Nasal Septal Perforation as The Initial Presentation for Systemic Lupus Erythematosus: A Case Report and Review of The Literature

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ABSTRACT

Background: Systemic lupus erythematosus (SLE) is a multisystem autoimmune disease with diverse clinical manifestations. Nasal septal perforation is a rare complication, typically occurring during active disease phases or in established cases. Its presentation as the initial symptom is exceedingly uncommon.

Case Presentation: Authors report a 35-year-old male presenting with recurrent mild nasal bleeding, nasal discomfort, fullness, and intermittent nasal obstruction. Nasal endoscopy revealed a large septal perforation. The patient also exhibited joint pain, stiffness, a mild butterfly rash, photosensitivity, and malaise that preceded nasal symptoms. Laboratory tests showed strongly positive antinuclear antibody (ANA) and IgM anticardiolipin antibodies, with normal urinalysis. Histopathology of nasal mucosa indicated mild lymphomononuclear infiltrate. Based on clinical and laboratory findings, diagnosis of Systemic Lupus Erythematosus (SLE) was established by the rheumatologist.

Conclusion: This case highlights nasal septal perforation as the initial manifestation of SLE, emphasizing the importance of considering autoimmune aetiologies in unexplained nasal perforations. Early recognition can facilitate prompt diagnosis and management of systemic disease.

Keywords: SLE, Nasal septal perforation, Epistaxis, ANA, Butterfly facial rash.

ثقب الحاجز الانفي كاول علامة سريرية على مرض داء الذئبة الحمراء: تسجيل حالة مع مراجعة المصادر

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الخلاصة

مقدمة: الذئبة الحمراء او الذئبة الحمراء الجهازية هي مرض مناعي ذاتي مزمن، يحدث عندما يهاجم جهاز المناعة انسجة واعضاء الجسم السليمة عن طريق الخطأ. من النادر جدا ان يكون ثقب الحاجز الانفي هو العارض السريري لمرض الذئبة الحمراء.

تسجيل حالة: رجل يبلغ من العر ٣٥ سنة اشتكى من رعاف، الم في الانف مع انسداد التنفس من الانف بين حين واخر. الفحص بمنظار الانف اظهر وجود ثقب في الحاجز الانفي مع احمرار على شكل فراشة على الوجه. عند مراجعة تاريخ المرض تبين ان المريض كان لديه الم مع تورم المفاصل. التحليلات المختبرية اثبتت ان المريض كان مصابا بداء الذئبة الحمراء وغير مشخص سابقا.

الخلاصة: قمنا بتسجيل حالة مرضية كان فيها ثقب الحاجز الانفي هو العلامة السريرية الوحيدة على مرض داء الذئبة الحمراء قبل تشخيص المرض.

الكلمات المفتاحية: الذئبة الحمراء، الذئبة الحمراء الجهازية، ثقب الحاجز الانفي.

INTRODUCTION

Systemic lupus erythematosus (SLE) is a complex autoimmune disorder characterized by the immune system mistakenly targets and damages the body's own healthy tissues. leading to widespread inflammation and organ damage. It can involve multiple organs, including joints, skin, kidneys, lungs, heart, brain, and blood vessels. The most common initial symptoms include joint pain, skin rashes, and mucous membrane ulcers. Nasal septal perforation is a rare manifestation, often associated with vasculitis or chronic inflammation, and typically occurs during active disease phases. Its presentation as the initial symptom is extremely uncommon, with few cases documented in the literature [1,2].

CASE PRESENTATION

A 35-year-old male presented to our ENT clinic with recurrent mild nasal bleeding, nasal discomfort, fullness, and intermittent nasal obstruction over several months. He denied recent trauma, nasal surgery, nose picking, or exposure to nasal irritants. He was a non-smoker and had no history of drug abuse.

On nasal endoscopy, a sizable perforation was observed in the cartilaginous septum, affecting both anterior and posterior regions, with crusting along the margins (Figure 1). No other abnormalities were noted on ear and throat examinations.

