Review Article

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The Clinical Manifestation of Primary Progressive Aphasia

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ABSTRACT

This review mainly focuses on language disorders. One type of language disorder is known as aphasia. Aphasia is generally occurring due to secondary lesions that appear on subcortical white matter that injures the left hemisphere and hence causes dysfunction in linguistic information such as grammar, syntax, morphology, semantics and other aspects of language. Aphasia is considered an acquired language disorder as it blocks the language usage as a whole. Aphasia can be of primary and secondary. Primary aphasia deals with problems associated with language mechanisms whereas secondary aphasia deals with problems associated from memory, attention and perceptual impairment.

Primary progressive aphasia also abbreviated as PPA, is a neurodegenerative disorder that is closely associated with any form of cerebrovascular accident such as stroke, cerebral tumours and brain injury that affects the middle cerebral artery and its branches. Primary progressive aphasia also tends to form when there is family history and 2-3 siblings receive it. It is led to believe that people inherit PPA due to the mutation of a particular gene known as progranulin on chromosome 17q21.31. While progranulin gene is mutated, a hyperphosphorylated protein is deposited in brain known as tau in the form of intraneuronal neurofibrillary tangles or otherwise known as pick bodies.

Keywords

Language Disorder, Aphasia, Primary Progressive Aphasia, PPA, Clinical Manifestation, Neurodegenerative Disorder.

Introduction

Language is a system used for communication with the help of symbols attached with objects and concepts; that involves the use of neural substrate and amplifies multiple recurring themes that helps in connections and functions of multimodal association cortex [1]. As language is described as multimodal cortex, any impairments or language disorders are associated with the processing of linguistic information such as grammar, syntax, morphology, semantics or other aspects of language [1,2]. Speech and language disorders are considered secondary due to vascular lesions in subcortical white matter and basal ganglia [3]. An example of language impairment or disorder is known as aphasia [1,2].

Aphasia is an acquired language disorder in which it comprises of any impairment of language as a whole [4]. This includes the difficulty in producing or even comprehending written and spoken languages such as context of well-preserved naming, articulation, prosody and repetition skills [1,2,4]. Aphasia can be divided into primary and secondary aphasias where primary is due to problems associated with language processing mechanisms and secondary aphasia develops through other problems associated with memory, attention, perceptual impairments [2,4,5].

Aphasia can also be classified into different types such as fluent and non-fluent. Fluent aphasia is also known as receptive aphasia, which comprises of impairments that affects the reception of the language such as verbal, written comprehension or in the repetition of words, sentences that is incoming [1,2,6]. Examples of fluent aphasia are Wernicke's aphasia, Transcortical sensory aphasia, Conduction aphasia, anomic aphasia [1,6,7]. Non-fluent aphasia is

also known as expressive aphasia, which comprises of impairments that affects the articulation but more predominantly affecting the verbal and written comprehension [1,2,6,7]. Examples of non-fluent aphasia are Broca's aphasia, Transcortical motor aphasia, Global aphasia [1,6,7].

This review aims to illustrate the aetiology, pathogenesis, pathophysiology, clinical manifestations and treatment for primary progressive aphasia, a form of neurodegenerative disorder or dementia that alters and dysfunctions an individual's language capacity.

Primary Progressive Aphasia

Primary progressive aphasia is a neurodegenerative disorder associated with cerebrovascular accident such as stroke, head injuries, cerebral tumors involving the middle cerebral artery or one of its many branches [5,6,8-11]. Primary progressive aphasia is resulted from the dysfunction in the left cerebral hemisphere, most predominantly the frontotemporal region [5,6,8,9]. Left cerebral hemisphere controls 96-99% of language functions of right-handed people and 60% of left-handed people [10]. Even though dominance with other hemispheres can also cause aphasia; injuries to left hemisphere is found to obtain milder aphasias in left-handed people than right-handed people [10].

Recent studies show that Alzheimer disease and frontotemporal dementia are associated with primary progressive aphasia as they manifest language deficits [5,9,10,12,13]. It is also shown in research that about 20% of stroke patients develop aphasia [10]. In United States America about 700,000 strokes occur each year and about 170,000 cases are related to aphasia [10,14]. Research undertaken by [10] states that African Americans are more likely to be affected with aphasia as they have a 2-fold higher chance than any other race in obtaining a stroke [10,14].

In recent research, it shows that dementia is very similar to primary progressive aphasia as these both conditions dysfunctions language potentials of individuals [14]. The rate of mortality is presented low as primary progressive aphasia is considered as a condition and not a disease [10,14]. The research also stated that men are more likely to be affected than females as they are bilateral of language function [5,10,14]. Research also shows that; any age groups can develop this particular disorder regardless for male or female. Age plays a significant role in recovering from aphasia. An individual aged 70 or above is less likely to recover compared to younger ages [10,14].

