Geschwind Syndrome in frontotemporal lobar degeneration: Neuroanatomical and neuropsychological features over 9 years

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A B S T R A C T

Geschwind Syndrome, a characteristic behavioral syndrome frequently described in patients affected by temporal lobe epilepsy (TLE), consists of the following features: hyper-religiosity, hypergraphia, hyposexuality, and irritability. Here we report the 9-year-clinical course of a case of Geschwind Syndrome that developed as a first and salient clinical expression of right temporal lobe variant of frontotemporal lobar degeneration (FTLD). Only one patient affected by frontotemporal dementia has previously been shown to present with Geschwind Syndrome.

MS presented at age 73 with 3 years of personality and behavioral symptoms. Her early symptoms primarily included hyper-religiosity, hypergraphia, and poor emotional regulation (irritability, impulsivity, disinhibition, egocentric behavior). Over nine years, other cognitive functions (word retrieval, memory coding and recall, set-shifting, famous face and building recognition) became affected; however, hyper-religiosity, hypergraphia, and scarce emotional control remained her most prominent deficits. Longitudinal cortical thickness and volumetric analyses revealed early atrophy in the right temporal pole, right amygdala, and right hippocampus, which progressively affected homologous regions in the left hemisphere. The present case describes an unusual clinical picture associated with frontotemporal dementia (FTD), in which the most salient symptoms originated and remained consistent with Geschwind Syndrome.

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1. Introduction

Starting with their seminal works published in 1974 and 1975, Waxman and Geschwind recognized that patients affected by temporal lobe epilepsy (TLE) could develop a specific constellation of symptoms that influenced their affect, personality, and cognition (Bear & Fedio, 1977; Waxman & Geschwind, 1974, 1975; see also; Devinsky & Schachter, 2009). In most cases, the characteristic behavioral syndrome described is interictal (i.e., present constantly, without a
specific relationship to individual seizures) and consists of the following features: hyper-religiosity (increased interest in philosophical, moral, and religious issues), hypergraphia (excessive compulsive writing often of a religious or philosophical nature), hyposexuality, and irritability of varying degrees. Geschwind attributed the syndrome to limbic system damage that accrued during seizures (Waxman & Geschwind, 2005). In the past few decades, several group studies and case reports have described one or more features of what is now referred to as Geschwind Syndrome (Garcia-Santibanez & Sarva, 2015; Hermann, Whitman, & Arntson, 1983; Okamura et al., 1993; Roberts, Robertson, & Trimble, 1982; Sachdev & Waxman, 1981; Tebartz van Elst et al., 2003; Woollacott et al., 1993; Roberts, Robertson, & Trimble, 1982; Sachdev & Waxman, 1981; Tebartz van Elst et al., 2003; Wuerfel et al., 2004). Some authors have suggested a major involvement of the right hemisphere, with hyper-religiosity and hypergraphia occurring more frequently in patients with non-dominant hemisphere TLE (Roberts et al., 2015; Wuerfel et al., 2004). Some other studies have more specifically implicated bilateral (Tebartz van Elst et al., 2003) or right hemisphere (Wuerfel et al., 2004) hippocampal damage in the manifestation of Geschwind Syndrome in TLE.

Although the behavioral triad of hyper-religiosity, hypergraphia, and hyposexuality is considered unique to TLE, some patients with frontotemporal lobar degeneration (FTLD) demonstrate symptoms that partially overlap with Geschwind Syndrome. The right-temporal lobe variant (RTLV) of FTLD, which is characterized by progressive atrophy that is most prominent in the right anterior temporal lobe often associated with impaired emotional regulation and social cognition and behavior, has been associated with hyper-religiosity in 15% of cases studied (Chan et al., 2009). In this case series by Chan and colleagues, one patient exhibited both hyper-religiosity and hyposexuality (but not hypergraphia). A separate recent case report described three patients with the semantic variant of primary progressive aphasia and new-onset creative writing behavior that was similar to hypergraphia (Wu et al., 2013). To date, only one patient who met the diagnostic criteria for Frontotemporal Dementia (FTD) without epilepsy and who presented with Geschwind Syndrome symptoms has been reported in the literature (Postiglione et al., 2008).

