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Case Report

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Case report: a rare presentation of nasal septal perforation due to pyoderma gangrenosum

Abstract

Background: Pyoderma gangrenosum (PG) is a rare dermatologic non-infectious neutrophilic disease that classically affects the lower extremities and is associated with inflammatory bowel disease. Rarely, it could affect the nasal septum, causing nasal septal perforation.

Methods: We reviewed the case of a 52-year-old male patient known to have PG with ongoing nasal septal perforation and reviewed his blood tests, computed tomography scan findings and histologic results.

Results: A diagnosis of nasal septal perforation due to PG was confirmed after exclusion of other common aetiologies. This was further supported by the presence of extensive ulceration of the nasal squamous mucosa with inflamed granulation tissue and abscess-like areas. These findings were consistent with the diagnosis of PG based on the histology report.

Conclusion: Although nasal septal perforation secondary to PG is considered rare, this presentation should still be kept in mind, especially when other possible causes of nasal septal perforation have been excluded and PG has been already well established.

Keywords: nasal septal perforation, neutrophilic dermatosis, pyoderma gangrenosum

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Introduction

Pyoderma gangrenosum (PG) is a reactive non-infectious inflammatory dermatosis falling under the neutrophilic dermatosis spectrum, which includes Sweet's syndrome and Behcet's syndrome.[1,2] It was first described in 1930 by Drs. Brunsting, Goeckerman, and O'Leary, who described the typical lesions of PG as enlarging necrotic ulcers with erythematous to bluish undermined borders surrounded by spreading erythema.[3,4]

Six major variants of the skin condition have since been outlined, including (1) ulcerative or classic, (2) pustular, (3) bullous or atypical, (4) vegetative, (5) peristomal, and (6) post-surgical PG (Table 1).[3]

Case report

A 52-year-old male patient was recently diagnosed with the vegetative type of PG as he only had skin lesions in the back without any other systemic manifestations. After the diagnosis, he developed several non-specific nasal issues that were progressive in nature. These included nasal stiffness, nasal obstruction, crustation, and a dull ache.

Previously, he had tried different remedies, either over-the-counter medications or those prescribed by his primary health care providers. These drugs included saline nasal rinses, local steroid sprays[5], and local antibiotics; however, despite the use of these medications, the patient's nasal symptoms continued to deteriorate. Therefore, he was referred to the otolaryngology specialized clinic.

On examination at the clinic, nasal bridge broadening was noted with extreme tenderness at the alar cartilage area. Nasal endoscopy was performed that revealed nearly total nasal septal perforation as well as erosion of the anterior sphenoidal wall. Moreover, there was widespread granulation tissue inside the nasal cavity.

A thorough and detailed approach was used to determine the cause of the septal perforation. There was no history of nasal trauma or surgery, cocaine abuse, or any granulomatous[6] diseases apart from PG. Various lab and imaging tests such as cytoplasmic and peripheral antineutrophil cytoplasmic antibodies (c-ANCA and p-ANCA), erythrocyte sedimentation rate (ESR), full blood count (FBC), and chest X-ray (CXR), among others, were ordered in order to arrive at a definitive diagnosis. The results of all these tests were inconclusive. In addition, a computed tomography (CT) scan was ordered, and the results were consistent with the clinical findings (Figure 1).

At that stage, it was prudent to exclude malignancy; therefore, multiple biopsies were taken under general anaesthesia (Figure 2).

The cytology assessment detected no malignant cells; however, there was noted extensive nasal squamous mucosa ulceration with inflamed granulation tissue and abscess-like areas consistent with PG.

Discussion

Nasal septal perforation is considered reasonably common in otolaryngology consultations. The aetiologies vary greatly from one another, and there is a myriad of possibilities that could cause them. These can be summarized into the categories listed in Table 2.

Because of the wide range of aetiologies for septal perforation, history-taking is crucial. Nasal perforations may be the first sign of various granulomatous disorders, including granulomatosis with polyangiitis (GPA) and systemic lupus erythematosus (SLE). Baseline work-up should include an FBC, ESR, serum urea, serum electrolytes,[7–9] urinalysis, C-ANCA, treponemal tests, ACE titres, a CXR, and a nasal swab. Routine biopsy of septal perforations to exclude vasculitis has been suggested[10–12] but biopsy rarely reveals anything other than chronic inflammation and is usually insufficient for a specific diagnosis.

