



A Very Rare Medical Case Report of Primary Progressive Aphasia (P.P.A)

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Abstract

A 68-year-old man, who has been suffering from a stuttering problem for three years. Before he encountered these problems, he was a social person and a professional journalist and political analyst who could study global issues and discuss them with his clients. He was familiar with three major languages of the world and had a university education. I had known him for more than thirty years and had a relative knowledge of almost all of his illnesses.

He was a very social person and an athlete, etc. That is why when one of his close friends told me about his medical condition, I visited him as soon as possible. Because it was unthinkable for me that this person, with these characteristics, would have a problem called stuttering.

To the extent that those around him would be worried. And when I did his first consultation, I believed with disbelief that he had aphasia.

What is aphasia?

Aphasia is a language and communication problem.

What is the problem? The person has the words in mind but can't find them, or can't form sentences correctly, or doesn't understand what others are saying.

Example: A person with aphasia wants to say "water," but may say "that thing... that we eat" or say a meaningless word. He knows what he wants, but he is at a loss for the "words".

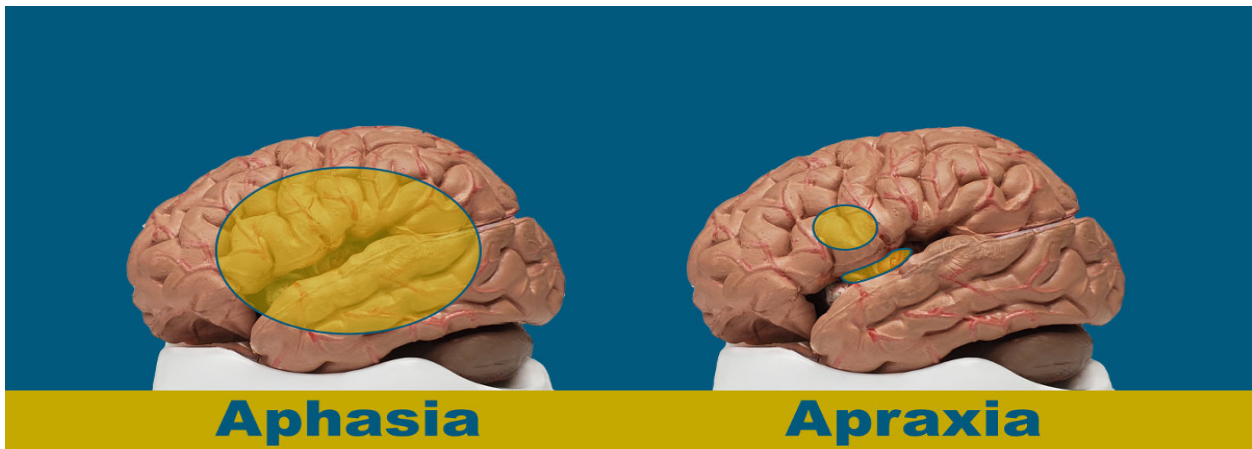
Causes: Damage to the language centers in the brain (usually the left side of the brain).

What is apraxia?

Apraxia is a problem related to motor planning in apraxia, the brain knows what it wants to do and the muscles are healthy and strong, but the brain cannot send the command "how to do" to the muscles. The connection between thought and action is broken. They will automatically wave their hand and say goodbye without thinking.

Causes: Damage to parts of the brain that store movement maps it can be difficult to distinguish between the two. At least when first encountered, the two disorders often occur together, for example, after a stroke, so it is sometimes difficult to tell the difference.

A person may simultaneously be unable to find the right word (aphasia) and unable to move their lips and tongue to say the right word apraxia of speech.



Quick comparison table

Apraxia	Aphasia	Feature
Moving and doing work	Language and speech	Main area
Making voluntary movements such as (brushing your teeth or waving your hands)	,Word finding comprehension, reading and writing	Problem in
Muscles are healthy (not paralyzed)	The jaw and tongue muscles are healthy.	Muscle condition
Movement error(making a wrong or (clumsy movement)	Language error (saying the (wrong word	Error type

Off course the symptoms of aphasia depend on which type a person has.

Broca aphasia is sometimes called expressive aphasia. People with this type of aphasia may eliminate the words "and" and "the" from their language, for example. They may speak in short, but meaningful, sentences. They usually can understand some speech of others. People with Broca aphasia often have right-sided weakness or paralysis of the arm and leg.

