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Clinical Trials and Case Studies

Abdul-Wahab Al-Allaf *

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Case Report

Nasal Septal Perforation: First Presentation for Systemic Lupus Erythematosus

Zakaryia A. Younus [1], Omar Al-Allaf [2], Abdulmohsin Y. Saleem [3], Abdul-Wahab Al-Allaf [4*]

[1] Emergency Department, Gaziantep University Hospital, Gaziantep, Turkey,

[2] Walsall Manor Hospital, Moat Road, WS2 9PS, UK.

[3] Early Detection Childhood Disabilities Center in Dohuk, Iraq.

[4] Consultant Rheumatology, Hamad Medical Corporation.

***Correspondence Author:** Abdul-Wahab Al-Allaf, Consultant Rheumatology, Hamad Medical Corporation.

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Abstract

Systemic lupus erythematosus (SLE), is an autoimmune disease in which the immune system attacks its tissues, causing widespread inflammation and tissue damage in the affected organs. It can affect the joints, skin, brain, lungs, kidneys, and blood vessels. SLE's commonest presenting symptoms are joint pain and swelling with stiffness, skin rash, recurrent multiple mouth ulcers other organ involvement such as renal, haematological and serositis.

Nasal septum perforation is a very rare manifestation of SLE. It usually occurs in a well-diagnosed case of SLE and mostly is an underdiagnosed complication of lupus because it is asymptomatic, and patients are often not aware of their nasal problem (1 Reiter D, Myers AR. Asymptomatic nasal septum perforation in Systemic Lupus Erythematosus. Ann Otol. 1980; 89:78-80). Here we are reporting a patient with a nasal septal perforation to be the first and sole clinical presenting symptoms for SLE for almost a year before the diagnosis of SLE was confirmed. This is an even much rarer scenario.Nasal Septal perforation in lupus may be secondary to vasculitis or ischaemia with subsequent chondritis (2. Rahman P, Gladman D, Urowitz M. Nasal septum peroration in Systemic Lupus Erythematosus - Time for a closer look. J Rheumatol. 1999; 26:1854-5).

Keywords: sle; nasal septal perforation; epistaxis; ana; butterfly facial rash

Clinical presentation

A 35-year-old male visited the ENT consultation clinic, complaining of recurrent mild nasal bleeding, discomfort at the nasal cavity, fullness in the nose and nasal on and off nasal obstruction. There was no history of previous local trauma, surgery, application of topical agents, or environmental exposure to metals. He did not smoke or take any medication or drugs.

Nasal endoscopy showed a large septal perforation involving the anteroinferior and anterosuperior parts of the cartilaginous nasal septum with crusting over the edges of the septal perforation (Figure 1).

Figure 1: Nasal Septal Perforation.

The crusts were endoscopically cleared. An ear and throat examination were done, and no other significant abnormality was detected.

He has a history of joint pain and stiffness with mild puffiness of the small joints of his hands including PIPs, MCPs and wrists. He also reported some photosensitivity, tiredness (fatigue), and a vague feeling of discomfort or Illness (malaise). All these symptoms were there for some time before his initial presentation with the nasal symptoms. On examination he has mild tenderness over the above-mentioned joints, but without clinical swelling. He also has a mild butterfly rash (figure 2). He reports this to be more pronounced on exposure to sunlight.

Laboratory investigation revealed mild hypochromic microcytic anaemia, with strongly positive ANA and a Positive IgM anticardiolipin antibody with normal urinalysis. ANCA was negative.

A nasal septum mucosa biopsy was performed, and a histological examination revealed a mild lymphomononuclear cell Infiltrate of the corium and Irregular acanthosis of the epithelium.

Figure 2: Butterfly Rash

Discussion

Systemic lupus erythematosus (SLE) is an autoimmune disease in which the immune system can attack a wide variety of various tissues and organs and hence the widespread symptoms and signs and various abnormal laboratory tests. It is usually resulting in widespread inflammation and tissue damage in the affected organs [1-3]. It can affect the joints, skin, brain, lungs, kidneys, heart, and blood vessels. However, nasal septum involvement is very rare. In general, it occurs during exacerbations and in the context of systemic vasculitis. It is even much rare for nasal septal perforation to be the initial presenting symptom.

Our case has strongly positive ANA, positive IgM anticardiolipin Abs, with arthritis and Malar rash and photosensitivity which all points to SLE. However, he does not fulfil the EULAR/ACR diagnostic criteria as his score was 8 (6 points for the joints and 2 points for the positive Anticardiolipin Abs) and not 10, which is needed for the definite classification for SLE. (Aringer M, et al. 2019 European League Against Rheumatism/American College of Rheumatology classification criteria for systemic lupus erythematosus. *Arthritis Rheumatol*. 2019 Sep;71(9):1400–1412)

About half of all lupus patients experienced a characteristic rash called the malar or "butterfly" rash that may occur spontaneously or after exposure to the sun. This rash is so named because it resembles a butterfly, spanning the width of the face and covering both cheeks and the bridge of the nose. [1-4].

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 [4].

Ninety-Eight per cent of people with systemic lupus have a positive ANA test, making it the most sensitive diagnostic test for the entering criteria to diagnose the disease [5].

Antineutrophil cytoplasm antibodies (ANCA) are known to occur in some patients with systemic lupus erythematosus (SLE), with a prevalence of 15 to 20 % [6]. It points to vasculitis which could explain the nasal septum perforation. However, this was negative in our case

Between 70 and 80% of patients develop skin lesions during the course of the disease [4-7]. Approximately 20% have skin lesions as an initial presentation. The butterfly rash across the nose occurs in about 30% of patients with SLE [7]. and it is present in our patient.s

Nasal septal perforation, a communication between the nasal passageways [8, 9]. is usually discovered incidentally during clinical or radiological examination. A broad variety of presumed etiologies have been described, making the diagnostic approach heterogeneous (Table 1).

Traumatic /Iatrogenic	Septal surgery, Chemical cautery, Nasal Packing Nasogastric probe, Prolong nasal intubation Nasal fracture, foreign body, Digital manipulation
Nasal Drugs Abuse	Decongestant nasal, Nasal corticosteroids Cocaine
Occupational Exposure	Chromium fumes Powder glass
Inflammatory	Wegener's granuloma, Relapsing poly-chondritis (+++), Sarcoidosis, Systemic lupus erythematosus (rare), Rheumatoid Arthritis Crohn's disease (very rare), Dermatomyositis, IgG4 disease (very rare),

	VEXAS (++++)
Infectious	Syphilis, HIV, Fungal infection, Leprosy, Tuberculosis Leishmaniasis, Rhino sarcoma
Neoplasm	Carcinoma, Lymphoma
Idiopathic	?

Table 1: Aetiology of septal perforation

The aetiology of septal perforation should be sought in all patients. A complete history was followed by anterior rhinoscopy, and nasal fibroscopic exams with a biopsy of the lesion, which are the main means to correct the diagnosis and subsequent therapeutic management [9].

Nasal septum perforation is an uncommon feature of Systemic lupus erythematosus (SLE). In general, it occurs during exacerbations and in the context of systemic vasculitis [10,11]. Very rarely it can be a presenting sign, accompanying more usual manifestations of LE [11].

Our case is unique in that the nasal septum perforation was the presenting sign for SLE and hence we need to draw the attention of our ENT specialists and other physician to consider SLE as a possible diagnosis for all unexplained isolated nasal septal perforation

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