their voices. As the disease advanced, he required third-party confirmation to recognize acquaintances. During this period, episodic memory impairment became evident, characterized by repetitive questioning and frequent misplacement of personal belongings. Besides cognitive decline, behavioral changes emerged, including expansive mood, disinhibition with excessive familiarity, inappropriate humor with sexually suggestive remarks, and atypical behaviors such as hoarding garbage at home.

At 59 years of age, the patient was disoriented in time and space, hypoprosexic, and presented with fluent speech. Surface dyslexia was apparent with words like Renault, and he struggled to describe and name elements in the Cookie Theft picture. He was unable to draw named objects (e.g., flower, cat) or define nouns (e.g., scarf, bee, elephant). The brain magnetic resonance imaging (MRI) revealed significant atrophy in the temporal poles and hippocampal formations, predominantly in the right hemisphere, with lesser involvement of the frontal and parietal lobes. Additional findings included thinning of the corpus callosum, white matter gliosis in the right temporal pole, and atrophy with gliosis in the right pericalcarine region (Figure 1).

Neuropsychological assessment confirmed partial disorientation in time and space, mild attentional deficits, anterograde amnesia (both verbal and visual), altered semantic knowledge, prosopagnosia, and topographic agnosia. Executive dysfunction, social disinhibition, and expansive mood were also identified. These deficits led to mild impairment in instrumental activities of daily living.

A speech therapist identified semantic difficulties, with impaired access to object names and minimal benefit from either semantic or phonological cues. Visual agnosia was also suspected, as the patient frequently reported being unable to recognize objects. Therefore, considering the symptoms, imaging, and conclusions from neuropsychology and speech therapy, a diagnosis of right temporal variant of semantic dementia (rSD) was established.

A cognitive rehabilitation plan was designed, with an emphasis on multisensory object recognition. Pharmacological management was initiated with quetiapine and sodium valproate to address behavioral disturbances. At the one-year follow-up, there was a slight improvement in disinhibition; however, cognitive and functional deterioration persisted. Anomia and prosopagnosia worsened, ultimately leading to

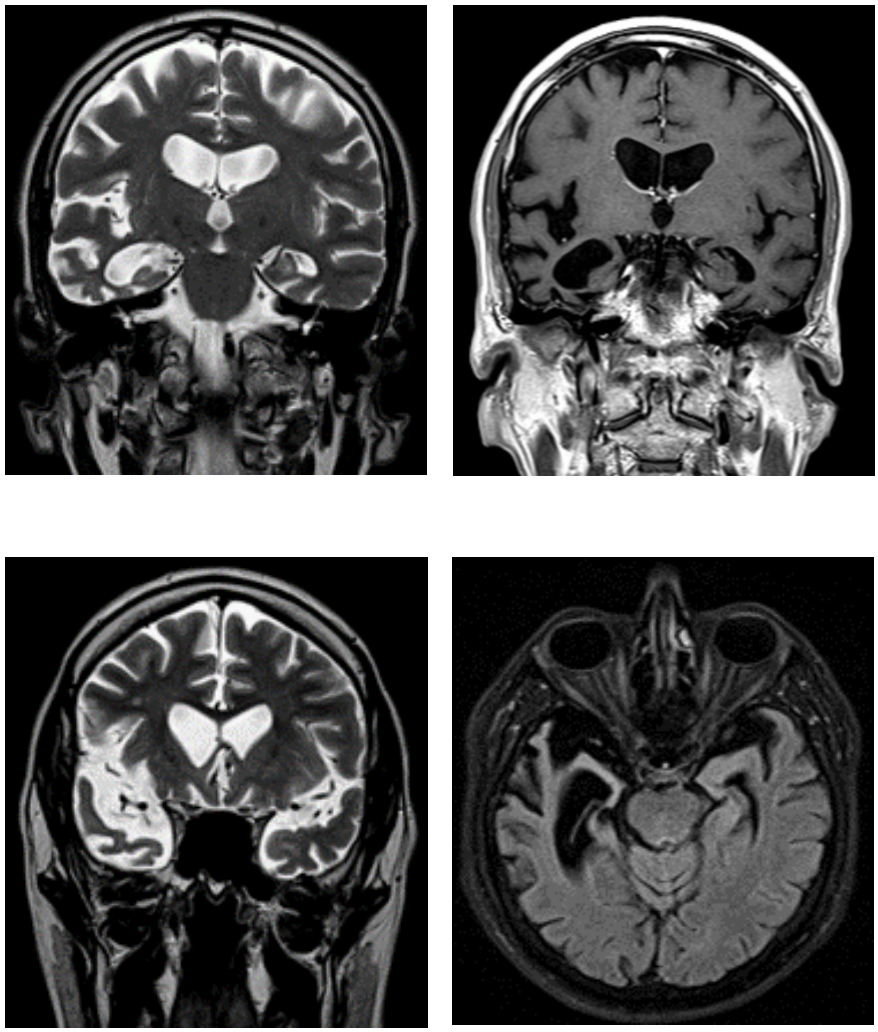

Figure 1. The brain MRI performed in 2021 (6 years after symptom onset) showing predominantly right-sided temporal lobar atrophy

Note. (A and C) coronal T2-weighted, (B) coronal, T1-weighted, (D) axial, T1-weighted.

