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Case report

Primary progressive aphasia: a case report on diagnostic issues

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Abstract

Primary progressive aphasia (PPA), a neurodegenerative condition of insidious onset which has language impairment as the most salient and significant initial feature may provide diagnostic challenges due to obstacles in confirming normal functioning in other cognitive domains. PPA is listed by the office of rare diseases (National Institute of Health) as a rare condition affecting less than 200,000 people in the entire United States population. The close similarities with the clinical signs of Alzheimer's disease and the behavioural variant of frontotemporal dementia in the later years, may also allow for a missed or false diagnosis if not recognised early enough. This case report describes a 63-year-old Caucasian female diagnosed with PPA and elaborates on the clinical presentation, the role of neuropsychology in arriving at a potential diagnosis and highlights a management approach.

Key words

neurodegenerative disorder, primary progressive aphasia, neuropsychological assessment, logopenic variant type

Introduction

With the increasing trend in life expectancy, it is no longer uncommon knowledge that neurodegenerative conditions are on the increase. The rarer sub-classes of dementias are now being seen more often in clinical practice, more disturbingly, in the younger age groups. The ability of clinical specialists to adequately recognise and diagnose these conditions remains imperative as this usually underpins the manner of support and therapy such patients receive.

According to the National Aphasia Association in the United States and seemingly defined by Mesulam,[1] Primary progressive aphasia (PPA) can be described as a syndrome spanning approximately 2 years, which comprises a progressive disorder of language function, with sparing of higher mental functions and general activities of daily living (ADL). Occasionally acknowledged as a diagnosis of exclusion in practice, PPA could be confirmed after assessment of clinical presentation, imaging, and linguistic testing.[2] This case report describes a 63-year-old White British female who was given a formal diagnosis of PPA after exclusion of other probable organic aetiological causes. Contribution of neuropsychological assessments to the diagnostic process is described.

Case history

A 63-year-old Caucasian female presented with a progressive cognitive decline, markedly in speech and language, of over 18 months duration, with notable difficulties in word finding, naming, spelling and comprehension. Some short-term memory deficits were also notable. There was no apparent change in general level of functioning as regards to the use of electronic devices, managing of finances, shopping, driving, maintaining personal hygiene and activities of daily living. There were also no changes in mood and no evidence of psychosis.

Two months after first presentation, patient was reviewed, and still presented with sustained language impairment with gradual progressive decline in word finding and comprehension. There were no changes to short term memory.

Assessments

Clinical assessment and investigation

Past medical history mentioned anxiety episodes in the previous year and benign breast cyst more than a decade back. Physical examination and haematological investigations were uneventful. She was on atorvastatin 20mg daily.

Results of MRI scan of head showed no evidence of ischaemia or haemorrhage. The midline structures appeared normal. There were involutional changes, although generalised and these appeared most marked around the left temporal lobe; no other abnormalities were noted.

Addenbrooke's Cognitive Examination III score was 67/100, which was below the cut-off score of 82. Difficulties were noted across several domains such as attention, memory, verbal fluency and language.

Neuropsychological assessment

Profile validity testing showed that subject was above cut-off on relevant scales. Intellectual functioning was within average range. Wechsler Adult Intelligence Scale (WAIS-IV) revealed a borderline IQ, with a score of 70. Wechsler Memory Scales, Forth Edition (WMS-IV) revealed that her deficits in memory were resultant from difficulties in the comprehension of language rather than secondary to memory impairment. Application of the Delis Kaplan Executive Function System (DKEFS), also further strengthened this observation. The test showed that executive function was intact, but seemed impaired due to significant word finding difficulties.

Assessment related to speech therapy also reported that the subject's problems arose from difficulty in language comprehension. In this regard, paucity in the use of verbs in subject's expressive speech as well as marked difficulties in word retrieval were noted, which were ultimately leading to a breakdown in communication.

Management

The patient was given a potential diagnosis of PPA. Though there is no known pharmacological agent with proven efficacy for PPA, the acetyl-cholinesterase inhibitor was tried on off-label medication basis. The patient was commenced on donepezil 5mg, which was increased after a month to 10mg. Patient continued to function at a high level, without any observable changes in personality or behaviour at the time, as is seen in most variants of PPA.

An extensive information pack from Alzheimer's research UK on understanding the condition and means of accessing help and support was offered to the patient.[4] A key worker was assigned and information on accessing PPA support groups was also provided. Speech and language therapist also remained involved. Duration of follow up and observations was for a period of around one year.

Discussion

Using a diagnostic criteria for PPA suggested by Mesulam,[1] we sought to further establish and confirm a potential diagnosis in our patient (table 1).

An article published by Gorno-Tempini,[3] elaborates on a proposed working classification of PPA and its variants. The article describes three possible variants: a nonfluent/agrammatic type, a semantic type and a logopenic type. The article further describes a potential diagnosis of PPA in terms of three aspects:

■ Clinical presentation based on specific speech and language characteristics

■ Supported by imaging, where there is known specific patterns notable on the scan

■ A definite pathology when there is available genetic data.

The progressive deterioration in speech abilities in the reported patient, coupled with the subsequent neuropsychological assessment and speech therapy reports suggested a possible logopenic variant type of PPA.

For purposes of research, further genetic analysis may have provided definitive pathological diagnosis and the significance. According to the literature, pathologic mutations in certain faulty genes (MAPT, Progranulin, C9ORF72) have been isolated.[5]

Conclusion

Based on the inclusion and exclusion criteria suggested by Mesulam, the reported case fulfilled the criteria deemed necessary for a potential diagnosis of PPA. This was also supported by generalized temporal lobe changes on the reported MRI scan. The neuropsychologial assessment findings were helpful in the diagnostic process.

This case report contributes to the literature base of one of the rare forms of dementia and may improve further awareness. Appropriate use of suggested criteria and neuropsychological assessments may prevent the risk of misdiagnosis and may help the patient in receiving appropriate supportive measures early.

Table 1: Characteristics in the reported case compared to the criteria adapted from Mesulam[1]

Criteria suggested by Mesulam[1]	Findings in the reported case
Inclusion criteria	
Language difficulty	Positive
Deficits in language should be the primary cause of any impaired activities of daily living	Positive
At onset aphasia should be the most prominent deficit.	Positive
Exclusion criteria	
Other medical disorders could better account for patterns of deficit	Negative
A psychiatric diagnosis could better account for cognitive disturbance	Negative
Prominent initial episodic, visual or visuospatial memory impairments	Negative
Marked prior behavioural disturbance	Negative

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