A physician's story of his own illness: Aphasia from possible stroke but more likely from encephalitis

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KEYWORDS Aphasia; cerebral infarction; encephalitis; physician patient

PREFACE (BY DR. STUART)

As physicians, we become accustomed to the notion that health turns on a dime, sometimes predictably but often without any apparent warning or reason. What follows is a case description written in the first person by a physician patient who has enjoyed a long and distinguished career in internal medicine. A Hopkins graduate, he practiced primary care and pulmonary medicine at my institution for 40 years before retiring into a part-time practice with our specialty clinic in multiple sclerosis. An avid reader and writer, he has been an amateur historian of medical and southern history and has been an active member of the Osler Society. His acute illness, detailed below, was striking in its remarkable behavioral presentation, and the climb back has been slow and painstaking. This story, as he tells it in his own words, serves as an important reminder of the uniqueness of individual experience in medical illness of all varieties, not just this one. I have included, for the record, my clinical recollection along with the necessary laboratory data for your review. But the more interesting read, I think, is in Dr. Lathan's words on the matter. When all is said and done, as the title alludes, we have never been entirely certain whether this was a case of herpetic encephalitis or cerebral infarction. As we can all appreciate, sometimes the data let us down and we must proceed on clinical intuition. That said, we elected to treat him for both, and his progress to recovery has been very gratifying. That he can even conceive of submitting this report to the journal, given his striking cerebral pathology seen on imaging, is a stunning testimony to his perseverance and hard work in years of recovery.

PRESENT HISTORY (BY DR. LATHAN)

In late September 2012, I finished a 10-day trip to Italy and flew home from Venice to Atlanta through New York City. I had a mild sore throat with congestion and a temperature of 99.4°F but improved after 2 days of good sleep (10 hours). The next morning I seemed okay and watched several hours of television footage of the Ryder Cup Golf

Tournament. In the afternoon, I started trying to read the newspaper but could not comprehend the words, because it appeared that there were no gaps between the words. There was actually no visual blurring but for several hours I could not read the paper, which frustrated me greatly. I became agitated and my wife could see that disaster was imminent. Shortly, an ambulance arrived and it took 4 attendants to force me into the vehicle. At the Piedmont Hospital emergency room, I had progressive agitation and immobilization requiring sedation.

My family requested a consultation from my neurologists, Dr. Doug Stuart and his father, Dr. Bill Stuart, who had worked with me in the Multiple Sclerosis Center of Atlanta. I was sedated and had no memory of what occurred for the next 60 hours but was later told that I was in intensive care under the care of Drs. Stuart.

CASE REPORT (BY DR. STUART)

The patient at the time of presentation on September 28, 2012, was a 74-year-old man brought in by ambulance for agitation and confusion. He and his wife had just returned from a trip to Italy 3 days prior and on the day before admission he complained of some nonspecific malaise with congestion. Over the course of the day watching sports, he was observed by his wife to have progressive agitation and delirium. He became combative when emergency personnel arrived and had to be compelled into the ambulance. In the emergency room, a head computed tomography scan showed an area of lucency in the left temporoparietal white matter but no hemorrhage. He became increasingly restless and combative, with cursing and loss of social decorum and modesty, completely uncharacteristic of him. Sedation with 7 mg of lorazepam and 15 mg of haloperidol was required to bring his behavior to a manageable condition.

His prior health was significant for severe osteoarthritic joint disease and spinal stenosis with a long history as an ultramarathon runner. He had had a hip fracture and knee replacements and several cervical spine decompression/fusions. He had a history of supraventricular tachycardia. There was no history of

Figure 1. Magnetic resonance images from (a) September 29, 2012, and (b) June 3, 2013.

tobacco or alcohol abuse or illicit/prescription drug abuse. He is married and is a semiretired internist/pulmonologist. He has no allergies. Medications at the time of admission included statins and naproxen.