Source: own elaboration.

the patient's inability to recognize close family members. Additionally, depressive and anxious symptoms emerged, seemingly related to his awareness of the disease. He continues annual follow-ups with neurology.

Discussion

Semantic dementia is generally characterized by fluent aphasia with anomia and impaired understanding of meanings. At early stages, atrophic changes are evident in the anteromedial temporal region, predominantly in the left hemisphere (9). Broadly speaking, right asymmetric degeneration correlates with a particular clinical presentation that not only features semantic language impairment but also encompasses behavioral symptoms, deterioration of other cognitive domains, and specific manifestations such as topographic agnosia and prosopagnosia (9). These clinical differences have led to the analysis of the clinical-radiological correlation of the rSD.

In general, the radiological pattern comprises asymmetric atrophy of temporal poles, middle and inferior temporal gyri, and insula, with right-hemisphere

predominance. Ipsilateral frontal atrophy may also be observed, specifically in the inferior frontal gyrus, straight gyrus, and orbitofrontal lobe (10,11). In the present case, the asymmetry and right predominance of hippocampal atrophy were quite evident, while frontal atrophy on the ipsilateral side was significantly less pronounced. Like most patients reported in the literature, there is involvement of the same left temporal regions but to a lesser extent (11), with discrepancies between both sides of more than 10% (10).

It has been found that left-sided variants with language impairment present a mirrored radiological pattern. However, although the involvement occurs in analogous contralateral areas, the clinical presentations of rSD and left-sided SD differ widely, even in advanced stages (3). On the one hand, the altered structures on the left side are essential for verbal and linguistic information, integrating sounds and letters into meaningful words; therefore, impairment in this region manifests as difficulty recognizing objects and generalizing the categories to which they belong (3,8). On the other hand, concepts predominantly integrated on the right side relate to non-verbal and visceral information, attributing value to social behaviors; thus, misinterpretation of these stimuli leads to lack of empathy, apathy, and behavioral traits (8).

Therefore, rSD is characterized by poorer performance in behavioral control (11) and earlier neuropsychiatric symptoms compared to left-sided SD (12,13), which may also present behavioral changes, but typically in advanced stages of the disease. This type of symptomatology in left-sided SD is usually correlated with greater degeneration in the right and frontal regions (5). In this regard, it should be noted that the typical radiological pattern of the rSD comprises predominantly right frontotemporal involvement, rather than being limited solely to the temporal lobe (10).

This aligns with the behavioral changes that were evident in our patient from the early stages of the disease, even though they were not the initial presenting symptoms. The predominant behavioral manifestation in our case was disinhibition, described in the literature as one of the main behavioral traits in right-sided variants (51-60%) (3,11). Other manifestations frequently mentioned include apathy (39-55%), compulsive behaviors (18-40%), loss of empathy (24-50%), changes in eating preferences (22-30%), as well as irritability or aggression (25-40%) and depression (28-45%) (11,14,15). In the case of our patient, hoarding behaviors were present, and as the disease progressed, depressive symptoms developed.

On the other hand, the initial and main complaint of our patient was anomia, characterized by fluent speech but semantic difficulties in neuropsychological evaluations. Studies regarding this variant have shown severe impairment in semantic language, as evidenced by deficits in tests such as the Boston Naming Test, as well as in verbal fluency, with greater impairment of semantic than lexical fluency. Additionally, episodic memory and executive functions may also be affected, but to a lesser extent (3,8), consistent with the neuropsychological evaluations of our patient.

Prosopagnosia is described as a feature that distinguishes right-sided semantic dementia from other variants of FTD. It is reported to affect up to 60-70% of patients with right-sided predominance (10,11,13), and it was one of the most evident manifestations in our patient. Regarding this issue, and related to the behavioral manifestations, several neuropsychological analyses have reported significant deterioration in the socioemotional functioning of patients with right-sided SD, including poor insight, alterations in recognizing emotions in faces, low scores on empathy scales, social awkwardness, and socioemotional sensitivity, which is not affected in left-sided variants (3,8).

