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Rheumatoid Arthritis Presenting with a Non-healing Ulcer, Diagnosed to be Pyoderma Gangrenosum

ABSTRACT

A middle-aged male diagnosed with rheumatoid arthritis presented with an acute large painful ulcer spreading over left elbow following an abscess drainage. Laboratory values showed normal procalcitonin, high C-reactive proteins, and sterile wound culture along with skin biopsy. The initial approach included antibiotic management but the ulcer proliferated despite parenteral antibiotics and regular dressing with a subsequent histopathological diagnosis of pyoderma gangrenosum. The patient improved with steroid and immunosuppressant drugs.

Key words: Dexamethasone and immunosuppressants, Neutrophilic dermatoses, Non-healing ulcer, Rheumatoid arthritis, Skin biopsy

INTRODUCTION

Pyoderma gangrenosum (PG) presents as a rapidly enlarging, very painful ulcer. It is one of a group of autoinflammatory disorders known as neutrophilic dermatoses. Pyoderma gangrenosum is an autoinflammatory disease (excessive response to an internal antigen) due to some form of neutrophil dysfunction. T lymphocytes and cytokines are involved. There may be a genetic predisposition. Pyoderma gangrenosum is a rare disease that affects males and females of any age but is more common in those aged over 50 years. It frequently is associated with an internal disease or condition. Its known associations include:

- Rheumatoid arthritis
- Inflammatory bowel disease (ulcerative colitis and Crohn disease)
- Myeloid blood dyscrasias including leukaemia
- Monoclonal gammopathy (usually IgA)
- Chronic active hepatitisGranulomatosis with polyangiitis
- PAPA syndrome
    - Behçet disease

CASE REPORT

A 56-year-old male patient presented with complaints of a large, non-healing ulcer over the left elbow joint, along with fever for the past 3 days. He was apparently alright 7 days ago when he noticed swelling and redness over the left elbow. He also started developing a fever in the past 3 days. The fever was of high grade, not associated with chills and rigors, and partially relieved by oral paracetamol. He was presumptively diagnosed to have cellulitis and underwent fasciotomy for the same. However, after the procedure, the patient reports that the incision site quickly developed into an ulcer. The ulcer kept increasing in size despite adequate dressing and administration of intravenous antibiotics (Ceftriaxone and Gentamicin) at a local hospital. However, as the above therapy was not providing any relief, the patient presented to our hospital for further evaluation and treatment.

On general examination, patient febrile with temperature 99.8 F, pulse 96/minute, regular, no radioraial or radio femoral delay, all peripheral pulses are well felt and pulse oximetry 98% on room air with Blood pressure 130/70 mm Hg in right upper limb in supine position. Patient was also found to have mild bilateral pedal edema which was pitting in nature. There was no pallor/icterus/clubbing/ lymphadenopathy.

On systemic examination, no significant abnormalities were detected in any of the systems. Local examination shows a solitary, large, and painful ulcer around the left elbow. The ulcer was advancing, erythematous, and had inflamed borders, along with a poor a healing base [Figure 1]. All routine blood investigations were performed, which revealed the initial high levels of white blood cells, high levels of C-reactive proteins, and positive rheumatoid factor levels while the other routine investigations were within normal limits [Table 1].

The patient was started on injectable antibiotics, clindamycin 600 mg thrice a day and cefoperazone 1000 mg

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Table 1: Investigation

Investigation	3/7/22	4/7/22	5/7/22	6/7/22	7/7/22	9/7/22	11/7/22	18/7/22	26/7/22	16/8/22
Hb (g/dL)	12	12.6		12.6	13	13.8	14.3	13.8	14.8	14.7
WBC count (/cmm)	36,880	34,060		25,760	17,810	18,680	16,920	16,280	13,930	10,050
Platelet (/cmm)	336K	371K		494K	555K	717K	880K	626K	279K	243K
PT/INR				1.4/1.35						
Total Bilirubin (mg/dL)	2.49			0.738		1.1			0.6	
Direct Bilirubin (mg/dL)	1.89			0.678		0.6			0.3	
Indirect bilirubin (mg/dL)	0.60			0.06		0.5			0.3	
SGOT (U/L)	23.6			37		20			21	
SGPT (U/L)	42.5			40.40		52			46	
GGT (IU/L)	217			151		185			66	
ALP (U/L)	153			144		110.34			68.07	
Total protein (g/dL)	6.23		5.7	5.82		6.50			6.99	
Albumin (g/dL)	2.94		2.33	2.7		3.29			4.18	
Globulin (g/dL)	3.34			3.1		3.21			2.81	
Alpha 1 globulin (g/dL)			0.77							
Alpha 2 globulin (g/dL)			1,00							
Beta 1 globulin (g/dL)			0.25							
Beta 2 Globulin (g/dL)			0.36							
Gamma globulin (g/dL)			1.00							
Myeloma band			Not detected							
Sodium (mEq/L)	123.5	126				129				
Potassium (mEq/L)	4.21	4.25				4.7				
Chloride (mEq/L)	87.21	92.5				98				
Urea (mg/dL)	49.01									
Creatinine (mg/dL)	1.73			0.88		0.76			0.80	
Fasting blood sugar (mg/dL)	119.09									
Random blood sugar (mg/dL)							137	130	107	189
Calcium (mg/dL)	7.54									
Uric acid (mg/dL)	6.80									
CRP (mg/dL)	225			202,60					1.0	
PCT (ng/dL)		1.10		0.438						
Urine culture		NoGrowth								
Rheumatoid factor (IU/mL)			37							
C ANCA (AU/mL)			Negative							
P ANCA (AU/mL)			Negative							

