Netherlands
The Journal of Medicine

PHOTO QUIZ

Fingertip necrosis and cervical lymphadenopathy

C. Bachmeyer[1]*, D. Orsini[1], J. Demay[1], S. Dautheville[2], S. Richard[3], O. Steichen[1,4]

[1]Department of Internal Medicine, [2]Emergency Unit, [3]Department of Oncology, Tenon Hospital (AP-HP), Paris, France; [4]Pierre et Marie Curie University, Faculty of Medicine, Paris, France, *corresponding author: tel: +33 (0)1 56016077, fax: +33 (0)1 56 017082, e-mail: claude.bachmeyer@tnn.aphp.fr

CASE REPORT

In January 2013, an 83-year-old man was admitted to the hospital for fingertip necrosis of both hands that had developed over three weeks. His medical history was remarkable for 70 pack-years of cigarette smoking until 1995, when excision for a vocal cord epidermoid carcinoma was performed. He denied use of any medication or illicit drugs. At examination, the patient was fatigued and had lost 5 kg. The tips of his 2nd and 4th fingers of the right hand and of the 4th of the left hand were necrotic (*figure 1*). Peripheral pulses were palpable. He did not report Raynaud's phenomenon. The heart exam was normal. An asymptomatic left cervical lymphadenopathy was found, hard and fixed, 6 cm in diameter. Routine blood cell count and serum creatinine were unremarkable. Thrombophilia work-up revealed a lupus anticoagulant and IgM anticardiolipin antibodies at 44 U (positive >15). The plasma level of platelet microparticles of the anionic phosphatidylserine was elevated at 7844/µl (normal 393-937) indicating *in vivo* platelet activation. Platelet aggregation induced by arachidonic acid and adenosine diphosphate was increased. Antinuclear antibodies were positive at 1/1280 with anti-SSA/Ro 60 kD antibodies. Cryoglobulinaemia was present at a very low concentration but could not be identified. Duplex ultrasonography of the upper extremity arteries was normal including palmar metacarpal arteries. Electrocardiogram showed a sinus rhythm. Transthoracic echocardiogram revealed no abnormalities; insertion of the transoesophageal echocardiography probe was unsuccessfully attempted. Aortic computed tomography scan was normal. 18-F fluorodeoxyglucose (FDG)-positron emission tomography detected accumulation of the tracer located to the left cervical lymphadenopathy and to right cervical lymph nodes. Fine needle aspiration of the left cervical lymphadenopathy was consistent with a metastatic localisation of epidermoid carcinoma. The search for the primary tumour was negative. Treatment with iloprost, heparin and aspirin was ineffective.

Figure 1. *Fingertip necrosis of both hands*

WHAT IS YOUR DIAGNOSIS?

See page 323 for the answer to this photo quiz.

ANSWER TO PHOTO QUIZ (PAGE 319)

FINGERTIP NECROSIS AND CERVICAL LYMPHADENOPATHY

DIAGNOSIS

A diagnosis of acral vascular paraneoplastic phenomenon was made.

DISCUSSION

Acral vascular paraneoplastic syndrome consists of Raynaud's phenomenon, acrocyanosis and gangrene developing in patients with malignancy and improving with the treatment of the underlying condition.[1,2] Indeed, in a review of the literature in 2002, 68 cases were identified associated with adenocarcinoma in 41% of patents (mainly lung and ovary cancers), but also with haematological diseases in 18% (such as lymphoma, leukaemia and plasmocytoma). Only a few patients had head and neck epidermoid carcinoma. Most patients had advanced cancer and the acral vacular syndrome preceded diagnosis of the malignancy. Pathogenesis is still disputed. Indeed, a vasoconstrictive substance produced by the tumour cells, microfragments of the tumour and hyperstimulation of the sympathetic nervous system have been advocated,[1] but also activation of coagulation and of platelets,[3] reported commonly in patients with malignancies and observed in our patient. Of note, multiple autoimmune abnormalities observed in our patient including antinuclear antibodies, cryoglobulins, and anticardiolipin antibodies, could play a role as described in the literature.[1,4] Interestingly, in our patient treatment combining cetuximab and carboplatin resulted in dramatic regression of the left cervical lymphadenopathy and of fingertip necrosis within three months. Indeed, an early diagnosis of the malignancy is mandatory since cancer treatment is the only intervention that can result in dramatic improvement of the lesions.

REFERENCES

1. Poszepczynska-Guigné E, Viguier M, Chosidow O, Orcel B, Emmerich J, Dubertret L. Paraneoplastic acral vascular syndrome: epidemiologic features, clinical manifestations, and disease sequelae. J Am Acad Dermatol. 2002;47:47-52.
2. Onitilo AA, Demos-Bertrand J, Depke J, Resnick JM, Engel J. Digital ischemia as a paraneoplastic consequence of squamous cell lung carcinoma. WMJ. 2012;111:138-41
3. Falanga A, Tartari CJ, Marchetti M. Microparticles in tumor progression. Thromb Res. 2012;129:S132-6.
4. Pham C, Shen YM. Antiphospholipid antibodies and malignancy. Hematol Oncol Clin North AmThromb Res. 2008;22:121-30.