CASE REPORT

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EARLY ONSET PROGRESSIVE NONFLUENT APHASIA

ERKEN BAŞLANGIÇLI PROGRESİF TUTUK AFAZİ

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Abstract

Progressive nonfluent aphasia is a slowly progressive degenerative disease characterized by atrophy in left hemisphere particularly frontotemporal. It is one of three subtypes of frontotemporal lobar degeneration (frontotemporal dementia). Unlike Alzheimer's disease it begins between 45-65 years of age and occurs equally in both sexes usually. The reported youngest case was 21 years old. Atrophy is seen in the left hemisphere more in temporal lobe on magnetic resonance imaging. Approximately half of the cases have family history. In early it might confuse with depression and therefore diagnosis may be delayed. Brain magnetic resonance imaging is important for verification of diagnosis. In this paper, a case who early onset progressive nonfluent aphasia was mentioned.

Keywords: Frontotemporal dementia, Progressive nonfluent aphasia, depression

Özet

Progresif tutuk afazi, sinsi başlangıçlı, yavaş ilerleyen, sol hemisferde (özellikle frontotemporal) atrofi ile seyreden dejeneratif bir hastalıktır. Frontotemporal lobar dejenerasyonun (frontotemporal demans) üç alt tipinden biridir. Alzheimer hastalığının aksine genelde 45-65 yaş arasında başlar ve her iki cinsiyette eşit oranda görülür. Bildirilmiş en genç olgu 21 yaşındadır. MRI'da sol hemisferde daha çok temporal lobda atrofi görülür. Olguların yaklaşık yarısında aile öyküsü vardır. Erken dönemlerde depresyon ile karıştırılabilir. Bu nedenle tanıda gecikilebilir. Beyin MRI tanıyı doğrulamada önemlidir. Bu makalede erken başlangıçlı bir progresif tutuk afazi olgusu sunulmuştur.

Anahtar Kelimeler: *Frontotemporal demans, progresif tutuk afazi, depresyon*

1. Introduction

Frontotemporal dementia (FTD) is the most common group of clinical syndromes associated with circumscribed degeneration of the anterior temporal and prefrontal lobes (Neary et al., 2005). It has been called frontotemporal lobar degeneration (FTLD) same time and non-Alzheimer disease type pathology. Behavioural changes are the presenting feature. It is dominate the clinical picture throughout the disease course. Cognitive impairments in executive function and qualitative changes in language occur. The absence of early neurological signs and findings of focal abnormalities in the frontotemporal lobes on neuroimaging contribute to diagnosis (Neary et al.,1998).

Progressive nonfluent aphasia (PNFA) may be speech production is effortful and halting, with speech sound (phoneme) errors, and simplified or "agrammatic" productions. Word and simple sentence comprehension, as well as recog¬nition of nouns are preserved. Although usually with some loss of comprehension for complicated syntactic constructions. Patients with PNFA rarely have underlying Alzheimer disease. Most have a tauopathy. This is especially true of patients who manifest both apraxia of speech and agrammatism. Language variant may appear relatively non-fluent, but they do not have the more specific features of agrammatism and apraxia of speech in the some patients (Rohrer et al., 2010).

2. Case

A 33-year-old male patient was admitted to the clinic with depressive symptoms. He divorced for years ago. He was treated with antidepressants for 2 years. It had been an increase in his depressive symptoms for the last one year. He also complained about forgetfulness, difficulty understanding, unwillingness, sadness and suicidal thoughts. His speech was hypophonic, slow and includes phonemic paraphasias. He occasionally stammered. He was apathic. In the neurological examination, could not be found a problem. His Hamilton Depression Rating Scale point was 18. His mother was diagnosed with dementia when she was 55 years old. Left hemispheric cortical atrophy and lateral ventricular asymmetry were detected in his cranial magnetic resonance imaging (MRI) (Figure 1, Figure 2, and Figure 3). Increase in theta-delta and alpha frequency was detected in his quantitative electroencephalography (QEEG) without any

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paroxysmal activity (Figure 4). Neuropsychological tests demonstrated impairment in cognitive functions. He was diagnosed with progressive non fluent aphasia.

