Perspective

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When you know too much for your own good: The Boston-Varon syndrome

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Introduction

It has been stated that knowledge is everything. However, sometimes this can actually be counterproductive. For example, the so-called "medical students' disease" (or second-year syndrome, intern's syndrome), has been described as a condition in medical students, who perceive themselves to be experiencing the symptoms of a disease that they are studying. (1) This syndrome is associated with an intense fear of contracting the disease in question. (2)

An identical syndrome could potentially be ascribed to those who care for patients, and in whom on of their family members suffer a condition that requires diagnosis. Here, we describe a variant of such syndrome, for which we have coined the term Boston-Varon Syndrome (BVS).

Case 1

A 24-year-old, previously healthy, woman, daughter of a clinician who cares for critically ill patients, was admitted to the hospital with fever to 41 ^{o}C, chills, and severe dehydration. Upon admission to the hospital, she underwent a complete microbiological assessment that was immediately followed by the administration of intravenous fluids and broad-spectrum antibiotics. Initial laboratory tests were significant for leukopenia (3000/µl) with normal differential and thrombocytopenia (90,000/µl). Despite the antibiotics and fluids, she remained febrile not responding to anti-pyretic agents and her transaminases tripled in less than 24 hours. A complete vasculitis workup was also non revealing. On her third hospital day, she developed a generalized maculopapular rash which did not improve with discontinuation of the antibiotics nor steroids. This rash brought up additional investigations which did not reveal the etiology of her condition. While she was undergoing work, her father, a seasoned critical care physician, constantly discussed her case with different consultants offering his own medical opinions as far as diagnosis and treatment. Moreover, this clinician developed significant anxiety symptoms regarding the potential complications of this acute illness. After several days of modifications in antibiotics, her symptoms improved and eventually she was discharged home. Some of her symptoms, including the maculopapular rash, remained for several weeks, for which her father looked for second and third medical opinions. This process took close to 6 weeks before she was able to go back to her normal activities. Upon writing of this article, she is doing well with no apparent long-term sequelae. The final diagnosis was sepsis of an unknown organism.

Through this process, her father was significantly concerned about a variety of lethal clinical conditions that could cause her signs and symptoms. In addition, he considered conditions that probably most clinicians would not consider initially, such as atypical presentation for vasculitis, B-cell lymphoma or a serotonin producing tumor. He encouraged the healthcare practitioners involved in his daughter's care to think for some of these unusual conditions, such as adult Still's disease.

Case 2

A 16-year-old, previously healthy, young woman, daughter of a lawyer who represents healthcare

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providers in lawsuits, began feeling slightly ill one evening. The next morning, she appeared to be "fighting a cold". As the day progressed, she complained of a sore throat, minor body aches and developed malaise. By the next morning, she suddenly felt much worse with a documented temperature of 40 ^{o}C. Four hours later, she was normothermic but she had generalized weakness. She complained of generalized abdominal pain, body aches and a worsening sore throat. She developed significant dysphagia followed by nausea, vomiting, and diarrhea. She also reported intermittent bilateral lower extremity numbness and tingling. She was taken to a free-standing emergency department (ED) where throat and nasal swabs were done. On initial exam, the throat was not particularly inflamed. She had intermittent tachycardia and persistently low blood pressure. Intravenous fluids were started and a complete laboratory work up done. However, her abdominal and overall body-pain were worsening. Morphine was offered and her father initially declined it, but her pain became intolerable and this medication was given. As expected, this therapeutic intervention dropped her blood pressure even more. She became extremely pale and groggy. An abdominal computed tomography (CT) and chest radiograph were done. Both were essentially normal but for possible gallbladder wall thickening. At that point she was not able to urinate and her blood pressure remained low despite 4000 cc on 0.9 Saline solution IV. There was no rash. And attempt to transfer to a large, tertiary care children's facility was done, but they declined acceptance pending increased, stable, blood pressures.