The Latest research also describes that patients with family history of dementia also suffers from aphasia as it is closely related to language impairment [9]. This mainly occurs due to progranulin gene mutation, which appears on chromosome 17q21.31 [9,15] and it is also stated that 2-3 siblings in the family develops primary progressive aphasia [9,15]. Frontotemporal dementia that occurs to 40% of the population tend to form via inheritance, while 20% of these population reach progressive aphasias due to the progranulin gene that is mutated in the chromosome 17q21.31 [15]. While the mutation takes place, an aggregated hyperphosphorylated tau proteins gets deposited in the brain in the form of intraneuronal neurofibrillary tangles or pick bodies [11,15]. But 60% of cases from frontotemporal degenerative disorder do not show any pathological changes from tau protein that is deposited in the brain rather they show pathological changes in cerebral cortex and hippocampus due to ubiquitin immunoreactive neuronal cytoplasmic inclusions [11,15]. In some cases of frontotemporal lobar degeneration with autosomal dominant transmission disease, the ubiquitin immunoreactive neuronal cytoplasmic inclusions appear within the neuron around the same regions of cerebral cortex and hippocampus [11,15].

Primary progressive aphasia is only diagnosed when an individual portrays symptoms that characterises language impairment [9-11,14-16]. This type of aphasia tends to only progresses gradually and usually found in patients who are aged between 52 – 60 years old [11,16]. As the onset of this condition progresses, impairment of language comprehension develops and later on the stages the patient might even tend to become mute as this prolongs the patient might also move to the stage of death [9-11,14,16].

As doctors conduct diagnostic testing for patients suffering from aphasia, they need to know whether the patient got this condition for any brain haemorrhage or from any cerebrovascular accident such as strokes, seizures or sometimes even due to herpes simplex encephalitis [10]. They also check for acute and chronic headaches, which could be due to cerebral tumors as these conditions lead to degenerative brain disorders and hence could lead to primary progressive aphasia [10,14]. Doctors categorises the steps to language impairment such as, firstly patient would have to undergo frontotemporal dementia, which tends to change patient's behavior and personality [15]. Secondly semantic dementia is the leading cause that tends to lead to loss of conceptual knowledge [15] and thirdly leading to progressive aphasia which causes impairments to language expression and thus only portrayed in the later stages of the life [15].

Anatomical considerations must also be taken care of while diagnosing patients suffering from aphasia. Many parts of both hemispheres contribute to the production and comprehension of language, so it's very important to make notes in differences in individuals to avoid confusion of correlated structures with function [10]. Patients who tend to have slow growing tumors may possess mild form of disease that lets the lesions to grow slowly, allowing the tissues to recover from functional deficits [10]. The severity of the condition can be evaluated from congenital abnormalities, handedness, initial severity of the illness, time since the onset, aetiology, and nature of the underlying vascular lesion and finally the age of the patient [10] Patient suffering from left hemispherical injury at a younger age may not have any residual language impairment whereas the status of the contralateral hemisphere is also important in diagnosing and also in estimating the prognosis for recovery [10,11,15]. Doctors must take special precautions while diagnosing aphasia as there are number of diseases that can generate aphasia as it is considered as a condition

[10,11,15]. Aphasias are normally checked by neuropsychologists and speech or language therapists [10]. Medical machines such as functional magnetic resonance imaging is used in order to identify the types of aphasias and the areas where it's potential to be or which areas of the brain can primary progressive aphasia can affect [17]. Currently there is no set treatment for primary progressive aphasia. But treatments such as Boston Diagnostic Aphasia Examination, the Western Aphasia Battery, the Boston Naming test, the Token Test, the Action Naming test and Aachen Aphasia test are used in order to find the potential of patients' recovery stage [10,17]. For each of these tests, language must be tested individually in order to examine for spontaneous speech, naming repetition, comprehension, reading and writing [11,17].

Conclusion

The aim of this review was to illustrate the aetiology, pathogenesis, pathophysiology, clinical manifestations and treatment for primary progressive aphasia. Primary Progressive aphasia is neurodegenerative syndrome that affects the left hemisphere of the brain. This occurs because of any form of cerebrovascular accidents such as strokes, cerebral tumors, head injuries, which involves the middle cerebral artery and its branches. It can also affect the frontotemporal regions as well. Diseases such as Alzheimer's disease, frontotemporal lobar degeneration, semantic dementia can also result in primary progressive aphasia. This could also happen due to particular gene mutation known as progranulin at chromosome 17q21.31. While the gene mutation takes place, an aggregated hyperphosphorylated protein known as tau is deposited in the brain in the form of intraneuronal neurofibrillary tangles or pick bodies.

There are no set treatments available for this particular condition, whereas there are treatments available that used with the help of medical machines such as functional magnetic resonance imaging in order to find potentiality of recovering status in patients.

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