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CASE REPORT

Cutaneous Kaposi Sarcoma During Treatment with Steroids and Rituximab for Pemphigus Foliaceus

Running Title: Kaposi Sarcoma, Steroids and Rituximab, Pemphigus Foliaceus

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ABSTRACT

Introduction: Pemphigus foliaceus is an autoimmune disease with bullous manifestations on the surface of the skin that requires long-term administration of immunosuppressive medications. Kaposi's sarcoma is one of the diseases that manifest in people with innate or acquired immune systems deficiency with the direct involvement of HHV-8 and causes polypoid skin lesions. **Case presentation**: This study was conducted on a 78-year-old man with a history of pemphigus foliaceus treated with immunosuppressive drugs. **Conclusion**: In this patient, a secondary polypoid lesion had been created around the anus and the results of pathological tests had confirmed the Kaposi's Sarcoma.

INTRODUCTION

Pemphigus foliaceus (PF) is an autoimmune disease with blisters on the surface of the skin that does not manifest on the bullous mucosa (1,2). The prevalence of PF varies worldwide so that it ranges from 1-0.5 per 1 million people in Western Europe and 6.7 patients per 1 million people in Tunisia. Although it is generally recognized as a sporadic disease around the world, it is endemic in Brazil located in South America (3).

Kaposi's sarcoma (KS) is a benign vascular neoplasm in immunosuppressed individuals, especially in human immunodeficiency virus (HIV)-infected individuals (4,5). Herpesvirus 8 (HHV-8) infection is involved in the creation of this vascular tumor (6). This disease is common in Eastern Europe, the Mediterranean, and North Africa in men with the age range of 64 and 72 years. It is also known as endemic in the Sahara Desert of Africa (6).

The present case study was conducted on an Iranian male patient with a history of PF who was treated with corticosteroids and rituximab. Five months after the first dose of corticosteroid, KS polypoid ulcerative lesion manifested.

CASE PRESENTATION

The case of this study is a 78-year-old Iranian patient suffering PF since 2019. He had bullous lesions on the head and neck of the dorsal body and upper and lower limbs in the form of skin blisters. In addition, no lesion was observed on the surface mucosa, including oral mucosa. A biopsy of 0.3 cm from the patient's skin showed intraepidermal acantholytic blisters. In addition, in the upper dermis area, moderate

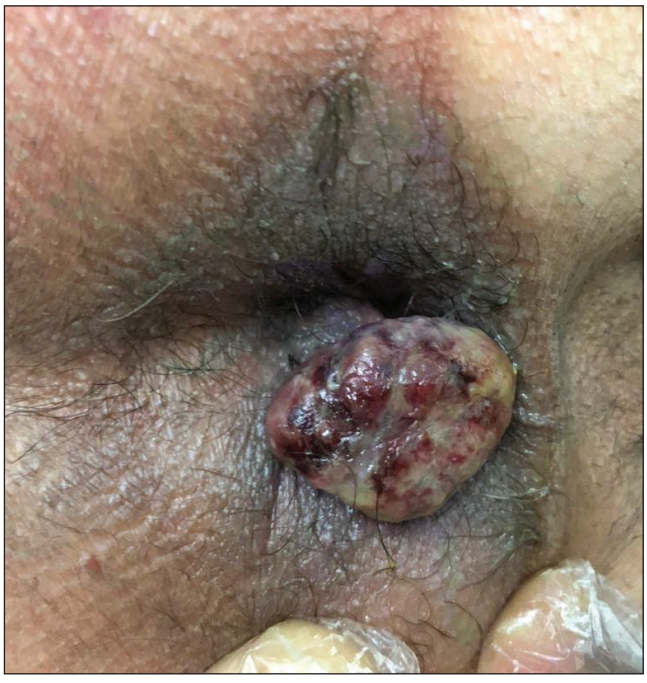

Figure 1. The lesion created in the anal area of the patient

infiltration of lymphocytic, neutrophilic, and eosinophilic cells was observed around the superficial blood vessels. Its result was confirmed diagnosis of PF.

The patient was treated with corticosteroid prednisolone pulse at a dose of 1 g/day for 3 days, followed by receiving an intravenous infusion of rituximab at a dose of 500mg / 50ml once per week for 4 weeks. In addition, after three days of receiving prednisolone pulse at the mentioned dose, he received 60 mg daily oral prednisolone for 2 weeks, followed by a dose reduction for 90 days. All blisters were resolved after the considered treatments. Five months (20 weeks) after receiving the first dose of corticosteroid, the patient noticed a lesion in the area around the anus that developed into an ulcerative polypoid after 1.5 months of growth (Figure 1). He underwent lesion resection surgery and the lesion was sent to the pathological laboratory for examination.

The results of pathologic tests showed that the polypoid lesion was at dimensions of 3.5 × 2 × 1 cm and resected tumoral lesion cells were positive in terms of infection with herpesvirus 8 (HHV-8), CD31, and CD34. Ki-67 was also positive in 40-45% of tumoral cells and the tumor cells were morphologically consistent with KS, confirming the diagnosis of KS.

DISCUSSION

KS is a benign angioproliferative neoplasm that causes polypoid lesions on the skin surface in three classes: 1) Classic KS, 2) Endemic KS, and 3) iatrogenic KS. The classic type of disease is more common in older men of Eastern Europe, the Mediterranean, and North Africa, but its endemic type is more common in Sahara Desert of Africa especially in people with the age range of 25-40 years. The incidence of KS is highly correlated with decreased immunity, so that as this disease is much more prevalent in HIV-infected people and transplant recipients who have lower immunogenicity than healthy individuals (4, 5) The iatrogenic types of KS are seen in patients who have used immunosuppressive drugs such as cyclosporine, corticosteroid, and rituximab (6). The case reported in this study also had a history of taking prednisolone and rituximab.

Based on the reports of the researchers around the world, KS can manifest during the treatment of bullous pemphigoid with steroids and methotrexate. Therefore, it can be suggested that the treatment of chronic autoimmune diseases with immunosuppressive drugs may increase the risk of iatrogenic KS. Another study (8) showed that a person was affected by KS with receiving short-term doses of immunosuppressive drugs, including steroids and methotrexate. The results confirm the efficacy of the medications received by the patient in the present study in the incidence of KS.

Another case report of iatrogenic KS (9) showed that a person with bullous pemphigoid treated with prednisolone and received clobetasol was affected with KS. This case study indicates the effect of immunosuppressive drugs on the incidence of this disease. The present study is a rare case in which the patient was affected by KS after being affected by PF and receiving the treatments. Similar case reports are rare around the world. Researchers can investigate the effects of immunosuppressive drugs such as those used on the case. Moreover, such studies try to prescribe a safer dose of the drug according to the patient's age and condition and reduce the incidence of such lesions by moderating the dose.

CONCLUSION

Finally, it can be concluded that the use of prednisolone and rituximab in the 78-year-old patient resulted in a lesion in the anus area. The cells of this polypoid lesion were positive for HHV-8 infection. The results showed that CD31 and CD34 were positive, confirming the iatrogenic KS.

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AUTHOR CONTRIBUTIONS

All authors contributed equally in this case and manuscript

CONFLICT OF INTERESTS

The authors declare that they have no conflict of interest.

ETHICAL STANDARDS

Patient's personal and private information are confidential and this study conducted by participants knowledge and consent.

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