AMERICAN JOURNAL OF MEDICAL AND CLINICAL SCIENCES, 2023
VOL 9, NO. 2, PAGE 1 – 3

REVIEW ARTICLE

Open Access

Explosive onset of SLE in white male in capital of Cherokee Nation

Caleb S. Alexander[1] and Prashant Kaushik[2]*

[1]Oklahoma State University, Center for Health Sciences, College of Osteopathic Medicine, Cherokee Nation, Tahlequah, Oklahoma, USA. [2]Chief of Rheumatology, Northeastern Health System, Tahlequah, OK; Associate Program Director, Internal Medicine Residency Program, TMG/OMECO at NHS, Tahlequah, OK; Clinical Professor of Medicine, OSU CHS Cherokee Nation, Tahlequah, Oklahoma, USA.

ABSTRACT

Systemic lupus erythematosus (SLE) is a major autoimmune rheumatologic condition that can cause severe life threatening complications. While it afflicts all ages, sexes, races and ethnicities, it overwhelmingly affects more females and non-white males. Due to these demographics, it is important to highlight this case of an explosive onset of SLE in a young, white male from a unique location in the USA. A 19-year old, white male first had symptoms following a change to carnivore diet, increase in exercise and prolonged sun exposure. Over the first few months, a rash that had started on his face and was originally thought to be a sunburn spread across the majority of his body. He then developed ascending joint pains and a loss of appetite. Over the course of about 6 months, he had gone from a relatively healthy, but overweight, teenager to requiring hospitalization for severe body pains and rash, photophobia, hyperacusis, lack of an appetite and nearly 30-percent loss of body weight loss. Since disease identification and receiving treatment, the symptoms have begun improving and he is resuming normal daily activities.

ARTICLE HISTORY

Received 11 Jan 2024
Accepted 23 Feb 2024
Published 01 Mar 2024

KEYWORDS

Systemic lupus erythematosus, white male, explosive onset.

Introduction

Systemic lupus erythematosus (SLE) is a major chronic systemic inflammatory autoimmune rheumatologic condition that can cause severe and life threatening complications [1]. In a study examining over 5000 cases of SLE in the United States, it was found that prevalence of disease was about 128.7 cases per 100,000 females and 14.6 cases per 100,000 males [2]. Looking further at males, the prevalence (95% CI) in the American Indian/Alaska Native (AI/AN) population is 53.8 (36.2–77.1) per 100,000, 26.7 (19.6–36.4) in the Black population, 11.2 (5.7–21.9) in the Asian/Pacific islander population, 8.9 (8.0–10.1) in the White population, and 18.0 (15.6–20.8) in those identifying as Hispanic. Further, a study of patients with SLE according to the Systemic Lupus International Collaborating Clinics (SLICC)/American College of Rheumatology (ACR) criteria found that 93.2% of patients were female with a mean (SD) age of 45.3 (13.9) years and 11.1 (9.2) years since diagnosis [3]. Considering these disease demographics, SLE may not be as high on the differential for possible cases among young, white males.

In January 2021, the first Division of Rheumatology was started at the Northeastern Health System, a health care facility in Tahlequah, Oklahoma, the capital of the Cherokee Nation (CN). It has been providing full-time Rheumatology services to a large population, including all of the Cherokee Natives, via a mixed-model offering both face-to-face and virtual Rheumatology visits [4]. This has been shown to be the optimal combination, overcoming the barriers to accessing care posed by distantly living patients, while also mitigating the limitations of virtual consultation [5]. It is at this clinic that a case of SLE with explosive onset in a young, white male was managed. This report expresses the details of this case to add to the available literature and emphasize the importance of considering SLE diagnosis in appropriate young, white, male patients.