Wernicke aphasia is sometimes called receptive aphasia. People with this type of aphasia may speak in long confusing sentences, add unnecessary words, or create new words. They usually have trouble understanding the speech of others.

People with **global aphasia** have trouble with speaking or understanding language.

Organic Causes:

Aphasia and apraxia refer to injuries or diseases that affect the structure of the brain and cause dysfunction of the language Centers and motor planning.

Since both disorders result from acquired brain damage, they share common organic causes.

Centers and motor planning

Since both disorders result from acquired brain damage, they share common organic causes.

1. Stroke (Stroke/CVA):

The most common and main cause of aphasia and apraxia is stroke.

Ischemic Stroke: A blockage of blood vessels, cutting off blood flow to part of the brain. If the blockage occurs in the left middle cerebral artery (MCA), it can involve both Broca's and Wernicke's areas and adjacent motor pathways, resulting in both aphasia and apraxia.

Hemorrhagic Stroke: Bleeding into brain tissue, which causes tissue destruction

2. Traumatic Brain Injury (TBI):

- Severe head trauma, especially in accidents, can lead to focal or diffuse damage to the cerebral cortex.
- If damage occurs in the left frontal and parietal areas (location of language and motor planning centers) aphasia and apraxia) develop.

3. Brain Tumors:

Tumors that grow near or directly in the areas of language and movement can compress or destroy surrounding tissues, causing symptoms of aphasia and apraxia.

Gliomas and meningioma are among the tumors that can cause these symptoms.

4. Neurodegenerative Diseases:

In these conditions, the disorders progress gradually over time:

- Primary Progressive Aphasia (PPA) type of dementia in which language problems are the only prominent early sign or symptom. This condition can also lead to apraxia of speech over time.
- Advanced Alzheimer's and Parkinson's disease: In advanced stages, they can cause speech and movement disorders by causing extensive lesions or disrupting neural pathways.

5. Other causes:

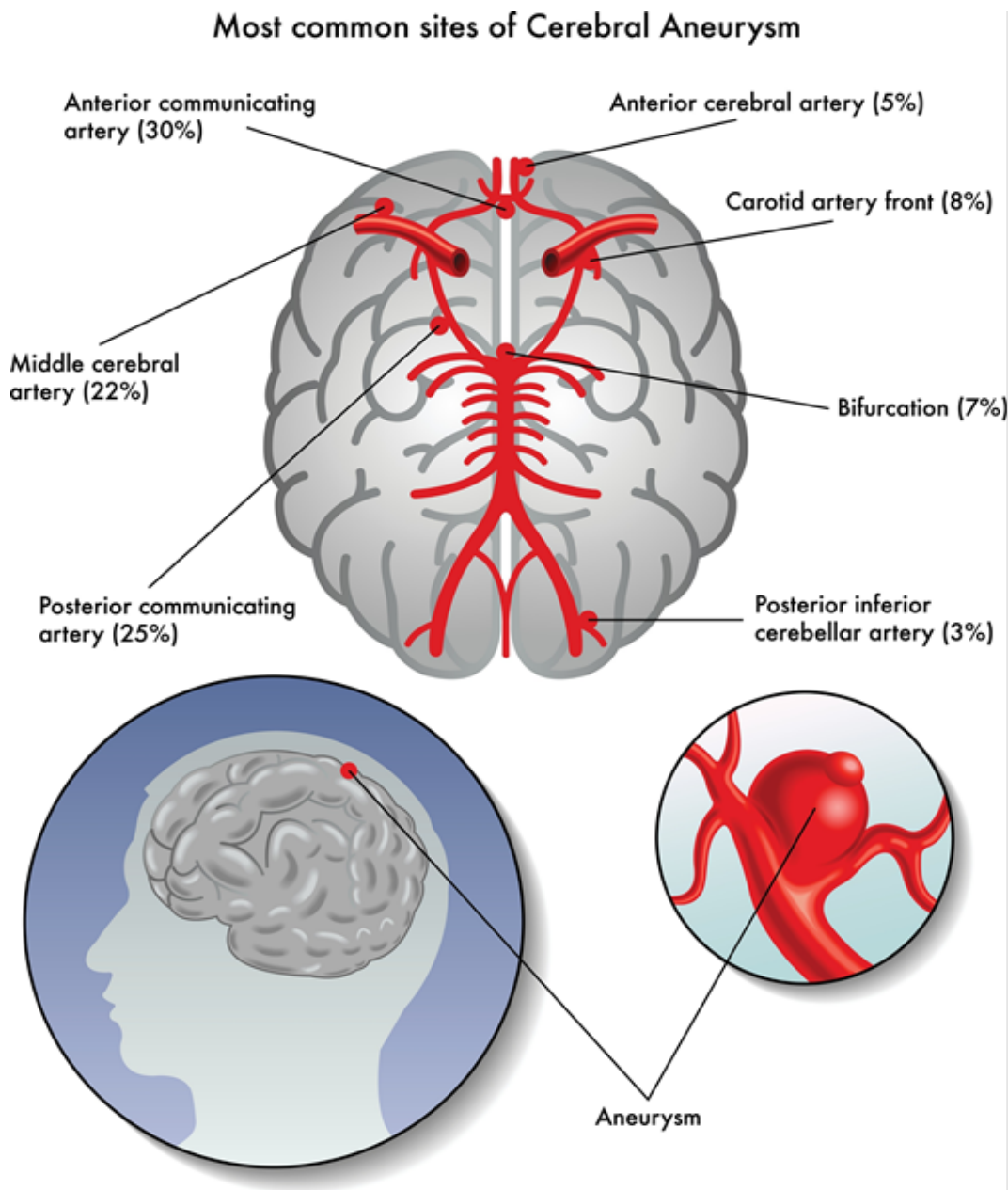
- Brain infections: Brain abscess, Meningitis, or Encephalitis, which cause inflammation and destruction of nerve tissues.
- Poisoning: In rare cases, certain poisonings or severe vitamin deficiencies such as vitamin B1 deficiency can temporarily or permanently affect brain function.

