

Health

Clinician Story: A Specialist Corrects Misdiagnosis of Sarcoidosis to HLH and Lymphoma

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THE CLINICAL CASE

A female patient in her early thirties came to me for a second opinion regarding a diagnosis of sarcoidosis based on a lymph node biopsy that showed granulomas. Per the guidelines for managing sarcoidosis, she had been treated with prednisone, but was getting worse rather than better. When I examined her, she had a 104 degree fever of unknown origin.

DIAGNOSIS AND TREATMENT

When I reviewed her case, I thought it was atypical for sarcoidosis, which I had seen many times. I started by reviewing causes for granulomatous inflammation (Initially, I find it helpful to do the broadest possible search with something like Google, rather than trying to zero in on one thing). The search led me to a disease that I had never heard of: hemophagocytic lymphohistiocytosis (HLH), which is a disease of the immune system that typically occurs in children, but, in rare cases, can affect adults. I don't typically handle pediatric cases, which explains why I had not encountered it before.

tests, including a ferritin blood test, which confirmed our suspicions that she had been misdiagnosed with sarcoidosis. My patient did indeed have HLH. We reviewed the information together and I emailed her the UpToDate patient education topic on HLH so she could do more extensive research on her own. The patient was well equipped to advocate for herself and went back and forth between her hematologist-oncologist and me with questions.

Ultimately, the patient was admitted to the Mayo Clinic for a bigger biopsy of her liver and a splenectomy. She was subsequently diagnosed with both lymphoma and HLH, and was started on appropriate treatment. Her oncologist has since told me the patient is responding well to treatment.

Let's recap what happened to my patient: Her initial diagnosis was inaccurate, which sent her down the wrong pathway for treatment; she continued to feel worse. UpToDate helped me to uncover the correct diagnosis, which is fortunate, since it's likely she would have been sent to another specialist. This would have cost her time, and she may have experienced further deterioration and weakening in her condition. HLH is quite serious; the course correction in her diagnosis may have saved her life.

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I next turned to UpToDate and found a comprehensive discussion of HLH, even for adults. I reviewed the list of criteria and symptoms together with the patient and discovered that most of them matched her symptoms. We ordered a few additional

There are two key lessons to learn from this case. First, we have to be careful in treating young and otherwise healthy patients. They have so much reserve to fall back on that sometimes we don't appreciate how sick they are. The same symptoms in an older, frail person would trigger alarms and possibly

faster treatment. Second, this patient had been assigned a diagnosis with a degree of certainty that was unwarranted. Once you offer a definitive diagnosis, some doors close. That was a disservice to her, in that it cost her time. The wrong diagnosis is much more harmful than saying, "I'm not sure."

TRUST IN UPTODATE

Medical decision-making often involves uncertainty, and UpToDate does a terrific job of helping physicians to navigate that. It's important for us to think about what we know, but perhaps more importantly, what we don't know, when making diagnoses and treatment recommendations.

There are four main scenarios within which I apply my medical knowledge and training, and I use UpToDate for all of them, albeit, for different reasons:

- First, as a specialist, I see patients on a daily basis who have conditions with which I'm quite familiar. However, I still check UpToDate to make sure I haven't missed anything. There may be several points I've covered, but UpToDate might raise the one additional point we should discuss.
- Second, there are conditions that I know quite a bit about but don't see as often, such as hypertension. I use UpToDate to double check my management plan and confirm it is still based on the most current clinical evidence.
- Third, there are conditions I encounter only occasionally. I know I've forgotten some of the specifics, so I use UpToDate to make sure I'm treating my patients according to current best practices.
- Fourth, there are some cases I've never encountered before. In the case of HLH, UpToDate helped me learn enough about the condition in a matter of minutes, which had a potentially life-saving outcome for my patient.

In addition to trusting UpToDate content to treat my patients, I value being able to access UpToDate within the EHR. The pop-up menus and suggestions are really helpful, and of course, highly relevant to my patient. I also use the drug interactions database from Lexicomp® on a daily basis, which ensures I'm keeping my patients safe from medication errors.

Lastly, to further engage patients in their care, I share UpToDate information written especially for patients. In the exam room, I show them graphs, tables, photographs, illustrations and other information, which I also email. I've found that patients appreciate the chance to further review the information on their own and share it with friends and family. When patients truly understand the nuances of their condition, they can participate more fully in their care and make more informed decisions.

Using UpToDate in my clinical practice gives me the confidence to take on challenging cases and provides a solid platform for life-long learning. I truly cannot imagine practicing without it.

BIOGRAPHY

Dr. Clark attended the University of Washington for his undergraduate studies, medical school and his residency. He then completed a pulmonary fellowship in Vermont. He has been in private practice in Anchorage, Alaska since 1995, specializing in pulmonary medicine. He is affiliated with Alaska Regional Hospital and Providence Alaska Medical Center. He also works at an outreach clinic in Homer, Alaska and serves as a clinical instructor at the University of Washington's Department of Medicine. In 2016, Dr. Clark was honored with the Washington Thoracic Society's Outstanding Clinician Award.

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