CASE REPORT

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Septum Perforation in a Patient with Asymptomatic Systemic Lupus Erythematosus

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Abstract

Background: Systemic lupus erythematosus (SLE) is an autoimmune disease in which organs and cells are damaged by autoantibodies and immune complexes.
This disease often affects the body organs, one of which is the vascular system that manifests itself as vasculitis. Rarely, this vasculitis can cause perforation and epistaxis in the nasal septum. Most people who present this manifestation in lupus are people who have a known lupus case and develop septal perforation over time.

Case Presentation: In this article, a patient with epistaxis and septal perforation was referred and, finally, she was diagnosed with lupus.

Conclusion: This study shows that other than granulomatous and eosinophilic angiitis (Wegner & Churg-Strauss), systemic lupus erythematous should be in mind in diagnosis of septal perforation.

Conflict of Interest: The authors declare no conflict of interest.
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Background

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease that can affect various organs in the body, especially the skin, kidneys, joints, blood, and central nervous system. The disease course includes periods of activity and recovery (1).

The disease has variable clinical manifestations that are caused in the early stages by autoantibodies and its attachment to tissues and immune complex. Diagnosis is based on clinical and laboratory manifestations. One of the rare manifestations is vascular involvement in the form of vasculitis in the nasal septum. Finally, there is septum perforation and epistaxis.

Septum perforation in SLE can be secondary to vasculitis or ischemia and chondrolysis (2).

Case Presentation

A 16-year-old girl presented with a complaint of massive epistaxis in the last few days. On initial examination, extensive septum perforation with at least 70% corrosion of the nasal septum was seen. No necrosis or inflammation was seen at the perforation edges (Figure 1). However, abundant crust was seen along with bleeding points. Erythematous-plaque lesions with a clear border with hypopigmented and itchy centers were seen on initial examination of the trunk and limbs without involvement of the head and face (Figure 2).

In further examination, malar rash and photosensitivity and asymptomatic oral ulcers were seen in the hard palate (Figure 3).

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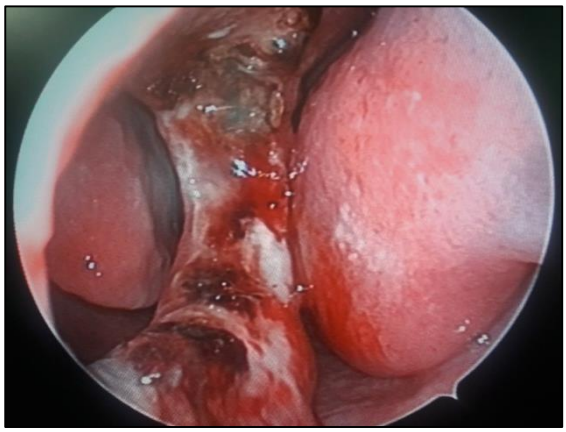

Figure 1. Large septal perforation involving the anteroinferior and anterosuperior parts of the cartilaginous nasal septum.

