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That purplish huge hole on the chest wall

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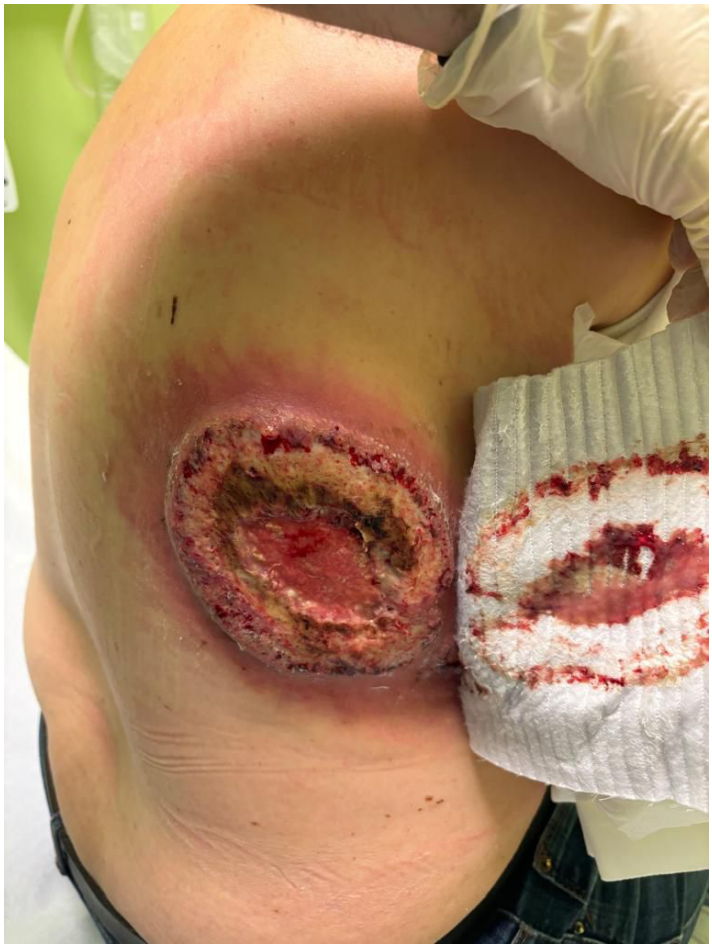
Ethics approval and consent to participate: as this was a descriptive case report and data was collected without patient identifiers, ethics approval was not required under our hospital's Institutional Review Board guidelines.

Informed consent: the patient provided consent for access to medical records at the time of admission.



Descriptive legend

A 54-year-old woman presented to our emergency department two months after the removal by Video-Assisted Thoracoscopic Surgery (VATS) of her middle lung lobe affected by severe inflammation and fibrosis. She was experiencing pain from a large ulcer (diameter 13 cm) starting

from the surgical scar. She suffered from Calcinosis, Raynaud phenomenon, Esophageal dysmotility, Sclerodactyly, and Telangiectasia (CREST) syndrome, and hypothyroidism. The laboratory results showed White Blood Cell count (WBC) 13,600/mm^3 (N 76%), C-Reactive Protein (CRP) 12 mg/L (nv<6), and Erythrocyte Sedimentation Rate (ESR) 86 mm/h (nv<20). Repeat skin swabs were always negative. Skin biopsy documented granulocyte infiltration in the dermis-hypodermis with tissue necrosis. For three weeks, the patient was treated with methylprednisolone 1 mg/Kg IV and colchicine 0.5 mg q12h orally. In the absence of a good response, oral cyclosporine 75 mg q12h was added. The ulcer gradually improved within a week. The pain was successfully treated with paracetamol 1 gr q8h IV.

Question: Given the patient history and the clinical presentation, what is the most likely diagnosis?

1) Pyogenic granuloma
2) Necrotizing fasciitis
3) Pyoderma gangrenosum
4) Martorell ulcer

Answer: The right answer is Pyoderma Gangrenosum (PG). PG is a rare ulcerative disorder that belongs to the category of neutrophilic dermatoses.[1] Despite its name, PG is not caused by infection or gangrene. The etiology is unknown. In 70% of patients, PG is associated with a systemic condition, including ulcerative colitis (5-12%), Crohn's disease (1-2%), solid tumours, and haematologic malignancies (20%).[2] The most common age of presentation is 30-50 years. Women are more commonly affected.[1] The pathogenesis is not fully understood.[3] PG is a diagnosis of exclusion both clinically and histologically. There are no specific lab markers.[1] Lesions are often initiated by minor trauma or scarring, and start with a small, red bump on the skin. Necrotic ulcers can develop rapidly over a period of 4-8 weeks. They usually exceed 10 cm in diameter and present with a purplish, undermined edge and surrounding erythema. Pathergy is the most important feature of PG. PG can affect any anatomical or surgical site. The skin over the tibia is a classic site. The head, oral cavity, and neck are rarely involved.[4] The differential diagnosis includes mycobacterial and deep fungal infections. Martorell ulcers and necrotizing fasciitis must be always excluded.[5] The treatment requires an interprofessional approach. The underlying systemic disease must be always treated.[6] Wound care and pain control are key features in the treatment. Debridement must be performed very cautiously because of the association with pathergy.[7] Systemic corticosteroids are the first-line therapy. Anti-TNF-alpha drugs such as etanercept and adalimumab have been used successfully.[8] The prognosis is generally good, but relapses are common.

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