Figure 1: MRI shows left temporal atrophy

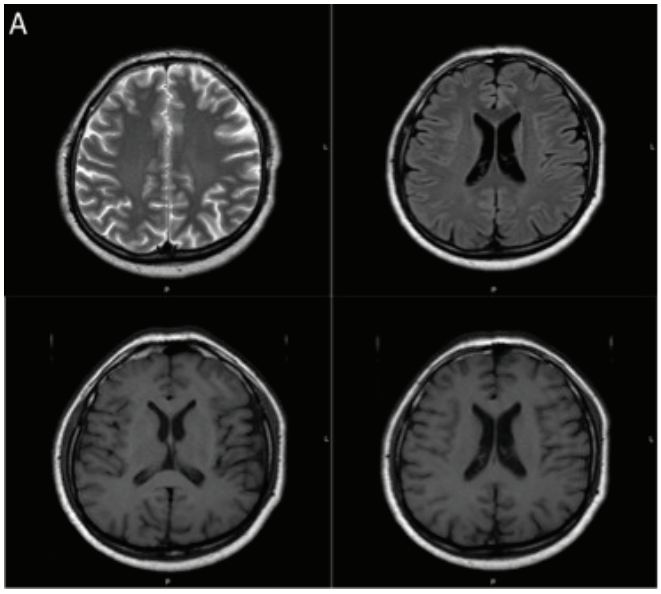


Figure 2: MRI shows left temporal atrophy

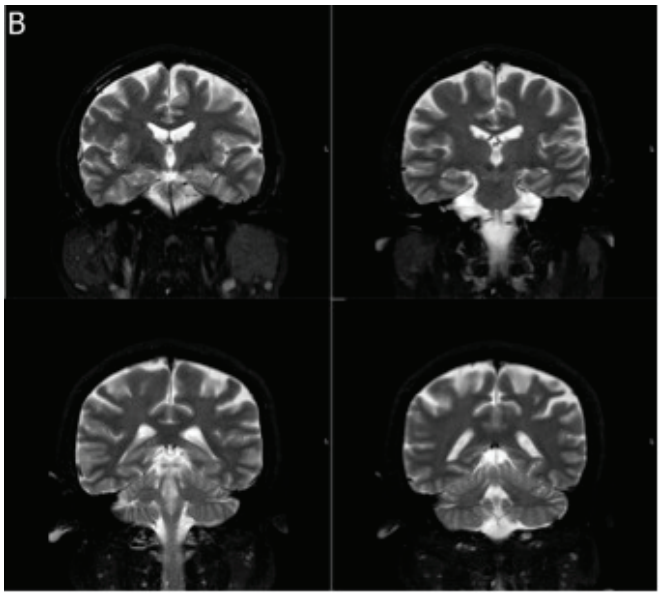


Figure 3: MRI shows left temporal atrophy

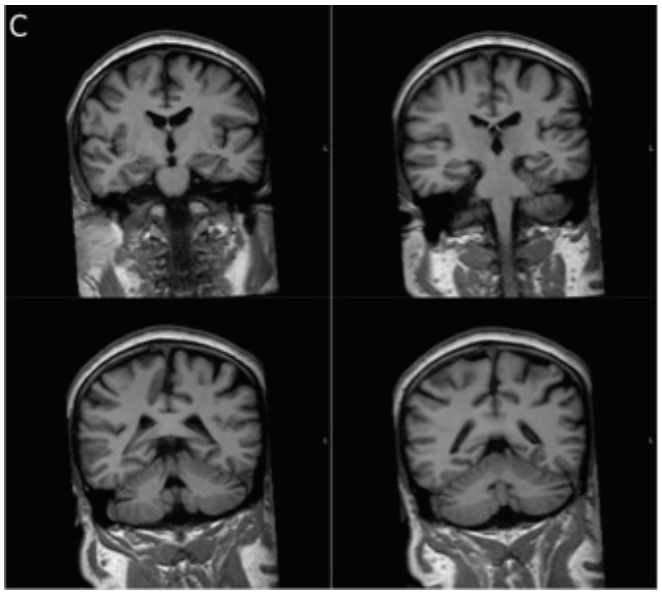
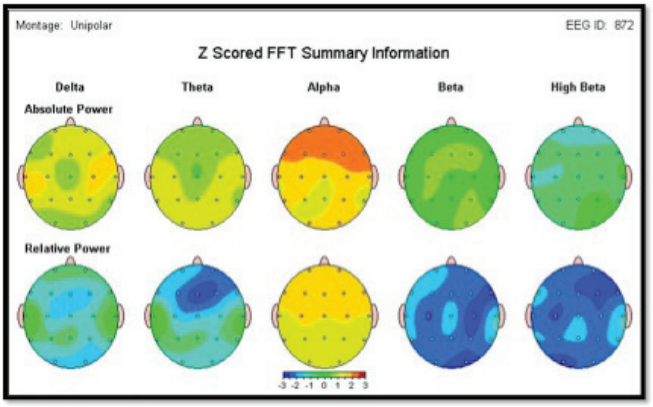


Figure 4: QEEG shows increase in theta-delta and alpha frequency



3. Discussion

For the first time in 1982, primary progressive aphasia (PPA) has been reported as a distinct clinical syndrome by Mesulam. In 6 cases, Mesulam has reported slow progressive aphasia symptom without comorbid dementia findings (Mesulam 1982). In subsequent years, clinical, imaging and postmortem properties of the syndrome have been defined. PPA is a neurodegenerative syndrome with progressive and isolated deterioration in language functions (Mesulam et al 1997). While mental functions such as attention, memory, visuospatial abilities, abstraction, judgment and behavior are maintained, an isolated and progressive deterioration in using and understanding words is seen at least two years long (Mesulam 2001). To date, a small number of PPA cases have been reported in the literature. Weak neurological findings in the right half of the body, asymmetric slowdown in electroencephalography, cortical atrophy, hypometabolism in the left hemisphere (especially in the frontotemporal and perisylvian regions) was found in most of these cases (Mesulam et al 1997). Two subtypes of PPA have been identified as non-fluent (PNFA) and fluent (Turner et al 1996). These have many common features

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with Pick disease and nonspecific lobar atrophies that cause focal degeneration in frontal and/or temporal lobes. In the light of this, it is believed that PPA and frontal lobe dementia can be addressed as a single clinical appearance (Mesulam 2001). PPA neuropathologically shows similar findings as nonspecific cortical degeneration with and without Alzheimer disease, Parkinson disease, Creutzfeldt-Jakob disease and spongiform changes (Karbe et al 1993, Mandell et al 1989).

PNFA is a slowly progressive degenerative disease characterized by atrophy in left hemisphere (particularly frontotemporal). It is one of three subtypes of frontotemporal lobar degeneration (frontotemporal dementia) (Neary et al., 2005). Unlike Alzheimer's disease it begins between 45-65 years of age and occurs equally in both sexes usually (Neary et al., 1998).

The youngest reported case was 21 years old (Snowden et al., 2004). Atrophy is seen in the left hemisphere more in temporal lobe on MRI. Approximately half of the cases have family history (Neary et al., 1998). In early it might confuse with depression and therefore diagnosis may be delayed. Brain MRI is important for verification of diagnosis.

Herein, we demostrated a case who early onset progressive nonfluent aphasia in this paper.

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