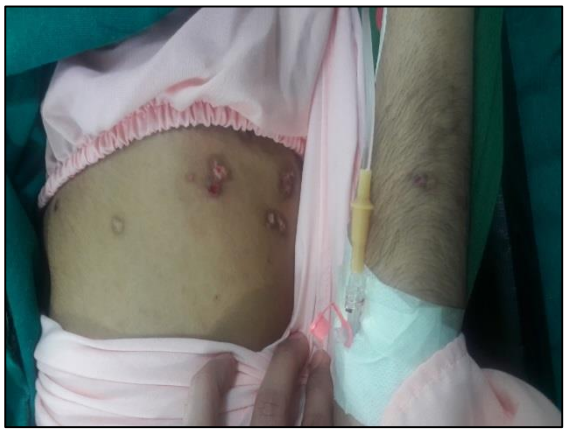

Figure 2. Skin lesions

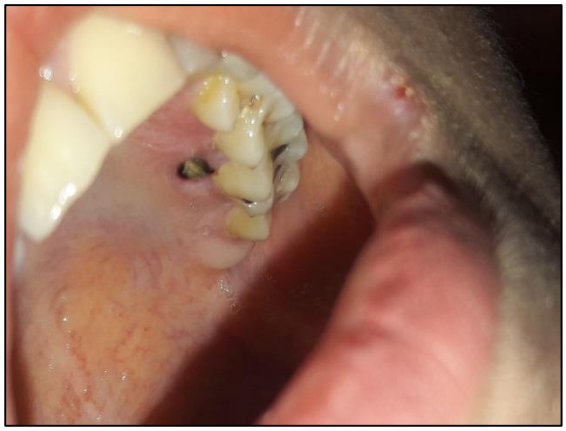

Figure 3. Mouth lesion

On the initial routine tests, coagulation and platelet counts were normal. Anemia was observed in initial CBC tests. Other findings were as follow:
(NL = 12-14) Hb = 10 (reticulocyte count ↑)
HCT = 35/9 (NL = 37-43)
PLT = 377000 (NL = 150-400)
Indirect Bilirubin serum = 3 (nl <0/4)
Direct coombs=(+)
Coagulation tests (PT, PTT, INR) were also normal.
The following tests were performed with suspicion of Wegener's disease:
Rheumatology tests, nasal endoscopy and biopsy of septal perforation edges, biopsy of skin lesions, sinus CT scan, lung CT

Rheumatology Tests
ESR = 49 (NL <20)
F ANA = (+) (1/320)
CPK = 299 (NL 24-170)
C3 ↑ C4 ↑ CH50 ↑
RF (-)
C-ANCA (-)
P-ANCA (-)
Anti ds-DNA (-)
Anti CCP (-)
Covid 19 IgM (-), IgG (-)
Anti Cardiolipin IgM (-), IgG (-)
Anti B2 Glycoprotein IgM (-), IgG (-)

Sinus CT scan: No lesions were seen on the left side except for septal perforation and middle turbinate (MT) concha bullosa (Figure 4).

Nasal endoscopy: No lesion was seen other than perforation.

The result of a nasal biopsy:
Nonspecific ulcer, granulation tissue and foci of acute vasculitis

The result of a skin biopsy:
Acute folliculitis
A normal lung CT scan was reported.

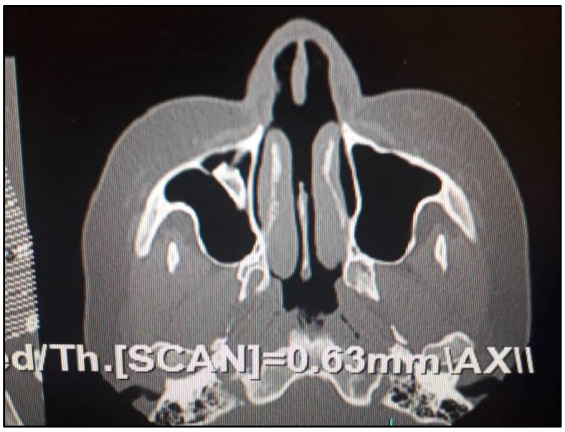

Figure 4. Nasal septal perforation in coronary paranasal CT scan.

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Finally, despite 5 out of 11 criteria, including: 1) Malar rash. 2) Hemolytic anemia. 3) Photosensitivity 4) ANA (+), and 5) Oral ulcers, corticosteroid treatment started for the patient with the diagnosis of SLE. She was then discharged with oral corticosteroids (30 mg daily) and methotrexate (15 mg weekly). After discharge, the patients had only two mild episodes of epistaxis in two months.

Discussion

Systemic lupus erythematosus is a disease that affects various organs and has a wide variety of manifestations. In each manifestation, patient may have more system involvement and the patient presents with the manifestation of involvement of the same system.

Vasculitis-induced septum perforation is a rare manifestation in lupus that often occurs during the disease and during periods of exacerbation and recovery in the known disease. According to precious articles, the history of perforation in rheumatologic diseases dates back to more than 50 years. For example, a study was performed on 12 patients in 1975. Three of patients had lupus, four had rheumatoid arthritis, and other had rheumatologic diseases. The above study ultimately concluded that septal perforation could occur in these diseases with or without epistaxis, and most of these diseases were diagnosed (3). However, our case was an unknown patient who had referred to a doctor with severe epistaxis and septal perforation, and finally, the patient was diagnosed with lupus after further examinations. In other articles, progressive septum perforation is reported in known rheumatologic diseases, sometimes with pain, epistaxis and septal perforation (3).

Conclusion

Systemic lupus erythematosus is a disease with various manifestations that can be easily ignored if not undergoes differential diagnosis diagnosed. Although even an unusual symptom such as septal perforation, especially in young women with no history of trauma or history of drug or substance abuse could put Wegner at the top of our list of differential diagnoses, with this case in mind, the importance of a clinical examination, laboratory examination, and good response to corticosteroids suggests other diagnoses, including lupus and churg-strauss syndrome.

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Patient Consent

Informed consent for publication was obtained from the patient

Conflict of Interest

The authors declared no conflicts of interest.

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