COMPLEX AUTOIMMUNE DISEASE MANAGEMENT IN INDIGENOUS YOUNG ADULTS:

A CASE REPORT

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Background:

Autoimmune diseases are characterized by systemic inflammation, where a dysregulated immune system targets normal body functions or organs.[1] There are over 80 autoimmune diseases, such as Type 1 Diabetes, rheumatoid arthritis, lupus, psoriasis and celiac disease. Studies have shown that women are 3 times more likely to suffer from an autoimmune disease than men, that risk increases furthermore if you are of indigenous descent.[2] Collectively, up to 7% of the population in North America are affected by some type of autoimmune disease, with Rheumatoid Arthritis being the most common (1%).[1] Despite aboriginal women being one of the most at-risk populations to be affected by autoimmune disease, information on their disease course and management is sparse. This is due to external influences such as lack of access to health care, lower socio-economic status to afford treatment, and indigenous populations' distrust of the current medical system.[3] The patient reported in this case study is a young indigenous female with multiple debilitating autoimmune diseases. It showcases the importance of multi-disciplinary approach to patient management and provides a screenshot of the impact of autoimmune diseases complications to indigenous women.

Clinical question:

How are patients with complex autoimmune diseases managed in the primary care setting?

Case History:

WS is a 28-year-old indigenous female with currently living in rural Manitoba. She has a past history of poorly controlled type 1 diabetes, rheumatoid arthritis, celiac disease, hypothyroidism and psoriasis. She was followed by her family physician since she was 19 years

old and has been admitted multiple times to the hospital due to exacerbations of her medical conditions. Her most recent admission to the hospital was from May 25, 2019 until May 31, 2019 due to a seizure.

Pre-admission:

On the afternoon of May 25th, patient's mother found her collapsed on the floor unconscious. Mother reported patient was non-verbal and groaning on the ground. She bit her tongue and had foaming in mouth. This lasted for more than 5 minutes. Her GCS was initially 8, increased to 12 when EMS arrived but dropped to 8 again during transport. Prior to this episode, mother said patient was complaining frequently of tiredness and had many staring spell lasting a few seconds 2 weeks leading up to episode; she had a tonic clonic seizure at the hemodialysis unit 2 weeks prior but with no workup.

In hospital:

Patient continued to have right gaze deviation with eyelid fluttering and was concerned for status epilepticus. Given 5mg IV midazolam x2, then intubated, sedated with propofol and midaz in ICU. Dilantin was loaded. CT of brain was done, which revealed normal findings. Patient then transferred to ICU. In ICU, EEG showed irregular but no definite seizure activity. MRI brain done on May 26 (day after) and neuro consulted. MRI showed enhancing lesion in the left frontal lobe, which looked like a focal thrombosis of a vein or leptomeningeal process. Neuro reports this to be the cause of her seizures.

She was extubated on May 27 and was slowly improving. She was transitioned to SC insulin on May 28th.

Upon her transfer to the wards, she was feeling tired but otherwise well with no other symptoms. She was discharged on May 29 to be followed up by her family doctor.

Past Medical History:

1. T1D:

- Diagnosed at 6 years old, initially presented with classic symptoms of diabetes and was found to be in diabetic ketoacidosis.
- Currently has diabetic neuropathy, non-proliferative diabetic retinopathy and elevated liver enzymes.
- BG (blood glucose) not well controlled, would have lows of 1-2 mmol/L weekly. During most recent hospital admission, would have BG range of 2.5 mmol/L to "high" on glucometer. Patient checks her BG QD but has some hypoglycemia unawareness. She has never been able to control her BG due to poor medication adherence and timely glucose checks, despite being followed up by endocrinologists and regular sessions with the diabetes educator. She has been hospitalized multiple times since 2009 due to her poor glycemic control.
- On Tresiba 11 units q. AM and Humalog 1 unit per 8 grams of carbohydrate, with correction factor [(current BG -8)/ 2].
- Feeling dizziness/light headedness every day, feeling nausea few times per week. No neuropathy in external limbs. No falls or LOC.

2. CKD Stage V:

- From complications of T1D
- Previously on peritoneal dialysis (PD) 3 times/ week, will be transitioning to hemodialysis in 2 weeks. PD tolerated well.

3. Rheumatoid arthritis and psoriasis:

- Patient reports being in pain 9/10, especially on left knee and left hip.
- Taking Tylenol 500mg, hydromorphone 300 mg for pain management and Humira.
- Has trouble sleeping due to constant itching, not well controlled.
- On exam, patient has large, cracked plaques around her body, specifically her nose, forearms, hairline and neck. There are scabs and bloody discharge in the middle of the plaques.

4. CHF and HTN:

- Admitted to ER due to a hypertensive crisis and diagnosed in 2018.
- Currently on furosemide, metoprolol and amlodipine for management.

5. Septic L hip arthropathy:

- Admitted to the ER in May due to a septic hip joint. Hip washout done on May 15th.
- No pathogens on culture, given 6 weeks IV ceftriaxone with vancomycin with hemodialysis.

