59 y.o.

- Truck driver
- Diabetes type II (polyneuropathy, lower legs arteritis)
- Ischemic cardiopathy (hart infarct 2002)
- Morbid obesity
- Cholecystectomia
- Gastroplasty
Treatment

- Diabetic diet
- Novomix 30-20
- Asaflow 80
- Coruno 16: 1/day
- Bisoprolol 5: 1/day
- Coversyl 5: 1/day
- Simvastatin 40 mg/day
- Cymbalta 75
- Lyrica 75
Patient hospitalized for multiple lower leg ulcers of unknown etiology, highly painful and developing over a few days and weeks

Septic choc (ICU) (Meronem/Vanco)

Pyoderma Gangrenosum (?) treated by Methylprednisolone during 3 months

A few weeks after stopping corticoid extensive relapse of the ulceration on the foots

(PG is an exclusion diagnostic!)
Excessive search for a secondary cause of PG (Colonoscopy, Pet/CT, no monoclonality, …)

**Treatment:** Corticoid / Azathioprin / Cyclosporin

? Anti TNF

…
Table 1: Associated systemic diseases

1. Inflammatory bowel disease 15%
2. Rheumatoid arthritis and systemic lupus erythematosus (37%)
3. Immune abnormalities:
   - Humoral-
     - Congenital and acquired hypogammaglobulinemia
     - Selective and complete hyperimmunoglobulin E syndrome
   - Cell Mediated-
     - Immunodeficient/Immunosuppressed
     - Congenital deficiency in leukocyte adherence glycoprotein
     - Defective neutrophil function
4. Hematologic Diseases-
   - Acute and chronic myeloid leukaemia
   - Multiple myeloma
   - Monoclonal gammopathy
   - Waldenstrom’s macroglobulinemia
   - Lymphoma-Hodgkin’s lymphoma, non-Hodgkin’s lymphoma
   - Polycythemia Vera
   - Large granular Lymphocytic leukaemia
   - Myelofibrosis
5. Liver Diseases:
   - Chronic active hepatitis
   - Cryoglobulin and hepatitis C
   - Primary biliary cirrhosis
6. Solid tumours like colon, bladder, prostate, breast, bronchus, ovary, adrenocortical carcinoma
7. Drugs – Alpha 2-b Interferon
8. Miscellaneous – Thyroid diseases, sarcoid, diabetes mellitus, HIV, COPD
<table>
<thead>
<tr>
<th></th>
<th>Differential diagnosis of PG</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Infections</td>
</tr>
<tr>
<td></td>
<td>Herpes simplex, impetigo, eczema</td>
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<td></td>
<td>gangrenosum, cutaneous tuberculosis, deep fungal infections</td>
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<td>2.</td>
<td>Vasculitis</td>
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<td></td>
<td>Polyarteritis nodosa, Wegener’s</td>
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<td></td>
<td>granulomatosis, mixed cryoglobulinemia</td>
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<td>3.</td>
<td>Thrombophilic states</td>
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<td></td>
<td>Livedoid vasculitis, antiphospholipid</td>
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<td></td>
<td>syndrome, factor V Leiden mutation</td>
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<td>4.</td>
<td>Venous insufficiency</td>
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<td>5.</td>
<td>Malignancies</td>
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<td></td>
<td>Squamous cell carcinoma, cutaneous</td>
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<td></td>
<td>lymphoma, metastatic carcinoma</td>
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<td>6.</td>
<td>Ext. of underlying IBD</td>
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<td>7.</td>
<td>Factitious</td>
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<td>8.</td>
<td>Insect bite</td>
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<td>9.</td>
<td>Drugs</td>
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<td></td>
<td>Isotretinoin, granulocyte colony</td>
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<td></td>
<td>stimulating factor, iodine and bromide</td>
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<td></td>
<td>overdosage, alpha 2b-interferon^{17}</td>
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</tbody>
</table>
### Table 3: Criteria (including clinical characteristics) for diagnosis of Pyoderma Gangrenosum

**Major criteria**

1. Rapid\(^a\) progression of a painful\(^b\) necrolytic cutaneous ulcer\(^c\) with an irregular, violaceous, and undermined border
2. Exclusion of other causes of cutaneous ulceration

**Minor criteria**

1. History suggestive of pathergy\(^d\) or clinical finding of cribriform scarring
2. Systemic diseases associated with PG\(^e\)
3. Histopathologic findings (sterile dermal neutrophilia ± mixed inflammation ± lymphocytic vasculitis)
4. Treatment response (rapid response to systemic glucocorticoid treatment)\(^f\)

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\(^a\)Characteristic margin expansion of 1 to 2 cm/d, or a 50% increase in ulcer size within 1 month; \(^b\)Pain is usually out of proportion to the size of the ulceration; \(^c\)Typically preceded by a papule, pustule, or bulla; \(^d\)Ulcer development at sites of minor cutaneous injury; \(^e\)Inflammatory bowel disease, polyarthritis, myelocytic leukemia, or preleukemia; \(^f\)Generally responds to a dosage of 1 to 2 mg/kg/d, with a 50% decrease in size within 1 month
<table>
<thead>
<tr>
<th>Initial therapy</th>
<th>Therapy</th>
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<tbody>
<tr>
<td>Incomplete response to corticosteroids</td>
<td>Corticosteroids + cyclosporine A</td>
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<tr>
<td>I. First choice</td>
<td>Corticosteroids + azathioprine</td>
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<tr>
<td>II. Second choice</td>
<td>Mycofenolate mofetil</td>
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<td></td>
<td>Dapsone</td>
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<td></td>
<td>Chlorambucil</td>
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<td>Sulfasalazine</td>
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<td></td>
<td>Minocycline</td>
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<tr>
<td>III. For recalcitrant PG</td>
<td>Cyclophosphamide</td>
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<tr>
<td></td>
<td>Infliximab</td>
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<td></td>
<td>Thalidomide</td>
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<td>Tacrolimus</td>
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<tr>
<td>Drug</td>
<td>Dose schedule</td>
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<td>-------------------------------------------------------------------------------</td>
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<tr>
<td>Oral steroids</td>
<td>Prednisone 0.5-1 mg/kg/d</td>
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<tr>
<td>Intravenous pulsed steroids</td>
<td>Methyl prednisone 1 g daily in 5% dextrose for 1-5 days</td>
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<tr>
<td>Cyclosporine</td>
<td>4-5 mg/kg/d</td>
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<tr>
<td>Azathioprine</td>
<td>1-2 mg/kg/d (50-200 mg)</td>
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<tr>
<td>Cyclophosphamide</td>
<td>1.5-3.0 mg/kg/d (oral) IV Pulse 500 mg/2wks or 1000 mg/month</td>
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<td>Dapsone</td>
<td>50-200 mg/d</td>
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<tr>
<td>Methotrexate</td>
<td>10-30 mg/wk</td>
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<tr>
<td>Mycophenolate mofetil</td>
<td>2-3 g/d</td>
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<tr>
<td>Thalidomide</td>
<td>50-200 mg/d</td>
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<tr>
<td>Etanercept</td>
<td>50-100 mg SC qwk</td>
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<tr>
<td>Infliximab</td>
<td>3 mg/kg IV infusion</td>
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<tr>
<td>Adalimumab</td>
<td>40 mg SC q wk</td>
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<tr>
<td>Tacrolimus</td>
<td>0.15 mg/kg twice daily</td>
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<td>Plasmapheresis</td>
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<td>IVIG, hyperbaric O₂ radiation, electron beam irradiation</td>
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