Case

A kind 19-years old male presented to the rheumatology clinic as his initial visit after a recent hospitalization during which a diagnosis of SLE was made by a non-Rheumatologist Hospitalist physician. His symptoms began about 8 months ago when he decided he wanted to lose some weight and started a strict carnivore-diet. He began at about 333 pounds, and was asymptomatic. Two months into the carnivore diet, he developed a smooth, light pink rash on his cheeks following a float trip on a river that was thought to be a sunburn. This rash persisted for about a month until they began trialing a mix of neomycin and hydrocortisone to the area per the recommendation of a friend with a similar "sunburn". This did not provide any relief and the areas on his cheeks began getting progressively more red. He was working with his brother in construction at this time, but noticed that he was becoming more sensitive to the building supplies over the next month or two. He noted pain beginning in his feet, then knees and then diffusely, and he also lost his appetite in this time period. By the end of the next 2 months, he had developed a rash all over his body, photophobia, hyperacusis and was becoming more ill with fatigue, malaise etc.

Contact: Prashant Kaushik, MD, FACP, FACR, Rh MSUS
Dip ABLM 1373 E. Boone St., Suite 2300, Tahlequah, OK 74464, Phone Number (918) 207-0025; Fax number (918) 207-0226.

In the early morning of the penultimate day of the year (about 7 months since the first symptoms), he was taken to the Emergency Room by his family. At this time he was down to 240 pounds (a loss of nearly 100 pounds in about 6 months). From the Emergency Room, he was transferred to a hospital in a nearby city for a higher level of inpatient care. At this time, he was diagnosed with SLE, treated with high dose intravenous pulse steroids and started on hydroxychloroquine, apparently by a non-Rheumatologist hospitalist physician. He remained in the hospital for about 5 days. Following discharge, he was seen by his primary care physician who recommended him to be seen by Rheumatology. In the meantime before the first Rheumatology appointment, he slowly gained back his appetite and was now holding his weight. His mother stated that he now looks better than he has in the past several months.

Despite the apparent improvements in condition, this young man had been taking the maximum dose of hydroxychloroquine (400 mg daily) and been on 10 mg of prednisone per day for over a month. He had begun developing moon facies and still has a significant malar rash and palpable purpuric rash on his arms. Further, he had the skin rash, fatigue, brain fog, left knee and neck pain and some chest pain when he woke up that lessened as he took big breaths and opened up his lungs. He also started experiencing Raynaud's phenomenon.

His family history was significant for a father who passed away from blood clots and related cardiac disease at 56 years of age, and 3 brothers with autoimmune conditions: oldest brother with type 1 diabetes mellitus, the second oldest with juvenile rheumatoid arthritis, and the third 'eczema'. He has 1 other brother and 3 sisters who are healthy. He is the youngest of the 5 brothers.

A detailed rheumatologic lab workup before the first Rheumatology appointment was as follows A complete blood count showed a normal white blood cell count, normal platelet count with hemoglobin was slightly low at 13.2 g/dl (normocytic and normochromic anemia). Serum AST, ALT and creatinine levels were normal. Complement C3 level was normal but complement C4 level was low at 13 mg/dL (normal range 15 to 53 mg/dL). Serum creatine kinase level was normal. Urinalysis was free of protein or blood. Lupus anticoagulant was not detected. Beta-2 glycoprotein IgG and IgM antibodies were negative. Anticardiolipin IgG and IgM antibodies were negative. Phosphatidylserine/prothrombin IgG and IgM antibodies were negative. QuantiFERON-TB Gold was negative. HLA-B27 was negative. Cyclic citrullinated peptide antibodies were negative.

Antinuclear antibodies were strongly positive at a ratio of 1:1280 nuclear, homogenous pattern, AC–1 ICAP pattern. Double-stranded DNA antibodies were significantly elevated at 80 IU/mL (> 10 being positive). RNP antibodies were positive at 4.2 Units. SSA and SSB antibodies were negative. Smith antibodies were significantly elevated at greater than 8 Units. Histone antibodies were positive at 2.8 Units. Rheumatoid factor was negative. Jo 1 antibodies were negative. RNA polymerase III antibodies were negative. Hepatitis B status was unimmunized. Hepatitis C antibodies were negative. Galactose alpha–galactose IgE antibodies were negative.