As the morphine wore off, she became more alert and conversant but otherwise her symptoms remained virtually unchanged. The ED physician's working diagnosis was "septic shock, possibly toxic shock syndrome". She was started on vasopressors (epinephrine) and empiric triple antibiotic therapy and admitted to the intensive care unit (ICU). Laboratory testing revealed elevated Epstein-Barr antibodies, possibly mononucleosis related, but virtually all inflammatory and antibody markers were significantly elevated and her presentation looked more like a bacterial infection rather than viral. White blood cell count and platelets were very low at 3000/µl and 58,000/µl, respectively.

Within about 48-72 hours of ICU admission, her hemodynamic parameters improved significantly. She was gradually weaned off epinephrine. Her appetite and energy slowly returned. She was moved to a regular floor. Antibiotics were incrementally peeled off but for oral doxycycline. She was discharged on day six essentially normal. It was as if she had never been sick. All swabs and cultures were negative. The cause of her sepsis remains a mystery.

In this case, her father, a very well-respected lawyer that specializes in defending healthcare providers, also reached out to clinicians that he identified as key opinion leader in the diagnosis and management of sepsis for advice and to vent some of his concerns. He also developed anxiety related to the potential complications of his daughter's clinical condition. Indeed, upon seeing his daughter's vital signs, he immediately thought "septic shock". When she did not urinate despite IV fluids in the context of her low blood pressures, he thought about hypoperfusion-related "acute kidney injury" as her body was shunting blood to the brain and heart. As a concerned father, his mind was racing. He could not figure out why she would be septic. She had no cuts, infected insect bites, etc. Was it the throat? It was painful but not significantly inflamed. Her abdominal pain was severe. Did she have an intestinal perforation? Was she bleeding? The CT ruled out any significant abdominal etiology. At that point, her father began second guessing himself. Perhaps he was overreacting. However, upon admission to the ICU, some of his thoughts were confirmed. His daughter was in septic shock. Once again, his mind started racing again. Was it bacterial? Was it some weird pathogen he did not know about? If so, how was it contracted? Was there a chemical toxin involved? Could this be vaping-related? Did some other person slip her something into a drink? Was this a weird autoimmune disorder? Would she experience sepsis-related disseminated intravascular coagulation? Would she get ischemic-related organ damage? This well-respected lawyer, remembered mortality statistics that he had seen through the years of practice. He actively encouraged the healthcare practitioners caring for his daughter to "think outside the box".

Discussion

The development of anxiety related to a clinical condition, when suffered by someone that is familiar with the illness is quite common. One of us has previously described his own experience when suffering from a life-threatening, catastrophic medical event. (3) In such instance a recollection of the events that lead to the illness as well as the concern for potential adverse outcomes was outlined. In some instances, the emotions that are felt can cloud clinical judgement. (4)

In our daughters' cases, both authors were aware that the mortality rate and long-term complications of sepsis remain elevated despite every known therapy available. (5-7) As such, the concerns for death and long-term sequelae were legitimate. Indeed, when we were confronted with the illness of our children, we both experienced severe anxiety, as we were well aware of these facts. More importantly, having dealt with situations that involve sepsis on regular basis (either medically or in the legal standpoint), we were aware of everything that could potentially gone wrong.

Even though anxiety is a normal and often healthy emotion. However, when we felt disproportionate levels of anxiety, due to our understanding of our daughter's illness, it could have become a medical disorder. Fortunately, both of them had good outcomes and our anxiety resolved.

Is there a way to avoid the BVS? The fear of a loved-one having a negative outcome is difficult to avoid when the fund of knowledge on a specific illness is vast. We both believe that knowing too much is a blessing (for our patients and clients) and a curse (for us). We suggest that clinicians who have family members or loved ones gravely ill, attempt to let their colleagues do their work and rather than try to find answers and offer clinical opinions, you focus your energy of supporting the person who is ill as well as those who are worried about the situation and maintain positive attitude.

The Boston-Varon syndrome is a real condition that need to be taught to health care students in an attempt to minimize the anxiety associated with this condition.

We are both happy (and thankful) that both of our daughters did well without any major complications, and that we are now much more relaxed about what happened.

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