Images in clinical rheumatology

Cutaneous Cryptococcosis in a patient with Behçet’s disease and colon cancer

Criptococosis cutánea en paciente con enfermedad de Behçet y cáncer de colon

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Introduction

Behçet’s disease is characterised by the classic triad of oral and genital aphthosis along with uveitis. However, it may be also associated with systemic involvement. Its cutaneous manifestations include erythema nodosum, pseudofolliculitis, purpura and migratory thrombophlebitis.

The following report describes a case of cutaneous cryptococcosis initially interpreted as vasculitis, which revealed an advanced stage of colon adenocarcinoma.

Clinical observation

The patient was a 67-year-old farm worker who suffered Behçet’s disease of 20 years evolution and who had been treated with colchicine 1mg/d. Although follow-up had been lost 5 years earlier, he came to the clinic due to the appearance of palpable purpura lesions on the back of his hands, thighs and forearms, with 2 weeks of evolution and with no oral-genital aphthosis. A general analysis was requested upon suspicion of cutaneous vasculitis and prednisone 20mg/d was indicated, pending further studies.

Two weeks later, the patient returned showing extensive necrotic and ulcerated lesions on the left hand (Figure), as well as the right thigh and forearm. Laboratory tests did not show systemic involvement. A biopsy of the lesions found dermal cryptococcosis, which was confirmed by culture. Treatment with fluconazole was indicated at doses of 400 mg/d and the reconstructive surgery service performed debridement of the lesions and local cures. A CT scan of the chest and abdomen confirmed multiple liver metastases from adenocarcinoma of the left colon, with no lung involvement. No skin grafting was proposed due to the short life expectancy of the patient, who died 4 months after diagnosis.

Discussion

Cutaneous vasculitis is a possible manifestation of Behçet’s disease. However, the slow-healing evolution of this patient and the absence of other manifestations of this disease evoked the possibility of cutaneous mycosis. Cryptococcosis is an opportunistic fungal infection of diverse manifestations, including cutaneous lesions that may be mistaken for vasculitis of a different origin in its initial stages. The most common risk factors are HIV infection, steroid therapy above 20mg/d, solid organ transplantation, rheumatic diseases under immunosuppression and neoplasms.

Figure. Ulcerated lesions on the back of the left hand, along with tendon exposure.
The main entry route of Cryptococcus neoformans is the airway, although haematogenous spread or spread through local dermal abrasions may be commonly facilitated in rural workers, as in the case of the patient described. Treatment with fluconazole or itraconazole is generally preferred, with surgical debridement of the lesions sometimes required. Amphotericin B and flucytosine are preferred in the case of haematogenous spread or neurological involvement.

In conclusion, cutaneous fungal infection should be suspected in patients with vasculitis-like lesions of slow evolution that are unresponsive to corticosteroids, and even more if there is a context of immunosuppression.

References