On examination, his general exam showed a normal temperature, with a blood pressure of 120/70 mm Hg, a pulse rate of 70 beats/minute, and respirations of 18 breaths/minute. His neck was supple with a normal range considering his multiple neck surgeries. Examination of the head, eyes, ears, nose, and throat was unremarkable, and his lungs were clear bilaterally. Cardiac examination was normal, and the abdomen was soft without guarding and with normal bowel sounds. No unusual skin rash was observed. Neurologically, he was initially agitated and combative but was able to answer simple questions correctly. Attention was distractible and he was disoriented. Language was fluent but confused. Cranial nerve exam was otherwise normal, as was motor and sensory testing within the limits of his cooperation. No obvious ataxia was seen and reflexes were normal.

In the hospital, a lumbar puncture demonstrated no white blood cells or red blood cells. Protein was 66 mg/dL, and glucose was 70 mg/dL. Cytology was normal and polymerase chain reaction testing for herpes simplex virus and varicella were both in normal range. Tests for syphilis, West Nile virus, arbovirus, and cryptococcal antigen were normal. The complete blood count and complete metabolic panel were normal, and erythrocyte sedimentation rate, troponin, thyroid-stimulating hormone, and ethanol and drug screens were unremarkable. Lactate and arterial blood gas were normal. A test for tuberculosis was normal and urine testing for *Legionella* was negative. The antinuclear antibody was negative, as was the antineutrophil cytoplasmic antibody. Rheumatoid factor was absent. Factor V Leiden genotyping was heterozygous.

Magnetic resonance imaging (MRI) performed on September 29, 2012 (*Figure 1a*), demonstrated a prominent area of left posterior temporal and insular increased signal on fluid-attenuated inversion recovery and T2 sequences without mass or

enhancement. There was mild sulcal effacement and a trace area of “bloom artifact” in the left insula felt to represent trace hemorrhagic change. When MRI was repeated, the area of increased signal had expanded medially and posteriorly but still showed no enhancement (*Figure 1b*). A chronic nonspecific arteriosclerotic signal was seen elsewhere in the bifrontal white matter.

An electroencephalogram performed on October 2, 2012, demonstrated bilateral theta slowing somewhat more prominent in the left temporal lobe, but no sharp waves or periodic complexes were seen. Echocardiogram was unremarkable. Because of the past history of supraventricular tachycardia, he was started on flecainide while in the hospital, as well as levetiracetam for seizure prophylaxis.

In the hospital, over the first several days, his behavior became progressively less agitated, but his language was noted to be obviously more aphasic with fluent jargon that was typical of a Wernicke’s aphasia. Behavior was impulsive and he required continuous bedside support. Aspirin and clopidogrel were added for the possibility that his presentation was due to infarction, but he completed a course of intravenous acyclovir for the clinical diagnosis of herpes simplex encephalitis. On the 10th hospital day, he was discharged to a rehabilitation facility for inpatient therapy.

FOLLOWING 2 DAYS LATER (BY DR. LATHAN)

On Monday morning, I woke up after 60 hours of sedation, and several family and friends surrounded the bed. I was able to speak but could not remember the names of anyone. I recognized my wife, Millie, but could not recall her name or any family or friends—no one!

I looked at the television and asked about the golf tournament and the football results. I was told, “That happened yesterday,” which did not make sense to me.

Later I was moved to a hospital room, and one morning I was visited by a good friend, Dr. John Cantwell. I tried but could not recall his name—however, I did know him and was

able to tell him that he was from Wisconsin. I was especially frustrated that I could not articulate the name of the golf course my wife and I had played a short time before.

About a week later, I moved again to a rehab center and was sent to speech therapy. To me, this was like starting over in first grade or kindergarten. I could recognize verbally only 10% of animals and other objects shown to me on flash cards. Slowly, after several days later and more therapy, I could recall my wife's name almost 100% of the time and my children's names about 70% of the time.

On one occasion, a nurse came by on a Saturday to watch football on television with me and my son, Rob, in the hospital room. Louisiana State University and another team were playing, but I could not recall either of the teams' names. I could say *Baton Rouge* but could not say the state's name. Another friend came to visit and asked me to list all of the teams in the Southeastern Conference (something I would normally be able to do easily); however, out of 14 teams, I could only name Alabama and Georgia.