Here, we describe the 9-year-clinical course of a patient with Geschwind Syndrome that developed as the first, most salient clinical expression of RTLV of FTLD. We report the disease progression through repeated neuropsychological assessments and longitudinal neuroimaging analysis.

2. Case description

MS, a 73-year-old, left-handed, English speaking woman presented with a 3-year history of progressive behavioral changes. MS had 12 years of education and worked as a cashier and secretary until she had children. When her children were older, she returned to work in the school system as a secretary and nurse. She first came to medical attention due to concerns that she might have Alzheimer’s Disease, although at that time no evidence of any cognitive abnormality was found. She was subsequently evaluated in the Frontotemporal Disorders Unit at Massachusetts General Hospital in Boston. During the visit, MS denied any significant problems with her memory, stating only that she occasionally went into a room in her home and forgot what she was intending to do. She denied difficulty recalling conversations, names, or words in discourse. She also denied difficulty with all activities of daily living, including dressing, hygiene, money management, and meal preparation. She continued to drive without impairment, and stated that she had never become lost or disoriented in a familiar area. The patient’s daughter concurred with the observation of no change in cognitive abilities, but reported a significant change in her mother’s emotional functioning throughout the preceding years. Specifically, she reported that her mother became angry much more frequently than she used to, without provocation, and that the level of her expressed anger was much more intense. This did not fit her prior emotional state, which was described by her family as gentle and kind. MS also appeared to have become more self-centered, and would often bring conversations back to herself. We formally quantified MS’s personality change by administering the NEO-Five Factor Inventory (NEO-FFI; Costa & McCrae, 1992); see Table 1. The patient and her daughter each completed NEO-FFI ratings for MS’s current and premorbid personality. The daughter’s assessment suggested changes from the patients’ premorbid personality in the form of decreased extraversion, openness to new experience, agreeableness, conscientiousness, and increased neuroticism. In contrast, the patient rated herself inversely, revealing a lack of insight into her changes in personality.

The patient’s daughter also reported that MS had become more religious: although she had been a lifelong churchgoing Catholic, she had recently begun watching religious shows, reading religious books, and talked about God much more frequently than previously. When asked about her current mood, MS reported that she was tired, busy, and had a lot to do, although she would still describe her mood as “happy.”

<table>
<thead>
<tr>
<th>Rater</th>
<th>Condition</th>
<th>Neuroticism</th>
<th>Extraversion</th>
<th>Openness</th>
<th>Agreeableness</th>
<th>Conscientiousness</th>
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<tr>
<td>Daughter</td>
<td>Premorbid</td>
<td>35</td>
<td>55&lt;sup&gt;a&lt;/sup&gt;</td>
<td>45&lt;sup&gt;a&lt;/sup&gt;</td>
<td>67&lt;sup&gt;a&lt;/sup&gt;</td>
<td>66</td>
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<td></td>
<td>Current</td>
<td>39</td>
<td>34&lt;sup&gt;a,b&lt;/sup&gt;</td>
<td>25&lt;sup&gt;a,b&lt;/sup&gt;</td>
<td>&lt;1&lt;sup&gt;b&lt;/sup&gt;</td>
<td>46&lt;sup&gt;b&lt;/sup&gt;</td>
</tr>
<tr>
<td>MS</td>
<td>Premorbid</td>
<td>37&lt;sup&gt;a&lt;/sup&gt;</td>
<td>53&lt;sup&gt;b&lt;/sup&gt;</td>
<td>42&lt;sup&gt;b&lt;/sup&gt;</td>
<td>51&lt;sup&gt;b&lt;/sup&gt;</td>
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<td>63&lt;sup&gt;a,b&lt;/sup&gt;</td>
<td>64&lt;sup&gt;b&lt;/sup&gt;</td>
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Scores are t values for women.

<sup>a</sup> Indicates that premorbid and current scores differ categorically in terms of NEO norms.

<sup>b</sup> Indicates that the patients’ scores and caregiver’s scores differ categorically in terms of NEO norms.
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