Netherlands
The Journal of Medicine

PHOTO QUIZ

A cutaneous ulceration with pulmonary mass

E. Marchiori[1]*, B. Hochhegger[1], M.D. Guimarães[2], G. Zanetti[1]

[1]Department of Radiology, Federal University of Rio de Janeiro. Rio de Janeiro, Brazil, [2]Department of Radiology, Hospital AC Camargo, São Paulo, Brazil, *corresponding author: tel.: +55 24 22492777, fax: +55(21)26299017, e-mail: edmarchiori@gmail.com.

CASE REPORT

A 74-year-old man was admitted with a one-month history of haemoptysis, fever, cough, and 8 kg weight loss. He reported a four-year history of a cutaneous chest wall lesion, with previous empiric treatment but no diagnosis. Physical examination revealed a large, painful, deep ulceration with elevated erythematous-violaceous borders and a necrotic and haemorrhagic base on the left side of the back (*figure 1*). Chest radiography and computed tomography revealed an irregular cavitated consolidation in the right lung (*figure 2A and 2B*). The ulceration spread and progressed rapidly, despite administration of systemic broad-spectrum antibiotics and antifungal medication. Laboratory tests, including those for cytoplasmic and perinuclear antineutrophil cytoplasmic antibodies (ANCA), were negative. Cutaneous lesion cultures were also sterile. The patient underwent skin and pulmonary biopsies. The skin lesion showed diffuse neutrophilic infiltration without angitis or granuloma. Histological findings of the pulmonary biopsy corresponded closely with those of the skin biopsy. Systemic treatment with prednisone led to improvement of the cutaneous and pulmonary lesions within four weeks.

Figure 1. *A large ulceration with elevated erythematous-violaceous borders and a necrotic and haemorrhagic base on the left side of the back*

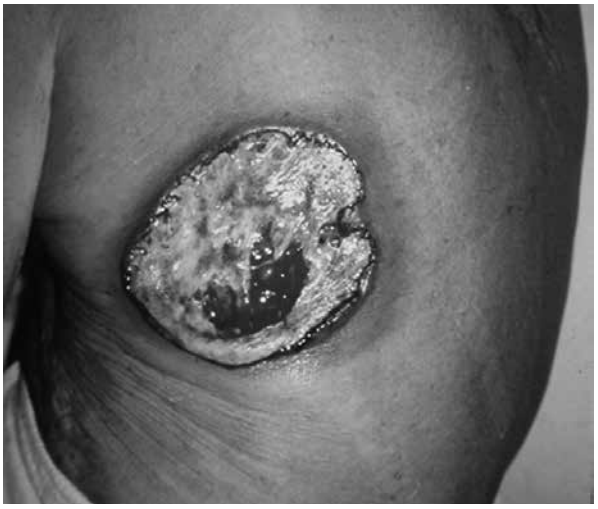
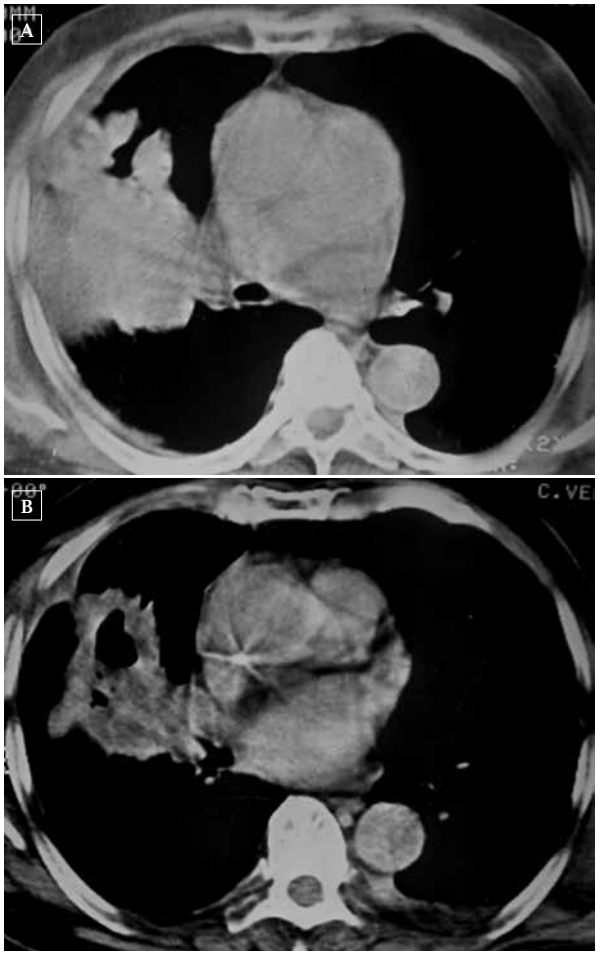