Because our case had none of these causes, our work was more difficult, both in diagnosing and controlling it.

The patient did not show any symptoms or complaints of these types of diseases, and our paraclinical examinations did not affect any of them.

In the initial history we had of the patient, we did not suspect any of these causes.

A CT scan of the brain and CT angiography were performed to discover and identify the causes of vascular insufficiency or congenital insufficiency such as cerebral aneurysm or asymptomatic ruptures of cerebral vessels, and then a brain MRI and various blood tests were reported to be completely normal.



When all our paraclinical investigations failed to find the cause of this problem in our patient, some of my colleagues finally recommended that we refer him to a psychologist to examine the problem.

Although at first, given my previous knowledge of the patient, I could not believe that the patient's problem was psychological. Nevertheless, I introduced him to my colleague, Dr. Eugene Sotiri, a psychologist, and I was actively present in all his consultations. Dr. Sotiri put the patient on various antidepressant treatments, including Citalopram.

But six months later, the patient told me that my problem was not psychological. Even Dr. Sotiri himself found it unbelievable that the case was caused by psychological factors, and he said that you should look into this issue in the neurology department and your own specialty to find the cause.

I wanted to get a PET scan of his brain here, maybe it would help us with the diagnosis.

When there are symptoms of chronic focal aphasia or apraxia, but structural imaging (MRI) and memory and dementia assessments are completely normal, this situation strongly rules out the most common causes (such as stroke, tumor, or typical Alzheimer's) and our focus should shift to focal neurodegenerative causes that manifest in the early stages without obvious lesions on MRI .

But unfortunately, in our country, to perform a PET scan of the brain , a special substance must be used that highlights the brain images, which this substance has not entered this country, at least so far, and there is no prospect of obtaining it at the moment.

For this reason, we also faced this real diagnostic impasse.

In consultation with our fellow doctors here, we all agreed that the case should be monitored as primary progressive aphasia. In this regard, Professor Ds.Iliriana Jaki emphasized this in her last consultation.

- Gender & Race: No significant predilection for any specific race or gender has been established, though some studies suggest a slight male predominance in some tauopathy variants.
- Prognosis: The condition is progressive. In cases of PPAOS, the deficit may remain isolated for years before spreading to other motor systems (like gait or limb movement).

Main hypothesis:

Primary progressive aphasia (PPA) is the main focus of diagnosis in these conditions, primary progressive aphasia (PPA) or other early forms of frontal and temporal lobe deterioration (FTD).

1.What is PPA?

