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The Official Journal of the American Academy of Neurology



Neurology Publish Ahead of Print
DOI: 10.1212/WNL.000000000201698

Right Brain: The Purple Scarf and the Cortical Ribbon

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Neurology® Published Ahead of Print articles have been peer reviewed and accepted for publication. This manuscript will be published in its final form after copyediting, page composition, and review of proofs. Errors that could affect the content may be corrected during these processes.

Equal Author Contribution:

Contributions:

Katherine Fu: Drafting/revision of the manuscript for content, including medical writing for content

Lealani Mae Acosta: Drafting/revision of the manuscript for content, including medical writing for content

Figure Count:

0

Table Count:

0

Search Terms:

[25] All Cognitive Disorders/Dementia, Creutzfeldt Jakob disease (CJD), Humanities, Narrative medicine

Acknowledgment:

We would like to acknowledge and sincerely thank the patient's family for their review of and consent to share this piece.

Study Funding:

The authors report no targeted funding

Disclosures:

K.A. Fu is serves as Deputy Editor of the Neurology® Resident & Fellow section; L.M. Acosta is the editor for the Humanities section of Neurology®

Preprint DOI:

Received Date:

2022-06-21

Accepted Date:

2022-11-01

Handling Editor Statement:

Submitted and externally peer reviewed. The handling editor was Whitley Aamodt, MD, MPH.

Abstract: A patient with probable Creutzfeldt Jakob disease (CJD) who was previously a high-functioning financial analyst remains fiercely independent, despite her family's concerns about her safety. As a trainee with limited experience in leading these difficult conversations, this neurology resident describes her experience reconciling the viewpoints of herself, the patient and her family. She reflects on the ability to reframe this delicate conversation from one of loss of independence in a patient with a rapidly progressive, neurodegenerative condition to one of safety, security and the care of her family.

She had a purple hat with a short brim, made of felt-like material. It sat quietly on her bed, along with the deep purple scarf that accompanied it. As the inpatient general neurology team, we were meeting our patient in her early 50s for the first time on rounds. Our ersatz livery of polyester white coats, blue nylon jackets and thin sea green scrubs surrounded her blue hospital gown peppered with small flowers. The natural sunlight from the window mixed with the fluorescent lighting of the hospital room, illuminating her two sisters sitting on emerald vinyl seats against the window. As the junior resident, I stood between the bed and her sisters. Although this was a direct admission from the neurobehavior clinic, we still asked the patient and her family to re-tell key pieces of her story.

She worked as a financial analyst in a small town. She was always the planner and organizer, the one her family relied on to take the initiative to coordinate family gatherings or her parents' medical appointments. Her sisters noticed that over the previous six months she seemed slower -- her responses, her walking, her work. She made unusual mistakes: mistaking the refrigerator for the freezer or not recognizing her keys, thinking they were the TV remote control. She rarely asked her sisters or parents, both of whom lived nearby in their small town, for favors or for help. However, she had stopped jotting down notes on important tasks, like reminders to buy certain foods before a family event or notes to herself when planning the work calendar for her month.

She appeared at her parents' house one evening, saying her car windows no longer worked. When they asked her to show them, she tried to slide the window down from the outside with just her finger, as if it was the touch screen of her phone. Shortly afterward, her family took her to a neurologist. The MRI of the brain was concerning enough that he referred her to a cognitive specialist for a second opinion. This specialist immediately admitted her to the hospital for an expedited work-up, to obtain all the tests in a few days that may otherwise take weeks to coordinate as an outpatient.

Our inpatient neurology team reviewed the outside MRI and compared it with the one from this hospitalization. In both sets of images, the edges of her brain on the diffusion weighted sequence lit up like the silver lining of the sun peeking through beyond the clouds: "cortical ribboning." It is ironic that a phrase that otherwise invokes a beautiful image of colorful ribbons and joyful celebration is associated with a pattern on brain imaging usually accompanied by a grim diagnosis: profound hypoxia, epileptic seizures, metabolic abnormalities such as hyperammonemia or hypoglycemia^{1,2}. But creeping in the corners of our minds was the most feared possibility in her case: Creutzfeldt Jakob disease (CJD).

No one on our team wants to believe this is what she has, and yet it is hard to convince ourselves the brain scans show anything else. Regardless, we do our due diligence: a thorough neurologic exam, blood and urine tests, an EEG to check for seizures or other abnormalities concerning for CJD, and a lumbar puncture to collect cerebrospinal fluid.

