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CASE REPORT

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Kaposi's Sarcoma in A Subject Treated for Dermatomyositis With Immunosuppressant and Steroid

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ABSTRACT

The incidence of Kaposi's sarcoma has increased in recent years mainly because its association with the acquired immunodeficiency syndrome and organ transplant recipients treated with immunosuppressive drugs, especially with cyclosporine. We present the case of a patient with dermatomyositis who developed Kaposi's sarcoma after receiving treatment with steroids, cyclosporine, and human polyclonal immunoglobulin.

KEYWORDS: Dermatomyositis. Kaposi's Sarcoma, Cyclosporine

INTRODUCTION

Kaposi's sarcoma has received special attention in recent decades due to its progressive increase in patients with acquired immunodeficiency syndrome and in transplant patients treated with immunosuppressive drugs.[1] Cyclosporine is an immunosuppressive agent used in transplanted patients to prevent rejection.[2] The incidence of malignant tumors in transplanted patients receiving cyclosporine is higher than in the general population. The most common tumors are non-Hodgkin lymphoma and Kaposi's sarcoma.[3,4] Cyclosporine is used to control diseases of an autoimmune nature.[5,6] We present the case of a patient diagnosed with dermatomyositis, treated with glucocorticoids, cyclosporine, and human polyclonal immunoglobulin, who subsequently developed a Kaposi's sarcoma.

CASE REPORT

A 55-year-old man presented to us in whom proximal muscle weakness began in the shoulder and pelvic girdle 2 months before admission. Physical examination revealed a heliotropic exanthema in the eyelids and decreased proximal muscle strength in the upper limbs and the lower limbs. The remainder of the examination was normal. Laboratory tests detected an erythrocyte sedimentation rate of 66 mm in the first hour. Serum glutamic-oxaloacetic transaminase values were 874 U/L; glutamic-pyruvic transaminase, 541 U/L; gamma-glutamyl transpeptidase, 135 U/L; alkaline phosphatase, 399 U/L; lactate dehydrogenase, 2,400 U/L; creatine phosphokinase, 10,026 U/L, and aldolase, 80 U/L. The determination of antinuclear antibodies, rheumatoid factor and other antibodies, including anti-Jo1, was negative. Complementary and immunoglobulin values were normal.

Electromyographic study was consistent with the diagnosis of inflammatory myopathy. Muscle biopsy showed the presence of a lymphocyte infiltrate surrounding muscle fibers and small vessels, with perifascicular atrophy and fat replacement, consistent with the diagnosis of dermatomyositis. No occult neoplasm was detected.7,8 Initial treatment consisted of glucocorticoids. After a month the patient had not experienced any improvement, with elevated muscle enzymes persisting. Therefore, treatment with human polyclonal immunoglobulin and cyclosporine was started, with good clinical response and decreased muscle enzyme values, normalizing within one month. The prednisone dosage was tapered to 30 mg/day.

Three months after starting cyclosporine, the patient developed several macular violaceous lesions on the skin of the trunk, face and limbs, as well as on the mucosa of the oral cavity. The diagnosis of Kaposi's sarcoma was histopathologically confirmed. Systemic involvement was ruled out and testing for viral infections, including the human immunodeficiency virus and cytomegalovirus, was negative at baseline and after 6 months. It was decided to discontinue cyclosporine and reduce the glucocorticoid dosage to 20 mg/day and continue with human polyclonal immunoglobulin. Six months after the onset of Kaposi's sarcoma, the patient was asymptomatic from his dermatomyositis. However, the Kaposi's sarcoma persisted, and he presented with new lesions. Radiotherapy was applied to the skin lesions, some of which remitted.

DISCUSSION

Kaposi's sarcoma has occasionally been described as

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being associated with dermatomyositis and polymyositis.[9-12] In our patient, Kaposi's sarcoma most likely developed secondary to immunosuppressive therapy for two reasons: the clear relationship between the onset of treatment with glucocorticoids and cyclosporine and the presentation of Kaposi's sarcoma, and the increase in the frequency of this tumor in patients undergoing immunosuppressive drugs. Kaposi's sarcoma has been described in association with the use of immunosuppressive drugs in patients who had received transplants.[3,4,13] It has been reported less frequently in patients with autoimmune diseases treated with these drugs.[10,11,14] The incidence of Kaposi's sarcoma has increased since the introduction of cyclosporine as the drug of choice in transplant patients. It is estimated that in these patients the possibility of developing Kaposi's sarcoma is 400–500 times higher if treated with azathioprine and glucocorticoids. This increase reaches 5000 times as much if receiving cyclosporine13, and furthermore the tumor develops earlier.[3,4] In most cases, treatment discontinuation results in partial or complete remission of the Kaposi's sarcoma.[3,4,11-14] Our patient was treated with human polyclonal immunoglobulin. This treatment is effective in controlling inflammatory myopathies15, and recently as demonstrated by Carmeli in a patient with polymyositis[12], may even cause the human polyclonal immunoglobulin lesions to return. Although control of the dermatomyositis was achieved in our case, the Kaposi's sarcoma lesions did not decrease. We also considered the possibility that we may have introduced the HIV with this treatment. However, repeatedly negative serological tests practically rule out this possibility. Another possible hypothesis about the pathogenesis of these 2 processes would be that dermatomyositis, being an autoimmune disease, acting on its own or in association with therapeutic immunosuppression, would have predisposed the patient to developing Kaposi's sarcoma. The strong relationship between Kaposi's sarcoma and immunosuppressive therapy, especially cyclosporine, suggests that this treatment is an important factor in the pathogenesis of this tumor. The immunocompromised state caused by this drug could facilitate the expression of viral agents. The virus most recently implicated as a causative agent of Kaposi's sarcoma is the human herpesvirus type 8, demonstrated in both Kaposi's sarcoma in HIV-infected patients as well as those of Mediterranean or African origin, and associated with transplant patients.[16]

CONCLUSION

The recent introduction of cyclosporine in the treatment of dermatomyositis may increase the number of Kaposi's sarcoma cases in dermatomyositis in the coming years. Healthcare professionals should be aware of such adverse events of cyclosporine.

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