It is a neurodegenerative disease in which language or speech problems (verbal aphasia or apraxia) are the only or main symptom at the onset of the disease, and memory and other cognitive functions are relatively preserved for years to come. As with our patient, this memory problem is quite true. The patient had a MOCA = 27, which strongly ruled out memory impairment.

Why is MRI normal? In the early stages of PPA, atrophy is very subtle or microscopic and may not be clearly seen in standard

2. Subsequent diagnostic steps:

Subsequent diagnostic stages of functional and biological assessments should be used to confirm or rule out PPA and other rare causes, in which structural MRI is normal.

A. Functional imaging,

which can focus on the following areas:

- PET scan (FDG-PET scan)

This test measures glucose metabolism (brain cell activity) instead of structure. In PPA, even if MRI is normal, a PET scan can show hypometabolism (decreased activity) precisely in the area responsible for the patient's speech (for example, the broca area of the irritated PPA).

- SPECT scans are similar to PET scans, but measure cerebral blood flow. Although PET is more accurate, SPECT can also show hypo perfusion in the affected areas.
- FMRI

Measures brain activity when performing a specific task (such as naming objects) and can show abnormal patterns of function.

B. Laboratory and Biological Evaluation

- Panels Autoimmunity / Vasculitis:

Although vascular symptoms have been ruled out, a thorough assessment is essential to rule out rare autoimmune diseases.

Or vasculitis (such as autoimmune encephalitis) that can cause focal symptoms and have a normal initial MRI.

- Cerebrospinal fluid (CSF) analysis:

To rule out rare infectious/inflammatory causes (such as prion disease or encephalitis) as well as to search for biomarkers of neurodegeneration (such as tau or amyloid proteins) that can predict the type of FTD / PPA pathology...

- Genetic testing:

If the patient is young (under 65) and has a family history, testing for genes such as C9ORF72 or the progranulin gene (GRN) can help determine the cause.

Temporary follow up:

Clinical follow-up:

The most important step is to repeat the clinical trials to determine if BAN / apraxia is really progressing. The progression of the disease strongly supports the PPA hypothesis.

MRI recurrence:

High quality MRI in 6 to 12 months can detect any mild, localized atrophy that was not detected in the initial scan.

Of course, due to the general condition of this patient, the process of himself and his family, I preferred not to do a Lumber puncture test for him. Because he did not prove a problem in this patient. My colleagues also confirmed this.

Treatment results:

Given that the cause of aphasia and apraxia is still unknown. And we do not have more diagnostic options available to diagnose underlying diseases at this time.

The treatments recommended by our colleagues were mainly dietary supplements for the nerves and sometimes treatment with antidepressants, none of which had the slightest effect on reducing the patient's problems.

At another point, the patient himself went to the Neuro20 clinic on the advice of his friends and was monitored for about three months by wearing sensor vests to stimulate and recover the disabled or weak nerves. Hemorrhagic strokes, etc. are used, which are in the experimental stage, and I can still say in our country that it has not been imported. And patients who are monitored in a private clinic provide contradictory results, which, of course, as proven by experience on this patient, use these sensor vests for three months and three days a week for one hour each day do not solve the problem.

His condition has been worse.

But the treatment of Speech Therapy was very early in the form of repetition of words and sentences in the patient's own language, which was relatively better for his development. Listening to the sound of words and changing the language and mouth, for a similar dialect, similar to what is done to get children started.

Although this method has not completely solved his problem, it seems that even as a treatment, it is a symptom that can be hoped for, which of course takes time.

Conclusion

Aphasia or apraxia in adults who do not have a history of chronic diseases or trauma and develops gradually poses many challenges to the medical staff, which is very, very different from children. In children, the state of growth and development of nerve cells, which should be sought in genetic roots, may lead to separate discussions, and so far, due to its frequency in communities, it has made good progress. But when it comes to adults, without trauma or a history of chronic illness, the task becomes more complicated.

Specialized facilities for final diagnostics make the task more difficult due to the lack of diagnostic facilities. We also look at the subject from a psychiatric point of view, but this is not the whole story we have encountered for the first time.

In patients who do not have a history of trauma or cancer or stroke, we should think about the problem of primary and progressive aphasia and monitor the patient from different perspectives by repeating Para clinic tests.

We must wait for the development of the patient's clinical symptoms and specialized facilities for more accurate diagnoses.

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