Nasal septal perforation due to PG is considered rare; therefore, this diagnosis remains a diagnosis of exclusion.[3] Head and neck manifestations of PG are known to have poor response to standard treatment.[13] The patient mentioned in this study had already received extensive steroid therapy to treat his skin lesions, and yet his lesions still progressed until finally his nasal structures were destroyed.

Table 1 Different clinical presentations of pyoderma gangrenosum and their associated systemic diseases

Pyoderma gangrenosum variant	Common location	Presentation	Associated disease
Ulcerative (classic)	Lower extremities	Rapid progression Violaceous undermined border Very painful	Inflammatory bowel disease (IBD) Arthritis Myeloproliferative disease
Bullous (atypical)	Face	Superficial bulla Blue-grey border	Myeloproliferative disease (i.e., acute myeloid leukaemia)
Pustular	Legs Upper trunk	Painful pustules Red halo	IBD
Vegetative	Trunk	Superficial ulcer No violaceous border	None
Peristomal	Near stoma site	Painful ulcer Violaceous undermined border	IBD Enteric malignancies
Post-surgical (procedural) (e.g., after nipple piercing)	Surgery site (breast, abdomen most common)	Rapid progression Active and undermined border Pain out of proportion to lesion	Fewer cases of underlying systemic disease (compared with classic form)

Table 2 Aetiologies of the nasal septal perforation

Traumatic causes	Surface irritants	Infections	Neoplastic	Inflammatory
Nasal surgery	Cocaine insufflation	Syphilis	Melanoma	Granulomatosis with
Nose picking	Fumes (chromic/sulphuric acid)	Tuberculosis	Squamous cell carcinoma	polyangiitis
Bilateral septal cauterization		Lepromatous leprosy	Adenocarcinoma	Sarcoidosis
Foreign bodies		Rhinoscleroderma Mucor	Lymphoma	Systemic lupus erythematosus

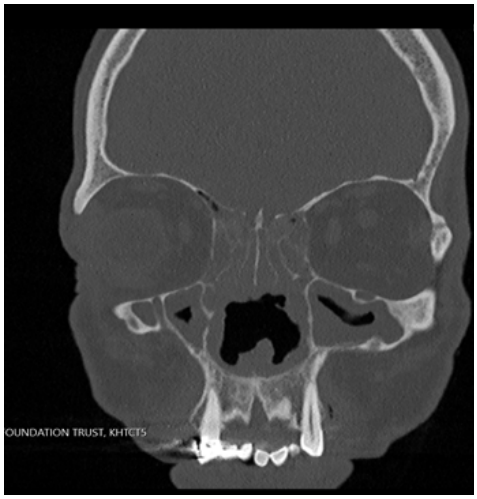

Figure 1 A coronal CT scan of the patients nose and paranasal sinuses.

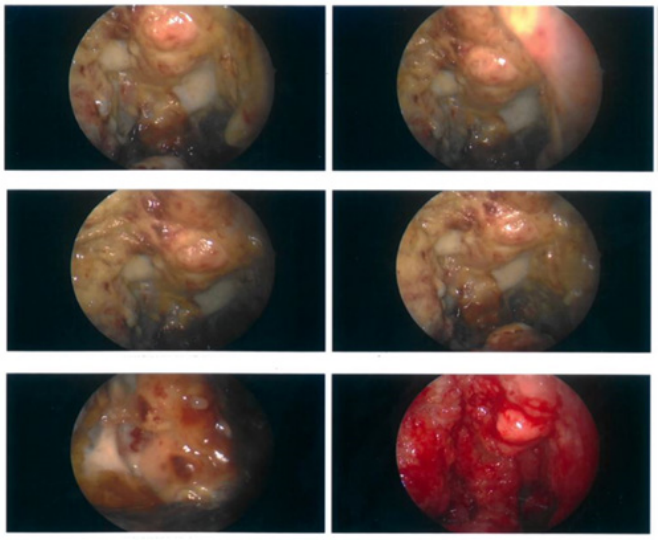

Figure 2 Intraoperative view of the nose.

Conclusion

Although nasal septal perforation secondary to PG is considered rare, this presentation should still be kept in mind especially when other possible causes of nasal septal perforation have been excluded and PG is already well established.

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Conflicts of interests

Authors declare no conflict of interests.

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