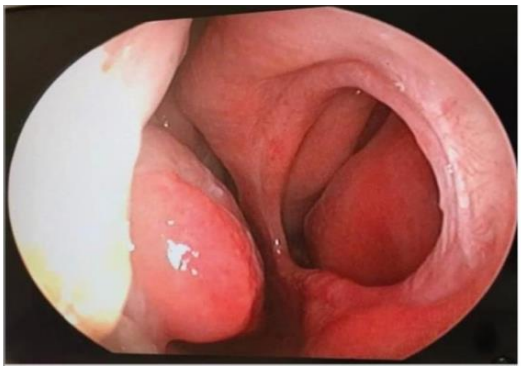

Figure 1: Nasal septal perforation

On direct questioning, the patient reported a history of joint pain and stiffness involving small joints of the hands (proximal interphalangeal, metacarpophalangeal joints) and wrists, along with mild puffiness. He also experienced photosensitivity, fatigue, and a vague malaise that predated nasal symptoms. Physical examination revealed mild tenderness over affected joints and a faint butterfly rash on the face, more prominent after sun exposure (Figure 2). No clinical swelling was observed.

Figure 2: Butterfly facial rash sparing the nasolabial fold

Laboratory investigations showed hypochromic microcytic anaemia (Hb=9.6 g/dL). Autoimmune screening revealed strongly positive ANA titres and positive IgM anticardiolipin antibodies. Urinalysis was normal. C-reactive protein and ESR were mildly elevated. ANCA was negative. A nasal mucosa biopsy demonstrated mild lymphomononuclear infiltrate and epithelial irregularities.

Based on the clinical features and laboratory findings, the patient fulfilled the EULAR/ACR classification criteria for SLE, with a total score of 14 points (ANA positivity: 6 points; joint involvement: 6 points; malar rash: 2 points; anticardiolipin antibodies: 2 points). Table 1

Table 1: EULAR /ACR classification criteria

Entry criterion
Antinuclear antibodies (ANA) at a titer of ≥1:80 on HEp-2 cells or an equivalent positive test (ever)

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If absent, do not classify as SLE
If present, apply additive criteria

↓

Additive criteria
Do not count a criterion if there is a more likely explanation than SLE.
Occurrence of a criterion on at least one occasion is sufficient.
SLE classification requires at least one clinical criterion and ≥10 points.
Criteria need not occur simultaneously.
Within each domain, only the highest weighted criterion is counted toward the total score§.

Clinical domains and criteria	Weight	Immunology domains and criteria	Weight
Constitutional		***Antiphospholipid antibodies***	
Fever	2	Anti-cardiolipin antibodies OR	
Hematologic		Anti-β2GP1 antibodies OR	
Leukopenia	3	Lupus anticoagulant	2
Thrombocytopenia	4	***Complement proteins***	
Autoimmune hemolysis	4	Low C3 OR low C4	3
Neuropsychiatric		Low C3 AND low C4	4
Delirium	2	***SLE-specific antibodies***	
Psychosis	3	Anti-dsDNA antibody* OR	
Seizure	5	Anti-Smith antibody	6
Mucocutaneous			
Non-scarring alopecia	2		
Oral ulcers	2		
Subacute cutaneous OR discoid lupus	4		
Acute cutaneous lupus	6		
Serosal			
Pleural or pericardial effusion	5		
Acute pericarditis	6		
Musculoskeletal			
Joint involvement	6		
Renal			
Proteinuria >0.5g/24h	4		
Renal biopsy Class II or V lupus nephritis	8		
Renal biopsy Class III or IV lupus nephritis	10		

Total score:

↓

Classify as Systemic Lupus Erythematosus with a score of 10 or more if entry criterion fulfilled.

2019 EULAR/ACR Criteria. From Aringer et al. Arthritis Rheumatol. 2019. PMID: 31385462

DISCUSSION

Systemic lupus erythematosus (SLE) is widely recognised as a disease that can attack a wide variety of tissues and organs, and hence the widespread symptoms and signs and various abnormal laboratory tests. It's a widespread inflammatory process that, if untreated, can lead to tissue damage in the affected organs [3-5]. SLE can affect the joints, skin, kidneys, bone marrow, lungs, heart, brain, and blood vessels. However, nasal septum involvement is very rare and in general, it occurs during the phase of the active disease or its exacerbations. It is much rarer for nasal septal perforation to be the initial presenting symptom. This may be due to vasculitis or ischaemia with chondritis [6].

Our case has strongly positive ANA, positive IgM anticardiolipin Abs, with arthritis malar rash and photosensitivity, which all point to SLE. Our case fulfilled the EULAR/ACR classification criteria as his score was 14 points; the entry criteria are the strongly positive ANA, and he had 6 points for the joints, 6 points for his rash and 2 points for the positive Anticardiolipin Abs [7].