6. Celiac disease:

- Diagnosed at 19 years old, currently on a gluten free diet, but difficult with adherence. Patient is also anemic and is on ferrous sulfate.
- Having diarrhea for past 2 weeks, diarrhea is watery in consistency, no blood in stool.
- Given Imodium for stool control.

8. Hypothyroidism: Well controlled, on levothyroxine.

9. Depression: Patient up at night a lot and is in bed all day, mood is low. On escitalopram 20mg.

Past surgical history: Tubal ligation in 2016

Family Medical History: Patient has very limited knowledge of her family medical history. She has a strong history of RA on her biological father's side.

Social History:

Patient is of indigenous descent and was in a foster care system since she was young. She does not know her birth family well but has a good relationship with her foster mother. Her older sister died of suicide when the patient was a teenager. Patient is currently on disability. She moved out to live with her sister in 2016 but was hospitalized multiple times due to inadequate management of her conditions. Currently is living with her foster mother and in the process of requesting for home care services due to the vast amount of caregiver burden experienced. Patient quit smoking 5 years ago and was a 1-pack- year smoker, she does not drink alcohol or do any recreational drugs.

Discussion:

This patient is a complicated case as she has multiple debilitating autoimmune disorders that are all poorly managed. It is important to report cases like this because it emphasizes the importance of a multidisciplinary approach to care for this patient. She is followed by hematology, endocrinology, rheumatology, neurology, as well as by her primary care physician (PCP). In addition, she regularly sees a therapist, a diabetes educator and a dietitian. She reported the meetings have helped her greatly because she learned CBT methods to help with her depression that could not be addressed in a single doctor's visit. Diet modification and education were crucial to her disease management due to her Celiac disease.

In the context of her recent hospitalization, literature review indicate people suffering from diabetes mellitus have a much higher risk of developing seizures (approximately 25%);

those experiencing diabetic ketoacidosis having an even higher risk.[4] The lesion (focal thrombosis) in her MRI scan could be from complications of poorly managed blood sugars, such as vascular endothelial dysfunction, atherosclerosis or systemic inflammation.[4] The risk is further increased for WS due to her multiple comorbidities, such as her congestive heart failure and hypertension, which are both independent risk factors for strokes and seizures.[5] The PCP thus has the responsibility to explain to her the potential reasons why this hospitalization happened, and the potential steps in the future to prevent seizures from happening again. Discussions include proper blood glucose management, timely follow-ups and having someone with her most of the time so she can get help early on in case similar episodes happen.

In the context of her residing location, she currently lives in a rural area in Manitoba. Even though she lives in Niverville which is a 45 min drive from the city center, she does not have a driver's license due to her medical conditions, which makes it extremely difficult for her to go to her many specialist appointments in the city. This is a problem that many patients face when they live in a remote area, as there is limited transportation services available, limited number of appointments and specialists that are available within their vicinity, making their disease management difficult.[6]

This patient also accurately reflects the socio-economic related challenges of healthcare management in young, indigenous women. Lower levels of education and lower income lead to poorer health outcomes, due to reasons such as not being able to afford drugs for treatment and accessing transportation for appointments.[6] WS reported that because her conditions are so debilitating, she was unable to keep a full-time job since she was 19, and was not able to get a university or diploma education after high school due to her illness. In Canada, 52.2% of children in foster care are Indigenous, but account for only 7.7% of the child population

according to Census 2016.[7] WS was raised in various foster homes since she was young. She found it very hard to establish a sense of belonging, learn about her family culture and learn about healthy lifestyle habits. These circumstances contribute to her lack of disease awareness and medication management, ultimately resulting in her recent hospitalization due to complications.

The primary care physician (PCP) plays a very important role in the management of this patient. The main problem for this patient is her struggle with medication adherence. Upon further discussion, she has trouble adhering to medication because she has little knowledge of the seriousness of her conditions, she doesn't have energy to get up and administer the topical creams, as well as other unwanted side effects. Patients like WS requires strict medication adherence because of the seriousness of the complications, thus it is the PCP's responsibility to educate patients on their medical conditions, as well as establish a treatment plan that the patient can follow. WS's PCP in this case then took the time to discuss ways to make medication adherence easier, such as getting help when applying the creams and talking about the consequences of not taking the meds. The PCP will make sure all her medications are up to date and all her medical conditions are being taken care for by the appropriate specialist. It is crucial to establish a trusting relationship with this patient in the PC setting, as the patient will require long term care and the PCP will be the first to know if new problems arise, or if existing problems progress. It is also very mentally burdening to live with so many medical problems at such a young age, thus providing adequate support such as home care and counselling is also important.

Conclusion:

Many patients that PCPs follow up are of similar nature, with multiple chronic diseases that require long term management. However, WS is much younger than the average patient and has had a lot more trouble with medical management. This young patient is definitely not the only patient suffering from chronic conditions that are difficult to manage. There are more patients in our community that have undiagnosed or underdiagnosed conditions that require our attention, especially indigenous patients since they are more likely to suffer from various health problems than the general population. Primary care providers have the important responsibility of understanding these barriers to access healthcare, having adequate cultural competency, and taking a multidisciplinary approach to patient care.

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