Hard to see: Systemic lupus erythematous presenting as cranial nerve six palsy: A Case Report

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Abstract

Systemic Lupus Erythematous (SLE) is an autoimmune disease that can affect multiple organs and can have a wide variety of symptomatic presentations. One rare presenting symptom is oculomotor nerve dysfunction, the following case report is of a 41-year-old female with a past medical history of hypertension and asthma, who was admitted for ocular dysfunction. On physical examination, the patient had an oculary palsy, but the rest of the neurological exam was unremarkable. Imaging modalities such as computed tomography (CT) scan head without contrast, brain magnetic resonance imaging, and computed tomography angiography (CTA) of the head and neck revealed no acute pathology. After a neurological etiology was excluded an autoimmune diagnosis was pursued. Lab results were positive for Anti-Smith, ANA, and Anti-DsDNA. The patient also at this time was also diagnosed with an acute kidney injury and a kidney biopsy that was done later revealed focal lupus nephritis. After starting immunosuppressive therapy, her symptoms improved.

Keywords: Systemic Lupus Erythematous (SLE); Anti-Double Stranded DNA (Anti-DsDNA); Anti-nuclear antibody (ANA); Computed tomography (CT); Computed tomography angiography (CTA)

1. Introduction

Systemic Lupus Erythematous (SLE) is an inflammatory autoimmune disease characterized by systemic immune complex deposition and autoantibody production. SLE is diagnosed approximately nine times more frequently in women than men, with an average age of diagnosis of 35. Typical presenting symptoms of SLE include arthritis, skin changes, and renal dysfunction. Isolated oculomotor nerve palsy is a rare sequelae of SLE. It develops in approximately 0.1% of patients with SLE before SLE is formally diagnosed.

In this case report, we present a case of SLE initially presenting as cranial nerve six palsy.

2. Case Report

A 41-year-old female with a past medical history significant for hypertension and asthma presented to the emergency department with new onset blurry vision that began the night prior. The development of her blurry vision was associated with a moderate headache. She denied eye pain.

The only significant finding on physical examination was cranial nerve six palsy on the left eye. Her cranial nerves two through twelve were intact otherwise. Her neurologic exam was otherwise non-focal. She had no deficits in her memory, speech, or cognition. She denied any history of head trauma. At this time, an acute cerebrovascular event or brain tumor was at the top of our team’s differential diagnoses. Subsequent work-up, including computed tomography (CT) scan
head without contrast, brain magnetic resonance imaging, computed tomography angiography (CTA) of head and neck revealed no acute pathology including stroke, hemorrhage, or aneurysm.

Given the lack of significant findings on brain imaging, the differential diagnosis was expanded to include viral causes of cranial nerve six palsy, atypical migraine, and autoimmune etiologies. On further interviews with the patient, she said she had pursued evaluation in the outpatient setting for her chronic symptoms of polyarthritis that so far, to her best knowledge, was unremarkable. The patient expressed interest in obtaining an autoimmune panel inpatient. Thus, the following laboratory studies were sent: serum rheumatoid factor, Scl-70, anti-Double Stranded DNA (Anti-DsDNA), anti-smith, and anti-nuclear antibody (ANA). Her lab results came back positive the next day for Anti-Smith, ANA, and Anti-DsDNA.

Other significant lab findings include an elevated creatinine. The patient denied a history of kidney disease. Her UA showed a significant elevation in urine protein. The results of her 24-hour-urine protein sample showed 1.6g/24 hours, indicating glomerular damage.

Given the patient’s symptoms of chronic, bilateral polyarthritis and cranial nerve six palsy, in addition to her autoimmune panel findings and kidney disease, SLE complicated by lupus nephritis was at the top of our differential.

The nephrology team was then consulted to pursue a kidney biopsy inpatient. The biopsy revealed focal lupus nephritis.

3. Discussion

Ocular motor nerve palsy are commonly caused by neoplasm, aneurysm, stroke, and hemorrhage. It is imperative to distinguish an autoimmune etiology of cranial nerve six palsy given the different treatment regimen required to treat autoimmune disease as compared to other causes. Our case highlights the range of causes associated with this unusual clinical entity.

As confirmed with renal biopsy, our patient’s primary disease process was her SLE. Treatment for SLE complicated by lupus nephritis and cranial nerve palsy typically includes steroids and immunosuppressants. The patient was started on 400mg of hydroxychloroquine daily and 50 mg of prednisone daily. She was referred to outpatient rheumatology and nephrology providers.

In her post-hospitalization outpatient appointments, the patient reported that her ocular symptoms had improved after starting these immunosuppressant medications.

Neuropsychiatric symptoms affect 37-90% of patients with SLE. These symptoms commonly include fatigue, headache, altered mood and cognitive impairment. SLE associated peripheral nervous system involvement is rarer, with an estimated prevalence of 2.2-8.2%. Currently, the pathogenesis of this disease process is poorly understood, but may relate to microvascular damage, immune complex deposition, or injury due to the production of antibodies.

This case is an example of the numerous ways in which SLE can present. Research shows that early (as compared to later) diagnosis of SLE leads to lower flare rates, reduced healthcare utilization, and lower health care costs. It is therefore essential for clinicians to pursue complete medical evaluations for patients with atypical symptoms to ensure these patients are optimized to achieve the best possible long-term health outcomes.

4. Conclusion

The patient’s ocular palsy improved once she began immunosuppressive therapy for SLE. A history and physical are important in autoimmune cases because they help with diagnosing early on. This study will benefit the medical community by advocating for a more thorough history and physical that will direct to an autoimmune workup. The workup could lead to an earlier diagnosis of SLE, leading to earlier treatment.
Compliance with ethical standards

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Disclosure of conflict of interest

No conflict of interest.

Statement of ethical approval

The present research work does not contain any studies performed on animals/humans subjects by any of the authors.

Statement of informed consent

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