SLE may be difficult to diagnose because its early signs and symptoms are not specific and can look like signs and symptoms of many other diseases [8]. These manifestations could also occur and develop over time, and hence, SLE patients may not initially fit the classification criteria, but with time, this will become clearer. For instance, we know that a patient with solely renal involvement may have the full-house criteria on renal biopsy but has few or no other criteria for SLE.

Authors know that almost all patients with SLE have a positive ANA test, making it the most sensitive diagnostic test required for entering the EULAR/ACR classification criteria [7,9]. Between 70 and 80% of patients develop skin lesions during the disease course [7,10]. Approximately 20% have skin lesions as an initial presentation. The butterfly rash across the nose occurs in about 30% of patients with SLE [10], and it is present in our patient, but was discovered later after his initial presentation with the Nasal septal perforation.

Primary vasculitis could cause a nasal septal defect; however, The present case was negative for antineutrophil cytoplasm antibodies C-ANCA and had no other clinical or lab criteria to suggest that. Interestingly, positive C-ANCA is known to occur in some patients with systemic lupus erythematosus (SLE), with a prevalence of 15 to 20 % [11]. It points to vasculitis, which could explain the nasal septum perforation in some of these cases, but this was not the scenario in our case.

Nasal septal perforation, a communication between the nasal passageways [12, 13], is usually discovered incidentally during clinical or radiological examination. Table 2[(2)] shows a broad variety of presumed aetiologies have been described, making the diagnostic approach heterogeneous.

Table 2: Etiologies of nasal septal perforation

Etiology	Examples
Trauma	Nasal injuries Nose picking Surgical complications
Nasal Surgery	Septoplasty Cautery Submucous resection Transseptal approach Excision of nasal tumor
Chronic Nasal irritation	Nasal sprays Decongestant, corticosteroids Drug abuse
Autoimmune diseases	Granulomatosis with polyangiitis (GPA), lupus, and rheumatoid arthritis
Infectious	Tuberculosis and syphilis
Tumors	Polymorphic reticulosis T cell Lymphoma Acute myelocytic leukemia Wegener's granulomatosi
Chemical exposure	Chrome Zinc
Idiopathic	3%

A good history and examination, and a wide variety of blood tests, as suggested by the clinical picture, are important to point to the possible diagnosis. This should be followed by anterior rhinoscopy, and nasal fibroscopic exams with a biopsy of the lesion, which are the main means to the correct diagnosis and subsequent therapeutic management[13]. In our case, the development of the typical malar rash and the photosensitivity prompted us to do the ANA and the other investigation and hence the diagnosis of SLE.

Nasal septum perforation is an uncommon feature of Systemic lupus erythematosus (SLE). In general, it occurs during the exacerbations of the disease in patients with well-established SLE. This most likely occurs in the context of systemic vasculitis [14,15]. Very rarely, it can be a presenting sign, accompanying more usual manifestations of LE and in our extensive journal review, we only found one previous case [15].

Additional details from the literature: The perforations are most likely to occur during SLE exacerbations, months or years after diagnosis. Epistaxis (nosebleeds) is a common initial symptom. A frequent association with Raynaud's phenomenon is noted, suggesting potential local vasospasms due to cold inspired air as a contributing factor. The perforations tend to increase in size, often independently of medication and are frequently managed conservatively. Some literature suggests that nasal septum perforation may be an early sign of an autoimmune disorder or a known disease with incomplete penetrance. A review of 140 cases of autoimmune-associated nasal septum perforations identified granulomatosis with polyangiitis (48%), relapsing polychondritis (26%), and cocaine-induced midline destructive lesions (15%) as more frequent causes.[١٦]

Conclusion

This case is unique in that the nasal septum perforation was the presenting sign for SLE, and hence we need to draw the attention of local ENT specialists and other physicians to have a high index of suspicion and to consider SLE as a possible diagnosis for all unexplained isolated nasal septal perforations.

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Conflict of interest

None.

Ethical Approval

The study was approved by the Collegiate Committee for Medical Research Ethics with approval code UOM/COM/MREC/24-25/APR27 on 9.04.2025.

Authors contribution

The patient whose case report was studied presented to the second author in his private clinic. The first author wrote the main manuscript text and share in reviewing the literature. The third and fourth authors shared in proofreading and reviewing the manuscript.

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