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Case Report

Iatrogenic Kaposi's sarcoma in a patient with bullous pemphigoid treated with an oral corticosteroid

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ABSTRACT

Kaposi's sarcoma is a multifocal angiogenic tumor disease whose principal causal agent is human herpes virus 8 (HHV-8). Herein, we report a rare case of iatrogenic Kaposi's sarcoma developing during oral corticotherapy. A 76-year-old, HIV-negative male presented with papulous, angiomatous lesions on the trunk and limbs, which appeared three months after the beginning of oral corticotherapy for bullous pemphigoid. We suspected iatrogenic Kaposi's sarcoma given the time to lesion onset in relation to the immunosuppressive treatment, together with histological and virological confirmation of HHV-8. The lesions began to subside when corticosteroids were tapered down to 10 mg/day. This was the first case reported in our setting and it emphasized the need for the rigorous monitoring of patients receiving immunosuppressants to avoid overlooking the side effects or rare complications of these treatments.

Key words: Kaposi's sarcoma; Oral corticotherapy; HHV-8; Bullous pemphigoid

INTRODUCTION

Kaposi's sarcoma (KS) is a multifocal tumoral disease whose principal causal infectious agent is human herpes virus 8 (HHV-8), identified in 1994 [1]. Four clinical and epidemiological subtypes have been described: the classic or Mediterranean, endemic, epidemic AIDS-related, and iatrogenic. Iatrogenic KS occurs in patients exposed to long-term immunosuppressive treatments (topical or oral corticosteroids and/or other immunosuppressants) associated or not with organ transplantation [2-8]. It often raises the problem of managing the disease for which the immunosuppressant was prescribed because of the need to discontinue the drug or reduce the dose. Herein, we report a rare case of iatrogenic KS occurring during oral corticotherapy in a patient with bullous pemphigoid, who was followed at the dermatology department of Yalgado Ouédraogo University Hospital, Ouagadougou, Burkina Faso.

CASE REPORT

A 76-year-old male, married, a retired journalist, was followed for bullous pemphigoid confirmed by histology of a bullous lesion and by indirect immunofluorescence. Oral corticotherapy (prednisone) 60 mg/day (1 mg/kg/day) was prescribed. Arterial hypertension was discovered the

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day that the oral corticotherapy was initiated and he was given amlodipine 5 mg/day. He was HIV-negative.

Three months after the beginning of corticotherapy, reduced to 30 mg/day, he developed firm, angiomatous, nodular lesions measuring 0.5 to 2 cm in their greatest dimension, locally erosive and sensitive on the soles and sides of the feet, without lymphedema (Fig. 1a). These lesions then spread to the thighs and arms (Fig. 1b). There were no buccal lesions. The possible diagnoses considered were Kaposi's sarcoma, hypertrophic cutaneous lichen planus, and sarcoidosis. A pathological examination of a lesion biopsy specimen revealed an orthokeratotic, slightly acanthotic epidermis overlying a regular basal layer. The dermis revealed fusocellular tumoral proliferation with slit-like channels containing red cells, some of which were extravasated. Spindle cells were organized in twisted fascicles. Their nuclei were hyperchromatic and often mitotic. They were associated with inflammatory lymphoplasmacytic infiltrate (Figs. 2a and 2b). This morphological appearance was consistent with Kaposi's sarcoma. For confirmation, a blood sample and a swab from an erosive lesion were obtained for the molecular diagnosis of HHV-8 by real-time PCR. HHV-8 DNA was detected in plasma and in the lesion swab sample, confirming the diagnosis of KS. Lung radiography in search of interstitial infiltrates predominating in the two lung bases, nodules, mediastinal lymphadenopathy, and/or pleural effusions was normal. Abdominal and pelvic echography revealed no hepatic or splenic abnormality or lymphadenopathy.

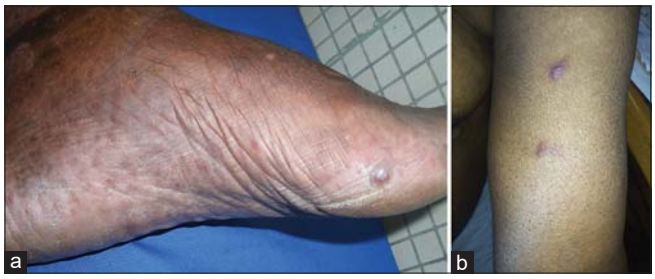

Figure 1: (a) Angiomatous papule on the internal side of the left foot. (b) Angiomatous plaques on the anterior aspect of the left arm.

