Scrofuloderma – still a diagnosis to bear in mind

Escrofuloderma – Um diagnóstico a ponderar no Mundo Ocidental

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Scrofuloderma – a case report

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ABSTRACT

The incidence of tuberculosis has been increasing worldwide. Contrarily, a recent decrease in Portugal has been reported. Cutaneous tuberculosis comprises a low percentage of all cases. We report a 70-year-old female with a 2-month-history of painful, nodular, suppurative lesions in the groin area. Previous history was remarkable for *Human Immunodeficiency Virus* infection and stage-IIIB cervical cancer. A skin biopsy, stained with PAS and FITE, Polymerase chain reaction on purulent discharge and mycobacterial culture of the skin were performed, leading to the diagnosis of scrofuloderma. Tuberculostatic therapy was initiated and complete response was observed.

This case depicts an uncommon variant of tuberculosis, highlighting the need for awareness of the cutaneous variants of tuberculosis that, although rare, can still present in clinic today.

RESUMO

A incidência de tuberculose tem vindo a aumentar globalmente. Em Portugal, porém, esta incidência diminuiu na última década. A tuberculose cutânea representa uma pequena percentagem de todos os casos. Apresenta-se o caso de uma mulher de 70 anos com nódulos supurativos da região inguinal, dolorosos, evoluindo há 2 meses. Como antecedentes relevantes, apresentava infeção por Vírus da Imunodeficiência Humana e carcinoma do colo do útero, estadio IIIB. Foram realizadas biópsia cutânea (coloração PAS e FITE), pesquisa de micobactérias por *Polymerase chain reaction* e estudo microbiológico por cultura, tendo sido estabelecido o diagnóstico de escrofuloderma. Iniciou terapêutiva tuberculostática com resposta favorável. Este artigo realça a importância do reconhecimento das formas cutâneas de tuberculose e a necessidade de manter um elevado índice de suspeição, sobretudo em pacientes imunodeprimidos.

Keywords: cutaneous tuberculosis; scrofuloderma;

INTRODUCTION

Over the last years the incidence of tuberculosis has increased worldwide especially due to the increasing incidence of *Human Immunodeficiency Virus* (HIV),1 being ultimately responsible for most deaths among these patients.2,3 In Portugal, tuberculosis is still an important health care issue, although its incidence has been dropping – with an estimate of 16,5 new cases for 100.000 inhabitants, with the majority of cases reported in the districts of Lisbon and Porto.4

Cutaneous involvement, although classical, remains an uncommon presentation of tuberculosis, comprising only 1-1.5% of all extra-pulmonary manifestations.5,6 Cutaneous tuberculosis can be acquired exogenously (direct inoculation) or endogenously (contiguous infection or hematogenous dissemination) and can present a wide variety of clinical features.7

CASE REPORT

A 70-year-old caucasian female was seen with a 2-month-history of multiple, painful, erythematous, nodular, ulcerated and suppurative lesions on the groin area, bilaterally (Fig. 1).

Physical examination revealed palpable inguinal lymphadenopathy on the right side, without any other significant clinical findings. The patient was afebrile and denied night sweats or other systemic symptoms.

Previous history was remarkable for HIV seropositivity diagnosed 8 years before, under antiretroviral therapy with a current CD4 count of 242/mm3, negative viral load, and stage-IIIB cervical cancer treated with a chemo-radiotherapy scheme that had finished 2 years before.

Given the clinical picture described, the main diagnostic considerations included cutaneous metastases, mycobacterial and deep fungal infections.

A skin biopsy showed a dermal tuberculoid granulomatous infiltrate with caseous necrosis, containing numerous multinucleated giant cells of the Langhans type accompanied by a rim of lymphocytes, plasma cells and occasional eosinophils (Fig. 2). PAS and FITE staining were negative. Polymerase chain reaction (PCR) performed on the exsudate revealed the presence of *Mycobacterium tuberculosis complex* and routine bacterial culture was positive for *Mycobacterium tuberculosis* (MTB) (confirmed only 4 weeks later). A CT scan showed contiguity between inguinal nodes and the overlying skin surface (Fig. 3).

The patient initiated treatment with a combination of pyrazinamide, isoniazid, ethambutol and rifampicin and her condition improved considerably in just two weeks of treatment. After 3 months of treatment only scarring were present (Fig. 4).

DISCUSSION

Scrofuloderma is a variant of cutaneous tuberculosis resulting from endogenous spread of the infection from a contiguous underlying focus, usually a lymph node or bone, with breakdown of the skin. The lesion starts as a subcutaneous, mobile nodule, which after attaches to the overlying skin leading to abscess formation and purulent discharge. This clinical manifestation is classically described in young patients,8 and is more often seen in the axillae, neck, groin and chest.9,10 It’s mainly associated to a state of lower immunity, with accompanying lower number of lymphocytes present in the granulomas and higher bacillary counts.

It’s interesting to note that other variants of cutaneous tuberculosis – such as Lupus Vulgaris and Tuberculosis Verrucosa Cutis, are associated with higher and intermediate immunocompetence states, respectively.11

Given the many clinical features of cutaneous tuberculosis and the fact that lesions resemble many other dermatological conditions, diagnosis can be difficult and requires correlation between clinical findings and diagnostic tests. In face of the delay in obtaining microbiological confirmation – 4 weeks, PCR can be of value. It has high sensitivity and specificity, with results in less than 24-48 hours. However, a positive result may not always correspond to an active infection and a negative result does not exclude it. 12,13 Therefore, despite its utility, clinical correlation is mandatory. In the setting of compatible clinical and histological features, even without a microbiological confirmation, tuberculostatic treatment should be started and, in these cases, the response to treatment can support the diagnosis.

In our patient, on clinical grounds alone, there was a possibility of cutaneous metastases from cervical cancer, given her medical history. However, the presence of typical tuberculoid granulomas in the skin combined with positive PCR for MBT, pointed to scrofuloderma, even in the absence of positive FITE and PAS stainings on histopathology. This was ultimately confirmed by isolation of MBT on routine bacterial culture (Löwestein-Jensenmedium) 4 weeks later. Sensitivity testing showed susceptibility to first-line tuberculostatics. The recommended treatment for cutaneous tuberculosis is the same as for systemic disease, as was carried out in our patient with good response.

This case is remarkable for several features: an uncommon presentation of tuberculosis – scrofuloderma - in a higher age group than average, maybe due to HIV-immunosuppression, with negative histochemical stain for mycobacteria in the cutaneous biopsy (unusual, although previously described), with a positive PCR for MBT and positive mycobacterial tissue culture.

Tuberculosis is still an important health issue and clinicians should be aware of atypical clinical presentations, especially in the context of immunosuppression. Timely investigation is key and can prevent further complications.

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FIGURE LEGENDS

Figure 1. Erythematous nodular lesions in the groin area with ulceration and suppurative content.

Figure 2. (a) Diffuse epidermal hyperplasia with dermal granulomatous infiltrate of tuberculoid type. (b) Central necrosis with numerous multinucleated giant cells of the Langhans type. Haematoxylin and eosin, original magnification (a) x 40, (b) x 200.

Figure 3. Chest-abdomen-pelvis CT showing a contiguity between the nodular lesions and the skin (red arrows).

Figure 4. Clinical appearance after three months of treatment.