Back home again, finally, 2½ weeks later, I was given a quiz in my kitchen by the speech therapist. I scored 0%, not able to recall names like *knives*, *spoon*, and *fork*. I was especially frustrated that I could not later say the word *microwave*. Therefore, it was very useful for me to begin weekly home therapy from the speech teacher and deliver "speeches" of diaries and especially newspaper articles on football games from the sports section. My graduation occurred on New Year's Eve, when I was able to make a fine speech about Georgia Tech beating Southern Cal and Clemson defeating LSU in the football bowl games.

After 3 months, I was certainly improving (the brain was "losing its charm"). I had a follow-up appointment with Dr. Doug Stuart, who felt that I was much better. The MRI result was almost back to normal (*Figure 1b*). At first, the diagnosis suggested the possibility of a stroke but later was that of encephalitis, which Dr. Stuart felt was more likely.

I was definitely better at word retrieval but still had difficulty with saying the word *encephalitis*. I would try and say this word, resting in bed, half-asleep at night. Often at night, I would also try to recall the names of friends. And when I could not remember the names, I would resolve to call them in the morning.

WHY DAD LIKED CEZANNE

(Essay written by my daughter, Caroline Lathan Stiefel, an artist, in November 2012)

Today I finally learned why my dad has always like Cezanne. Whenever he visited an art museum, my dad wanted to know whether they had any paintings by Cezanne, whom he considered a wonderful artist. I was somewhat curious how my dad, who was rather unschooled in the realm of art, had come to this conclusion.

About 7 weeks ago, my dad had a sudden episode that affected the language and speech area of his brain. I was seeing him in the hospital in Atlanta and he was in good spirits but mainly had trouble with speech, calling every object rice and every place South

Carolina. Upon returning home to Pennsylvania, I have called him regularly. Our conversations have been longer than usual and his first sentences usually sound normal, but then "incorrect" words and phrases slip in, especially as he attempts to describe a particular event or memory.

This morning, out of the blue, he started telling me about the first and only art class he took in eighth grade in Chester, South Carolina. His teacher mainly taught ceramics (ceramics took a long while to get out of him, with my guessing various occupations until I got it right). The teacher, Mr. Rash (his son is now a great writer—Ron Rash), said that his first assignment was to draw a still life. Getting still life out of Dad took quite a while. He first said, "We are making, you know, those things like copying glass, fruit, you know, a bowl." And finally, I said, "A still life?" And he said, "Yes!"

My dad said there was one boy in the class whose drawing was by far the best. Mr. Rash came over to him and said, "This is very good. It looks almost like a 'Collette.'" I said, "Collette?" And Dad said, you know that artist that starts with a "C." I immediately thought of his favorite artist, "You mean Cezanne." "Yes," he said, "Cezanne!"

My dad asked Mr. Rash who Cezanne was, and Mr. Rash responded, "You do not know Cezanne? Well, you cannot come back to class until you have looked up his work." So, my dad went to the Chester Library and studied the books and encyclopedia on Cezanne and from that point on, Cezanne became his favorite artist.

EPILOGUE (BY DR. LATHAN)

I still frequently say *days* instead of *hours* and *husband* in the place of *father*. It helps to go through the alphabet, starting with "A, B, C . . ." to try to remember words. Also, I will pause on a name, but a friend or relative will be able to provide it to me or give me a clue.

Dr. Stuart, after his 3-month follow-up, recommended that I should retire working from the Multiple Sclerosis Center. He also recommended that I not drive anymore. All of my medications were reviewed.

Five years later, I have really enjoyed retirement. In general I feel well and work on improving my speech and recall but still struggle. Often I wake up in the middle of the night to recall someone's name. I have not played golf in 2 years but enjoy swimming 3 to 4 times a week, finding water aerobics very helpful. I sleep very well (9–10 hours a night) and take a nap every afternoon. I have a good diet but still could lose 15 lb. I read every day and have published three books, one in the last 2 years, and I have been working on two other books. I am not bored or depressed at all.

ACKNOWLEDGMENTS

I am indebted to my daughters—to Caroline for ideation and the essay and to Stewart for her astounding typing and research—and to my son, Rob, for his help on football. I also thank Dr. Doug Stuart for saving me from the intensive care and my wife, Millie, for her tenacity and love and for calling relatives and friends.