CRP: C-reactive proteins, WBC: White blood cell, PCT: Procalcitonin

plus sulbactam 500 mg twice a day. The ulcer was proliferating despite proper dressing and antibiotics. Since the wound culture was not showing any organism; hence, skin biopsy was done by a dermatologist. It showed mild neutrophilic infiltration without any other pathology. The patient was started on dexamethasone 4 mg 3 times a day along with topical applications of Human Recombinant Epidermal Growth Factor and Mometasone after taking into consideration the clinical findings, those of the antecedent of mutilans rheumatoid arthritis (RA), and histopathological findings that had previously suggested pyoderma gangrenosum (PG). The patient started improving on this regimen, with a decrease in the fever spikes, along with a decrease in white blood cell (WBC) counts. The ulcer margins stopped spreading and

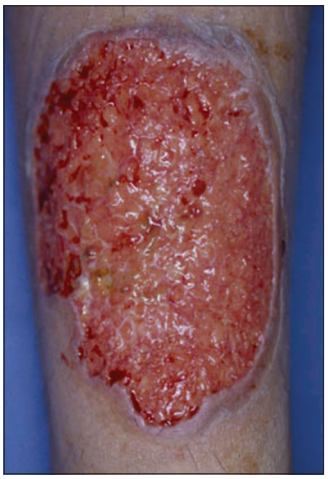

Figure 1: Solitary painful rapid growing superficial ulcer with irregular borders and violaceous edges around the left elbow with advancing, erythematous and inflamed borders, along with a poorly healing base

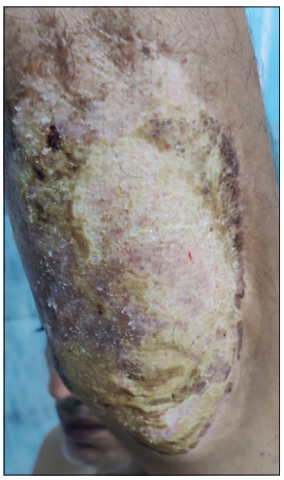

Figure 2: Solitary superficial ulcer showing signs of healing and inflammatory changes subsided after 6 weeks of treatment

inflammation subsided after 6 weeks of treatment and started showing signs of healing [Figure 2]. Blood sugar levels were monitored, and the patient was given subcutaneous insulin based on sugar levels to prevent the high levels of blood sugar levels due to corticosteroid intake. Gradually, the patient stabilized and was discharged. The patient was switched over to oral steroids (prednisolone) on discharge. In view of persistently high WBC counts, he was additionally started on an immunosuppressant, azathioprine 50 mg twice a day, for which he has shown substantial improvement.

DISCUSSION

PG is an chronic, recurrent, idiopathic, and autoinflammatory disease (excessive response to an internal antigen) due to some form of neutrophil dysfunction. It is also known as neutrophilic dermatoses. The median age for the condition is 40–60 years.[1] There is involvement of T lymphocytes cells and that cytokines. 25% of the patients with PG have positive Pathergy.[1] There may be a genetic predisposition which is seen in some patients. PG presents as a rapidly enlarging, painful ulcer.[2] The name PG is a misnomer. The condition is not an infection (pyoderma), nor does it cause gangrene. It is characterized by pathergy (ulcer occurring at sites of trauma, with ulcer extending past area of trauma).

In many people (more than 50%), PG is linked to systemic disease, primarily RA and inflammatory bowel disease. The arthritis-related lesions are frequently ulcerative. These lesions can affect the entire body, even though they normally only affect the lower limbs.[3] When other causes of similar-appearing cutaneous ulcerations are ruled out, the diagnosis is made. These other reasons include infection, cancer, vasculitis, vasculopathy, venous insufficiency, collagen-vascular disorders, diabetes, and trauma.[4]

Topical treatment as well as local and systemic treatments are used to treat PG. The treatment of the ulcers requires daily application of wet dressings with petrolatum (to prevent stress to the underlying tissue during removal) and antibiotic to combat infection. Avoid surgical debridement to prevent further spread of the disease .With AQ5 initial immunosuppressive medication, the majority of individuals with PG experience improvement and require little subsequent maintenance. Alternative therapeutic methods include tacrolimus, thalidomide, infliximab, plasmapheresis, systemic treatment with corticosteroids, and mycophenolate mofetil and ciclosporin, which is rarely used if ineffectively. [5] These medications are typically used in conjunction with corticosteroids to lessen their dosage and try to treat refractory instances. In PG instances that are resistant to other treatments, biological medicines including fusion proteins and anti-TNF-alpha can also be administered.

CONCLUSION

We present a patient with PG who was successfully treated with cortisone and azathioprine for RA. Comprehensive testing ruled out co-existing autoimmune conditions, proving the link between PG and autoimmune conditions such RA. PG can be difficult to diagnose and has fatal effects if left untreated; therefore, it is obvious that the diagnosis should alert doctors to the risk of concurrent RA. On the other hand, RA should alert us to PG's potential future emergence. All things considered, our case contributes to the body of knowledge regarding the manifestation and treatment of PG in connection with RA.

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