We finish our testing and tell her that soon she can return home. Her two sisters had slept in the room during her stay. Early one morning, as our team files out of the room after rounds, one of the sisters slips me a note: "She will want to go back to work. She will want to drive. She is fiercely independent."

As the junior neurology resident, I promise to return in the afternoon to discuss her expectations for life after the hospital and upon her return home. I feel the weight of the impending conversation and briefly doubt myself, reluctant to dive the depths of these sensitive topics with this patient and her family as I remain hyperaware of the magnitude of our discussion. Nevertheless, I remind myself that even though I sign my notes "PGY-2 neurology resident," I am her doctor.

As I walk towards her room that afternoon, I remember my other firsts. My first goals of care conversation was as an intern about code status with an elderly gastric cancer patient. My first death pronouncement was in the ICU right after I watched the patient's blood pressure dip and her heart monitor flatten. The first time I explained to an overwhelmed mother why we would not recommend a feeding tube in her young son with an aggressive brain tumor. This will be my first time discussing with a patient the heavy implications of her disease on her independence and the rest of her life.

Her sisters remain seated against the window, while the patient sits along the edge of her bed, facing them. We discuss the events of the past several months and her understanding of her hospitalization. We reach solid ground, a shared understanding of her likely diagnosis and hospital course. Then we brace ourselves to begin discussing the beyond. She says she would

like to go back to work, to go back to living by herself, to go back to driving. We chat about concerns that I heard: her poor work performance, her sisters' recent recollection of her minor driving accident, worries about safety at home, particularly around stoves and knives. Slowly, we wade across this river of concerns and reach the other side. The side where I take a deep breath and say, "I would recommend that you take a break from work," "I would recommend that you not drive," and "I think we would all feel better if you could stay with your parents or family for the time being."

With each utterance, I sense her deflating, slowly. Each sentence was like another tap of a chisel, introducing in both her heart and my own heart a fine crack, again and again, not enough to shatter, but enough to weaken and wound. Because, as the disease strips her of her memories and her abilities, I feel I am the one stripping her independence, one life activity at a time. I know it is not me. I am not the jailor who says she is imprisoned. Instead, I am the air traffic controller who sees the trajectory of the plane and guides its way to avoid harm, just as I see the markers of her decline and hope to guide her out of harm's way.

I wonder if my reframing our conversation to prioritizing her safety and well-being and that of her family will suffice. I am, after all, persuading her to surrender several activities that defined her pre-disease independence. Nevertheless, by emphasizing her family's reassurance with these changes, we could, perhaps, create shared agreement. It is a delicate act, balancing both her and her family's different goals. Years later, I strive to recenter conversations of loss to ones that focus on other meaningful elements of life and encourage a transition towards gradually accepting change.

I ask her how she feels. She thinks deeply. Her sisters sit silently with their legs crossed. Their gazes fix intently on their sister. She looks down at her fingers, fidgeting with the edge of the hospital blanket. She looks up and to the right at her purple felt hat and purple scarf sitting on the square wooden side table next to the hospital bed, searching for familiarity. Even though her memory is poor, and she cannot recall all details of the hospitalization, she senses the conversation's gravity and its implications. She nods. "I knew it was serious when they told me to come to the hospital, directly from the clinic." She agrees to take a break from work, to live with her parents, and to stop driving.

The final morning before discharge, I walk into the room. She sits at the edge of her bed, her wool, cream-colored sweater contrasts with her flowing black skirt like diffusion restriction on a DWI sequence. On her head was her purple hat, and the purple scarf was wound about her neck. Her notebook is open, and it is obvious that she has traced, over and over again, the outlines of her multiple attempts to copy the intersecting pentagons I had asked her to copy when she was first admitted. The lopsided figures do not quite intersect as they should. The corner of one of the pages has been torn out. As our team files out of the room after rounds, she hands me the folded torn edge of the paper, upon which she has scrawled in looping cursive, "Thank you, Doctor."

Epilogue: This conversation was a brief window into this patient's journey. She was seen one additional time in neurobehavior clinic, where they discussed the diagnosis of CJD based on positive test results (CSF 14-3-3/RT-QuIC), her poor prognosis and the limits of medical therapies, while offering available resources. She died ten months after this hospitalization.

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Neurology published online December 16, 2022

DOI 10.1212/WNL.0000000000201698

This information is current as of December 16, 2022

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