A Rare Case Study of Behcet Associated with Pyoderma Gangrenosum

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Abstract

Behcet's disease is a rare, chronic, recurrent disorder characterized by inflammation of blood vessels involving the entire body. We present a case of a 45-year-old male patient with non-healing ulcers on the left lower limb, scrotal ulcer, and oral ulcer for seven years. Evaluation for Behcet's disease was performed, and the patient was found to have Behcet's pyoderma gangrenosum. Despite treatment with immunosuppressive therapy and anti-inflammatory agents, the symptoms persisted. The primary goal of management in Behcet's disease is to induce and maintain remission, improve quality of life, and prevent irreversible damage. However, the choice of the best treatment is challenging, and there still exists a gap in the treatment of Behcet's disease despite several research advancements.

Keywords

Behcet's disease, non-healing ulcers, pyoderma gangrenosum, immunosuppressive therapy, anti-inflammatory agents.

1.Introduction

Behcet's disease, also known as Behcet's syndrome, is a disorder of uncertain origin that causes inflammation in multiple bodily systems. It is characterized by inflammation in blood vessels throughout the body [1]. This disorder can give rise to a range of symptoms that may initially appear unrelated, including mouth ulcers, skin rashes and lesions, genital sores, and inflammation of the eyes. Similarly, pyoderma gangrenosum is a type of cutaneous vasculitis that is characterized by an accumulation of neutrophils and mononuclear cells at the site of the reaction [2]. Primary pustular lesions can develop in both Behcet's disease and pyoderma gangrenosum, and neither condition has a distinctive histological appearance.

2.Epidemiology

The estimated occurrence of this condition is relatively low, ranging from 1 in 1,000 to 1 in 10,000 cases along the Old Silk Route. However, the reported prevalence of this condition is notably higher in Asian countries, ranging from 13.5 to 20 cases per 100,000 people [3].

3.Case report

Behcet's disease, also known as Behcet's syndrome, is a rare, chronic, and recurrent disorder characterized by inflammation of blood vessels that affects the entire body. The disease presents various symptoms such as mouth sores, skin rashes and lesions, eye inflammation, and genital sores. In this case report, we present the clinical features and treatment course of a 45-year-old male patient who presented with a non-healing ulcer on the left lower limb, scrotal ulcer, and oral ulcer persisting for 7 years.

Upon examination, the patient had oral pigmentation over the palate with a healed ulcer, multiple split skin graft wounds due to resulting graft failures, and a leg ulcer measuring 2x15 cm occupying two-thirds of the leg. The patient was admitted to the rheumatology ward for further evaluation, and it was found that he was HLAB51 negative [3]. However, an ulcer biopsy revealed Behcet's pyoderma gangrenosum and culture from the swab taken from wounds showed Pseudomonas and Klebsiella, while the swab from the leg ulcer showed Acinetobacter and E. coli.

Based on the culture sensitivity report, the patient was administered Tab. Linezolid 600 mg, Tab. Cephalexin 250mg, Inj. Meropenem 500 mg, in addition to anti-inflammatory agents of Tab. Prednisolone 30mg and Tab. Tofacitinib 5mg. The patient received immunosuppressive therapy of Inj. Adalimumab 30mg (3 doses) due to severe muco-cutaneous manifestations [4]. However, despite treatment, the patient's symptoms continued to persist.

Table 1: Drugs Prescribed

DRUGS	DOSE	FREQUENCY	TREATING INDICATIONS
T. LINEZOLID	600mg	BD	Infection
T. CEPHALEXIN	250mg	Q6H	Infection
INJ.MEROPENEM	500mg	BD	Infection
T. PREDNISOLONE	30mg	OD	Anti-inflammatory
T. TOFACITINIB	5mg	BD	Ulceration in skin
INJ.ADALIMUMAB	30mg	Weekly once	Pyoderma Gangenosum

4.Discussion

This is a description of pyoderma gangrenosum, a rare skin disease characterized by the breakdown of a nodule or pustule, leading to the formation of a progressively enlarging ulcer [5]. About 50% of patients with this condition also have a related systemic disease, including inflammatory bowel disease, arthritis, and monoclonal gammopathy [6]. Behcet's disease is diagnosed by the hallmarks of mucocutaneous lesions [7], involving oral, genital and conjunctival mucosa [8,9]. The primary goal of managing this disease is to induce and maintain remission, improve quality of life, and prevent irreversible damage, and exacerbation of mucocutaneous and articular disease. Immunosuppressive agents form the mainstay of treatment in severe, life-threatening, and symptomatic manifestations. Treatment options include local preparations of fluorinated corticosteroids, nitric silver digestion, local anaesthetics, systemic preparations of corticosteroids, sulfonate, gamaglobulin, and large doses of vitamin C. However, the results of treatment may still be slow and incomplete.

5.Conclusion

This case report highlights the challenges in managing Behcet's disease and the need for a personalized treatment approach. The presence of multiple microorganisms in the ulcer cultures and the lack of response to treatment suggest that the case may be more complicated than typical Behcet's disease. Further evaluation and monitoring may be necessary to determine the most effective treatment plan for this patient. Despite several research advancements, there still exists a gap in the treatment of Behcet's disease.

6.References

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