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DIAGNOSIS

## High Altitude

By LISA SANDERS, M.D.

The middle-aged woman lay in the intensive-care unit, observing the tumult around her. “At least I won’t die alone,” she said to herself. She squeezed her husband’s hand. Though she hadn’t felt well for more than a month, it was not until earlier that day that it occurred to her that she might be dying.

In the afternoon she had taken her teenage daughter shopping. As her daughter disappeared down an aisle, the woman struggled to keep up. Her breath was rapid and ragged. She could hear her blood pounding with each heartbeat. Suddenly the whole right side of the world seemed to go out of focus, color and shapes blending together. She didn’t want to frighten her daughter, so she said nothing. “Please just let me get home,” she prayed silently. She drove with her bad eye shut, and when she got home she promptly lay down. When the phone rang, she put it to her ear without opening her eyes. “Could you come to my office right now?” Dr. Andrea Needleman, her new physician, asked. “And bring your husband?”

The patient saw Needleman the day before, because her regular doctor of nearly 20 years didn’t seem to be listening to her. She was 50 and had always been healthy — until now. The symptoms began while she was visiting friends in Ecuador. Their house was high in the Andes; when the fatigue and weakness first hit her, she figured it was the altitude and would pass. But it didn’t. Then the nausea and [diarrhea](#) started. In the airport, on the way home, she felt so sick and weak that she couldn’t even carry her purse. Since returning, all she wanted to do was sleep.

In her office, Needleman pulled a couple of pages from a file. The blood tests provided some answers but prompted important questions as well. The patient was severely anemic — that’s why she felt so weak and tired. Her platelets, an essential component of the clotting system, were low, and her kidneys were hardly working. She needed to go to the hospital. “Now?” the patient asked, her voice quavering. Yes, now. Needleman wasn’t sure what was going on, but she was very sure that her patient needed to be in the hospital until they figured it out.

When the patient arrived at the emergency room, her [blood pressure](#) was 225/115 — terrifyingly high. A [CT scan](#) showed no sign of a [stroke](#), but her intermittently [blurred vision](#) suggested that her spiking blood pressure was taking a toll. The initial workup revealed an important clue. The I.C.U. doctor saw fragments of red blood cells in her blood. From that, he knew that abnormal clots were forming inside the blood vessels. These intravascular clots were causing the damage: her red blood cells were being torn apart as they were forced through the vessels. That was why she was so anemic. The platelet-filled clots also obstructed the blood vessels, starving the kidneys and other organs. As Needleman explained the results, she saw rising alarm in her patient’s eyes. “It’s going to be O.K.,” Needleman said, with as much confidence as she could

muster. “We know how to treat this.”

Only a few diseases will cause this kind of intravascular clotting; all are potentially deadly. The I.C.U. doctors were particularly worried about H.U.S. (hemolytic uremic syndrome) or its cousin [T.T.P.](#) (thrombotic thrombocytopenic purpura.) H.U.S. is a disease linked to a diarrheal illness usually caused by *E. coli* O157:H7 bacteria, which release a toxin that injures the lining of the blood vessels, causing intravascular clots to form. Although the patient’s symptoms fit, H.U.S. is usually seen in very young children.

T.T.P. can present a similar picture and, although rare, is more common in adults. The immune system mistakenly destroys a naturally occurring clot-buster, allowing clots to form willy-nilly inside vessels. Most patients also have neurological symptoms — this patient had blurred vision. [Kidney injury](#) is less common but can occur. The treatment for T.T.P. and severe cases of H.U.S. is to replace the plasma, the fluid that carries red blood cells, which helps to get rid of the toxin in H.U.S. and provides the missing clot-buster for patients with T.T.P.

Other diseases were much lower on Needleman’s list. Certain severe infections will cause the coagulation system to form these abnormal clots. But the patient had no signs of infection. [Malignant hypertension](#) can also cause these clots, but that is usually seen in people with longstanding, poorly controlled [high blood pressure](#). While the patient’s blood pressure was elevated when she came to the hospital, she had no history of being treated for hypertension. Needleman ordered tests to look for evidence of H.U.S. or T.T.P.; blood cultures to look for hidden infection were already sent. The patient was on a powerful medicine to control her wildly elevated blood pressure. The next morning, the daily plasma exchanges started. By week’s end, her red-blood-cell and platelet counts were rising, and her blood pressure was nearly normal. She felt better. And she was coming to terms with her unusual diagnosis.

Needleman, however, was not. By now the blood test ruled out H.U.S., but the specialists she consulted weren’t at all sure this was T.T.P. either. While the patient seemed to be responding to treatment, her kidneys were still not working. It would take a kidney [biopsy](#) to tell them if this was T.T.P. or something else.

The patient was reluctant. Wasn’t her response to therapy proof that the diagnosis was right? Needleman shook her head. Her case was atypical enough that she felt they needed a definitive test.

And she was right. This wasn’t T.T.P. The biopsy pointed to something much lower on Needleman’s list. The most likely cause, the pathologist said, was malignant hypertension — a physiological crisis seen in people with chronically untreated high blood pressure. Needleman was surprised and asked her patient if she’d ever received a diagnosis of high blood pressure. The patient said that her previous doctor occasionally told her that her blood pressure was high. But he hadn’t wanted to treat her, she reported, because at other times her blood pressure was normal.

How and when to treat high blood pressure has changed drastically over the past 20 years. As our understanding of the damage done by hypertension has increased, so has our willingness to treat even modest elevations. The current guidelines put a “normal” blood pressure at less than 120/80 and recommend treatment after two readings over 140/90.

High blood pressure is called “the silent killer” because it is usually asymptomatic and leads to other diseases — like heart disease and stroke — that do the killing. And yet sometimes blood pressure can get so high that it can cause devastating injury. Although only 1 percent of those with hypertension will develop malignant hypertension, high blood pressure is so common that a mere 1 percent can translate into hundreds of thousands of cases each year. By contrast, there are, on average, only 300 to 400 cases of T.T.P. a year in the U.S. Doctors are taught to treat T.T.P. even before the diagnosis is certain, but we are also taught that an unusual presentation of a common disease is far more likely than even a classic presentation of a rare disease. That was certainly the case with this patient.

Now, nearly six months after she became ill, the patient’s vision is back; so is her energy. Her blood pressure is well controlled on two medications. But her kidneys still aren’t working, and she is reminded of her illness three times a week, when she goes for [hemodialysis](#). Still, she remains hopeful that her kidneys will recover and grateful that she found a doctor who listened.

*Lisa Sanders is the author of "Every Patient Tells a Story: Medical Mysteries and the Art of Diagnosis."*

*If you have a solved case to share with Dr. Sanders, you can e-mail her at [lisa.sandersmd@gmail.com](mailto:lisa.sandersmd@gmail.com). She is unable to respond to all e-mail messages.*

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