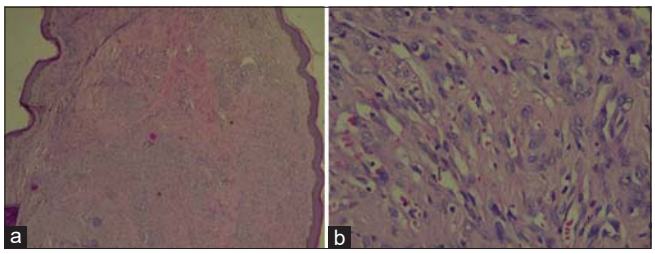

Figure 2: (a) Nodule biopsy specimen showing a regular epidermis overlying a dermis with vascular proliferation. (b) Vessels lined by an atypical endothelium with extravasated red cells.

As the disease progressed, the lesions increased in number to twenty. Prednisone was tapered first by 5 mg then by 10 mg every fourteen days. At a dose of 10 mg/day, the lesions began to subside. There was no recurrence of the bullous pemphigoid.

DISCUSSION

The iatrogenic subtype of KS was originally described in patients who had undergone organ transplantation, in particular kidney transplants followed by high-dose immunosuppressants [9]. Since then, several other cases have been reported in patients who have not had organ transplantation yet have been receiving immunosuppressants, including systemic and topical corticosteroids, for a variety of disorders (blood diseases, kidney diseases, atopic dermatitis, asthma, chronic inflammatory disease [8,10-14].

Herein, we report the first case of iatrogenic KS in a patient with skin phototype VI observed at the department of dermatology and venereology of Yalgado Ouédraogo University Hospital, the national reference center in Burkina Faso. This case occurred in a patient who had been receiving oral corticotherapy for bullous pemphigoid (pemphigoid diseases account for 4.6% of hospital admissions at our institution [15]). Some cases of KS in patients followed for an autoimmune bullous dermatosis (bullous pemphigoid or pemphigus vulgaris) treated with oral corticotherapy [10,14] and/or local corticotherapy [5,6] have previously been reported in the literature, yet not in sub-Saharan Africa.

The cases of iatrogenic KS described in the literature in patients with bullous pemphigoid occurred in elderly subjects, as in our patient (76 years), most seventy years old or older [2,5,14].

The time to onset of KS in our patient was 3 months after the start of corticotherapy. This corresponds to the time to onset (1-36 months) observed by other authors such as Tourlaki et al. [14] and Tremblay and Friedmann [2]. This time-lapse is in support of the iatrogenic nature of the disease in our patient, particularly as he had no sarcomatous lesions before oral prednisone was prescribed. His lesions developed at a prednisone dose of 30 mg/day, which is close to the dose of 8 to 25 mg/day recorded by Tourlaki et al. [14].

In addition to the oral corticosteroid (prednisone or methylprednisolone), which was the only drug taken by our patient, some reported cases had also received other associated immunosuppressants for the treatment of their bullous pemphigoid, either a dermal corticosteroid or mycophenolate mofetil [2]. Among the patients who applied a very strong topical dermal corticosteroid to their bullous lesions, without oral corticosteroids, some also took methotrexate [5].

In our patient, KS involved only the skin, with lesions on the limbs, as has been reported by some authors whose patients were followed for bullous pemphigoid [2,14]. However, in iatrogenic KS, mucosal involvement has frequently been described. Tourlaki et al. described duodenal involvement in one of their patients [14]. We did not perform gastrointestinal fibroscopy in our patient as he had no gastrointestinal warning signs.

Paraclinically, our patient's HHV-8 infection was confirmed by molecular diagnosis. However, such investigations are not routine as they are costly and performed in a research setting. HHV-8 is endemic in sub-Saharan Africa, where 30% to 60% of asymptomatic adults have markers of the infection [16]. In studies by other authors, patients did not always undergo HHV-8 serology and/or molecular biology techniques such as real-time PCR, yet the disease was confirmed histologically.

The modulation of the immunosuppressive treatment is the principal therapeutic weapon in controlling the progression of iatrogenic KS. In our patient, the lesions began to subside at the dose of 10 mg/day of oral corticosteroids. Some authors have reported complete remission of KS lesions when the immunosuppressant involved was decreased or discontinued [6,8,14]. Other authors, however, had to initiate specific treatment for KS (a course of bleomycin, intralesional injection of vincristine, intravenous vinblastine, radiotherapy) in order to obtain remission or stabilization of the lesions [4,5,14]. Worsening of the lesions and rare cases of death due to disseminated intravascular coagulation or septic shock have been reported [10].

With regard to the course of bullous pemphigoid in our patient, it did not recur during the tapering of corticotherapy. However, a longer follow-up period is required for the better evaluation of the course of the disease. We would also like to note that most authors report improvement of their patient's bullous pemphigoid with no recurrence on discontinuation of the immunosuppressive treatment over a longer period than in our case [5,6,14].

CONCLUSION

This rare case an HIV-negative patient with iatrogenic KS induced by long-term oral corticotherapy prescribed for the treatment of bullous pemphigoid highlights the need for rigorous and close patient monitoring in order to avoid overlooking the side effects or rare complications of such treatment. Decreasing the dose of the immunosuppressant or its complete discontinuation is generally followed by regression of KS.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

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