What is Primary Progressive Aphasia (PPA)?

"**Aphasia**" is a word used broadly for any acquired language impairment, when a person loses language abilities due to damage in certain areas of the brain. Most aphasias are caused by strokes or traumatic brain injuries, however PPA is caused by a disease in the brain.

"**Progressive**" refers to the symptoms of PPA worsening over time. People who have an aphasia caused by an acute injury such as a stroke may recover some of their communication abilities, whereas PPA unfortunately worsens over time. PPA tends to progress slowly, with changes being noticeable over the course of several months or years, rather than days or weeks. Many people with PPA can live independently or with minimal support for years after their symptoms begin.

"**Primary**" refers to the language symptoms being the primary characteristics of the syndrome, particularly in the first two years. People with PPA may eventually develop symptoms in other areas such as memory, personality, decision-making, and movement as the disease progresses.

What are the subtypes of PPA?

Nonfluent/Agrammatic: Individuals with the nonfluent or agrammatic subtype PPA most often have difficulty understanding and using complex grammar. The person may use short, simple sentences, mix up word order, or leave out small grammatical words (e.g. "the," "is," "for," etc). Many individuals with this diagnosis also have motor speech impairment. Speech may be slow, effortful, and articulation abilities often decline over time.

Semantic: Individuals with the semantic subtype of PPA often have the most difficulty knowing the meaning of specific words and retrieving the names for actions and objects. Their speech is usually fluent and clear; however, it may be "empty" due to using vague words (e.g. "We went to that place with this person who did everything for us.").

Logopenic: Individuals with the logopenic subtype of PPA often have the most difficulty retrieving specific words for ideas they would like to communicate. Therefore, they may use pauses and fillers (e.g. "uh," "um," etc) while searching for words. Sometimes individuals with this subtype produce phonemic paraphasias, and have difficulty repeating phrases and sentences on testing due to an impairment in auditory working memory.

Not everyone with a diagnosis of PPA clearly fits into a single subtype. A person may have clinical symptoms of more than one subtype. Fitting into a subtype can help health professionals hypothesize about the underlying pathology or what to expect in the future, but it's not necessary for diagnosis.

Are there treatments for PPA?

There are currently no medications or other treatments that have been proven to cure PPA specifically, although research is ongoing. There are some medications that can boost thinking skills or treat symptoms such as anxiety or depression, however they will not cure or stop the progression of the disease.

Many people with PPA and their families benefit from speech-language therapy, psychological counseling, support groups, adult day health programs, and/or other support resources. Although these will not directly affect the course of the disease, they can help patients and caregivers develop strategies to work around communication impairments and give them tools to cope with the diagnosis.

Please refer to the PowerPoint presentation for further information regarding treatment.

Is PPA genetic?

Most cases of PPA do not run in families, but there are rare cases that are genetic due to a mutation in a single gene. If one parent has this gene, each child has a 50% chance of also having the gene. Typically, a person with a genetic form of PPA has several family members who developed PPA, Frontotemporal Degeneration (FTD), or Amyotrophic Lateral Sclerosis (ALS) at a relatively early age (50s or 60s).

How is PPA related to Frontotemporal Degeneration (FTD) and Alzheimer's Disease (AD)?

When a neurologist diagnoses a condition such as PPA, there are generally two "levels" to the diagnosis- the *clinical syndrome*, which is based on the symptoms the person is experiencing, and the *underlying disease*, which is the physical process in the brain causing the symptoms. PPA is a clinical syndrome because it is diagnosed based on clinical symptoms. It is most often caused by one of two underlying diseases: Frontotemporal Lobar Degeneration/Frontotemporal Degeneration (FTLD/FTD) or Alzheimer's Disease (AD). These diseases are caused by different proteins in the brain malfunctioning and damaging the brain tissue around them.

Often it is alarming for a person with PPA to hear that he/she has an atypical form of Alzheimer's Disease. However, it does not mean that the person will develop Alzheimer's *Dementia*, which is a clinical syndrome where the primary symptoms are memory related. It means that the person has the same underlying disease in his/her brain, but the disease is causing different symptoms depending on where the proteins are located. Both AD and FTD proteins can occur in various brain areas, causing different clinical syndromes.

Neurologists try to determine which underlying disease a person has by looking at biomarkers. Biomarkers are proteins found in spinal fluid or through special kinds of PET scans. These tests are important because new medications are being developed to target specific proteins.

What are the purposes of MRI and PET scans?

MRI scans show the structure of the brain, which allows neurologists to rule out other causes of impairment such as strokes or tumors, and to look for shrinkage in the language areas of the brain as expected in PPA. Finding the locations of the shrinkage may help the neurologist explain why a person is experiencing particular symptoms.

FDG-PET or glucose PET scans show the functioning or amount of activity in the brain, helping a neurologist better understand which areas of the brain are not working properly. Often areas that are not working functioning correctly will be visible on a glucose PET before there is atrophy visible on a MRI scan. However, they can be expensive and many insurance companies do not cover them. Amyloid PET scans show the presence of the protein amyloid beta in the brain. This protein is associated with AD and can help determine if a person's PPA is caused by AD pathology. Tau PET scans are a new kind of scan, currently being researched. We hope that eventually tau PET scans will accurately show the presence of the protein tau in the brain, which is associated with both AD and FTLD/FTD. When a person's brain has tau but not amyloid, the underlying disease is likely FTLD/FTD.

References

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