In terms of clinical-radiological correlation, it has been shown that rSD patients with prosopagnosia exhibit greater bilateral volume loss in temporal poles and the fusiform gyrus (11,16), extending to the right ventral occipitotemporal cortex (17). These regions are related to the facial recognition process that includes perception formation, encoding, and comparison with stored memories, allowing access to semantic information (18). Thus, it is known that different brain regions represent individual domains of knowledge about an object (e.g., shape, sound, utility), while the bilateral anterior temporal lobes play a key role in integrating these different fragments of information into a whole multimodal concept (3).

Our patient was unable to recognize the faces of his relatives but initially managed to do so by their voices, a skill that was lost as the disease progressed. This is particularly relevant, as voice recognition is usually compromised, which correlates with the

multimodal semantic impairment previously exposed (3,9). This is also related to another of the patient's manifestations: difficulty recognizing everyday objects, which may be attributed to visual agnosia.

Additionally, involvement of various regions such as the lingual gyrus, posterior parietal, cingulate, and parahippocampal cortices can lead to disorientation due to topographic agnosia, another distinctive feature of this particular variant. Our patient presented this type of agnosia as one of the initial symptoms, with difficulty reaching known places when given the names of reference points, stemming from an inability to recognize the concept of structures used for orientation. This manifestation is typically associated with bilateral or right-sided lesions, and in this case, the patient exhibited significant atrophy of the right hippocampal formation (9).

There is ongoing debate regarding the nosological classification of this clinical presentation. While some authors define right-sided cases as a mirrored variant of SD, others consider them a distinct subtype within the FTD spectrum. Some authors support the former view, emphasizing anatomical symmetry, while others argue for a separate classification, often referred to as the right temporal variant of FTD, based on clinical, radiological, and pathological distinctions. Ramírez et al. highlight this controversy, referencing a study that identified two subgroups among patients with right temporal atrophy: a behavioral variant linked to tau pathology and a semantic variant characterized by prosopagnosia, anomia, and topographic agnosia, associated with TDP-43 proteinopathy (14,19). This supports the hypothesis that rSD may not simply reflect contralateral involvement but rather a separate clinicopathological subtype with a distinct genetic basis.

Recognizing this distinction is crucial to improve classification frameworks and diagnostic accuracy, where multiple potential confounding factors may influence cognitive performance, such as a history of lymphoma, alcohol use, and hypertension. Additionally, there was no family history of neurodegenerative diseases. In this context, genetic or pathological confirmation access would have been especially valuable for diagnostic certainty. This poses an important limitation for our case study, as the diagnosis relies solely on clinical and imaging findings. Our patient was seen through a private consultation at a research center. Furthermore, insurance-related barriers prevented genetic testing. These limitations reflect the reality faced by many patients with rare neurodegenerative variants in low and middle-income countries, where comprehensive diagnostic resources remain scarce. This highlights the need for broader diagnostic access and for longitudinal studies to improve the characterization of rSD and its overlap with other FTD spectrum variants.

Conclusions

The presented case illustrates the unique clinical and neuropsychological features of rSD and its association with a distinctive asymmetric radiological pattern. This correlation underscores the critical role of the temporal region in integrating various perceptions to develop meaningful multimodal concepts.

Nonetheless, the case also highlights the diagnostic hurdles encountered in low to middle-income settings, as clinical diagnosis alone may be challenging due to overlapping features with behavioral FTD and left-sided SD. Therefore, there is a pressing need for more comprehensive characterization through longitudinal studies to tailor individualized therapeutic interventions effectively.

Authors' contributions. María Fernanda Acosta: data curation, formal analysis, methodology, investigation, writing-original draft, writing-review and editing, visualization; Juan Felipe Quintero-Moreno: conceptualization, data curation, methodology, investigation, resources, writing-original draft; Margarita Giraldo: data curation, resources, supervision, validation; Daniela Torres Urazán: conceptualization, data curation, investigation, writing-original draft; Liliana Hincapié-Henao: data curation, resources, validation; Yesica Zuluaga-Castaño: data curation, resources, validation; David Aguillon: project administration, supervision, validation; Francisco Lopera: project administration, supervision, validation.

Ethical implications. This case report was approved by the ethics committee of the medical research institute of the University of Antioquia. The data used for this article are not publicly available due to them containing information that could compromise the patient's privacy and anonymization. All data used were taken from the private and confidential medical history of the pa-

tient. Written informed consent for publication of medical records, including images was obtained from the patient and the patient's son.

Conflicts of interest. The authors declare that they have no competing interests.

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