Shared decision making process yielded a plan to proceed with treatment of refractory SLE with an explosive onset. Due to the severity of this case, it was strongly recommended to add belimumab to the current maximal daily dose of hydroxychloroquine. Further, it was recommended to taper the prednisone as follows: decrease the prednisone dose by 1mg/day every 2 weeks until it can be discontinued (meaning 9 mg/day for the next 2 weeks, then 8mg/day, and so on). It was also recommended that activity can be resumed as tolerated, however UVA and UVB exposure should be limited and SPF >50 sunscreen should be generously applied during prolonged sun exposure. Lastly, it was recommended to follow up in the Rheumatology clinic in about 4 months with surveillance-labs repeated about 3 weeks prior to the visit. Our office is available to be called with any concerns, and he can be seen earlier if necessary. He and his mother were very appreciative of the detailed plan of care.

Discussion

There are multiple factors contributing to the importance of sharing this case of SLE in a young, white male in rural Oklahoma. First, this patient's demographics are atypical to the common population that this disease afflicts. As described above, the prevalence of SLE among white males is estimated to be between 8 and 10 per 100,000 population [2]. Further, a study of emergency department visits among patients with SLE has found that the majority of patients were female (90.54%) [6]. This study also expresses that among the white SLE patients, the mean age was 53 years and only 8.54% were in the 18-30 year age group. While the exact distribution of males to females for each racial/ethnic category by age bracket is not explicitly stated, it can be extrapolated - using the equation: (% male in study)*(% white in age group) - that the number of white males between the ages of 18-30 in this study is less than 1% of the population. The rarity of patients matching the demographics of our presented case strengthens the necessity to share these details to improve care for future patients.

Another interesting finding in this case is the presence of multiple different autoimmune diseases in the patient's brothers. This is significant as there is often familial clustering of autoimmune diseases, many of which have been associated with specific genes. For example, a functional single-nucleotide polymorphism (rs2476601, encoding R620W) in the intracellular tyrosine phosphatase (PTPN22) has been associated with increased risk of type 1 diabetes mellitus, rheumatoid arthritis, SLE and Hashimoto thyroiditis [7]. Although genetic analysis has not been completed for our patient, future investigation could reveal the presence of this gene mutation.

He had not been on any obvious drug that could have induced lupus. Hence, the exact etiology and clinical significance of the positive anti-histone antibodies remains obscured. Nonetheless, in SLE, the body is known to become a 'warehouse' of various disease-specific as well as non-disease specific autoantibodies.

Conclusion

While SLE is not as common in young, white males as it is in other populations, its unusual presentation should not be

overlooked. This disease and its typical presentation are well described in literature; however, the explosive onset in this young white male patient should serve as a reminder that 'diseases just don't read the book'.

References

1. Bernatsky S, Boivin JF, Joseph L, Manzi S, Ginzler E, et al. Mortality in systemic lupus erythematosus. Arthritis Rheum. 2006; 54(8): 2550-2557.
2. Izmirly PM, Parton H, Wang L, McCune WJ, Lim SS, et al. Prevalence of Systemic Lupus Erythematosus in the United States: Estimates From a Meta-Analysis of the Centers for Disease Control and Prevention National Lupus Registries. Arthritis Rheumatol. 2021; 73(6): 991-996.
3. Arnaud L, Furie R, Morand EF, Aringer M, Peschken C, et al. Burden of systemic lupus erythematosus in clinical practice: baseline data from the SLE Prospective Observational Cohort Study (SPOCS) by interferon gene signature. Lupus Sci Med. 2023; 10(2): e001032.
4. Kaushik P, Ashraf A, Marak CP, Baker J. Virtual Rheumatology during COVID-19: 'A gift in a seemingly ugly package'. Oklahoma State Medical Proceedings. 2021; 5(2).
5. Kaushik P, Bastible S, Marak C. On telemedicine in rheumatology. Arthritis Care Res. (Hoboken). 2022; 75(6): 1385-1386.
6. Dhital R, Pokharel A, Karageorgiou I, Poudel DR, Guma M, et al. Epidemiology and outcomes of emergency department visits in systemic lupus erythematosus: Insights from the nationwide emergency department sample (NEDS). Lupus. 2023; 32(14): 1646-1655.
7. Criswell LA, Pfeiffer KA, Lum RF, Gonzales B, Novitzke J, et al. Analysis of families in the multiple autoimmune disease genetics consortium (MADGC) collection: the PTPN22 620W allele associates with multiple autoimmune phenotypes. Am J Hum Genet. 2005; 76(4): 561-571.