Figure 2. *Computed tomography scans (A and B) obtained at the level of the lower lobes showing an irregular cavitated opacity in the right lung*



WHAT IS YOUR DIAGNOSIS?

See page 156 for the answer to this photo quiz.

© Van Zuiden Communications B.V. All rights reserved.

ANSWER TO PHOTO QUIZ (PAGE 152)

A CUTANEOUS ULCERATION WITH PULMONARY MASS

DIAGNOSIS

The aggressive nature of the patient's ulcers, in combination with the negative findings of laboratory tests, cultures for bacteria and fungi, and histopathological examination, allowed the diagnosis of pyoderma gangrenosum (PG).

PG is a chronic inflammatory disease of unknown aetiology, characterised by neutrophilic infiltration of skin and lung tissues.[1,2] Its cutaneous manifestation is characterised clinically by erythematous-violaceous nodular lesions or pustules that progress and enlarge rapidly to painful ulcers of variable size, with irregular, loose borders and necrotic and haemorrhagic bases, preferentially located on the lower limbs.[3,4] The pathogenesis of PG remains unclear, but may involve derangement of immunity and/or neutrophil function.[1]

PG has been reported in association with various systemic diseases involving basic immunological disorders, such as inflammatory bowel disease (ulcerative colitis, Crohn's disease), polyarthritis, vasculitis, lymphoma, paraproteinaemia, leukaemia, rheumatoid arthritis, gammopathies, multiple myeloma, and active chronic hepatitis.[1-4]

Although PG is basically considered to be a dermatological disease, its clinical appearance has some systemic aspects.[2] Extracutaneous manifestations of PG are uncommon and have been reported in the bones, lungs, trachea and bronchi, liver, heart, pancreas, spleen, kidneys, and central nervous system.[3] Although systemic involvement is rare in PG, the lungs are the most commonly affected organs.[4] The main clinical manifestations in the lungs are solitary or multiple pulmonary nodules or masses, with or without evidence of central necrosis and cavitation, pleural effusion, and infiltration. The lungs of patients with PG also show marked intra-alveolar neutrophilic infiltration.[1-4] Skin biopsy is essential for diagnostic confirmation. Histological examination basically reveal aseptic inflammatory neutrophilic infiltrates.[4]

The differential diagnosis of PG primarily involves consideration of Wegener's granulomatosis, which can be associated with similar skin lesions and systemic involvement. The absence of upper respiratory tract involvement or kidney alteration, laboratory findings (cytoplasmic and perinuclear ANCA negativity), and pathological findings of tracheal and skin lesions ruled out Wegener's granulomatosis in our case.[2,4]

Steroid therapy is considered to be the first choice to control the systemic manifestations of PG, especially the pulmonary form.[2,4]

REFERENCES

1. Kanoh S, Kobayashi H, Sato K, Motoyoshi K, Aida S. Tracheobronchial pulmonary disease associated with pyoderma gangrenosum. Mayo Clin Proc. 2009;84:555-7.
2. Kasuga I, Yanagisawa N, Takeo C, Koga M, Kiyokawa H, Yonemaru M, et al. Multiple pulmonary nodules in association with pyoderma gangrenosum. Respir Med. 1997;91:493-5.
3. Kitagawa KH, Grassi M. Primary pyoderma gangrenosum of the lungs. J Am Acad Dermatol. 2008;59(5 Suppl):S114-6.
4. Bittencourt M de J, Soares LF, Lobato LS, Mançano AD, Leandro HS, Fonseca DM. Multiple cavitary pulmonary nodules in association with pyoderma gangrenosum: case report. An Bras Dermatol. 2012;87:301-4.

© Van Zuiden Communications B.V. All rights reserved.