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Case Report

Kaposi's sarcoma concurrent with granulomatosis polyangiitis

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Abstract

Kaposi's sarcoma (KS), a malignant vascular tumor, can develop in patients who receive corticosteroids or immunosuppressive therapy. We report a patient with KS and granulomatosis polyangiitis (GPA). To the best of our knowledge, this is the first report regarding the co-occurrence of these two diseases. The physician must be aware of the risk of cancer in patients with rheumatism.

Keywords: Kaposi sarcoma, granulomatous poliangiitis, vasculitis

Introduction

Kaposi's sarcoma (KS) is a rare malignant vascular tumor widely reported after renal transplantation, iatrogenic immunosuppression, and in patients with acquired immune deficiency syndrome (1). KS is less commonly described in patients with rheumatic diseases receiving long-term corticosteroids and/or immunosuppressive therapy (2).

We report the case of a patient concomitantly diagnosed with KS and granulomatosis polyangiitis (GPA).

Case Presentation

A 70-year-old woman with diabetes mellitus and hypertension was admitted to the hospital with fever, weakness, and widespread pain. She had coarse rales at middle and base of the right lung, bilateral pretibial edema and purplish purpuras on the fingers and sole of the left foot. Abnormal laboratory investigations included hemoglobin level, 10.1 g/dL; leukocyte count, 8.9×10^3/μL; thrombocyte count, 676×10^3/μL; sedimentation rate, 80 mm/h; C-reactive protein level, 204 mg/L; creatinine level, 1.3 mg/dL; blood urea level, 58 mg/dL; serum albumin level, 27 g/L; spot urine protein/creatinine ratio, 1.13; and positive c-ANCA. Renal ultrasonography revealed bilateral normal-sized kidneys. Chest X-ray revealed nodular lesions, and computed tomography revealed multiple lymph nodes (largest, subcarinal, 11×9 mm) and bilateral pleural effusion. Cytology of the pleural effusion revealed benign cells. Superficial perivascular dermatitis was observed on the skin biopsy of the left foot. Furthermore, she suffered from epistaxis, and in nasal examination, septal perforation was observed. Renal biopsy demonstrated focal necrotizing/crescentic glomerulonephritis with vasculitis. On the basis of these findings, she was diagnosed with GPA, and pulse steroid (1 g/day for 3 days) and intravenous cyclophosphamide treatments were initiated. After the third cycle of cyclophosphamide, purplish skin lesions on the left foot worsened, and new lesions developed on the left knee. Incisional biopsy was performed again, and KS was diagnosed on pathological examination. Because of the diagnosis of KS, cyclophosphamide was discontinued, and corticosteroid was gradually decreased. The patient was negative for human herpes virus-8 (HHV-8) and human immunodeficiency virus. She was referred to the medical and radiation oncology departments and was evaluated as stage III KS, and palliative radiotherapy and systemic chemotherapy were administered.

Discussion

We report the case of a patient with KS and GPA. This is the first study in literature to report the coincidence of these two diseases. The physician must be aware of the risk of cancer in patients with rheumatism.

Louthrenoo et al. (2) reviewed 24 cases of KS associated with immunosuppressive treatment of rheumatic disease, and since 2003, a few other cases have been reported (3, 4). All these patients developed KS after immunosuppressive treatments, unlike our case. Considering the number of patients with rheumatism who are receiving corticosteroid and/or immunosuppressive treatments, it is an extremely rare complication. Some evidence supports that steroids have a direct role in stimulating tumor development and growth (5). Gou et al. (6), reported the high levels of steroid receptors in KS lesions and that receptors

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can be upregulated by exogenous steroids and inflammatory cytokines. In autoimmune and/or inflammatory diseases, the risk of de novo cancer development is increased. Moreover, immunosuppressive therapies decrease the immune system's ability to recognize and eliminate malignant cell clones and may have direct mutagenic properties (7). In our patient, the co-occurrence cannot be explained by the effect of steroid or immunosuppressive agents. However, the deterioration of KS lesions was probably related to the therapy.

To date, only four cases of KS development after immunosuppressive therapy for GPA has been reported (3, 4, 8, 9). GPA is generally considered as a primary vasculitis; however, infectious agents, drugs, or cancer could be etiological factors. Vasculitis occasionally develops secondarily to malignancy and may occur in patients with previous or concomitant cancer because of risk factors predisposing to both conditions. In addition, inflammatory responses provoked by the underlying neoplasm contribute to the pathogenesis of malignancy-associated vasculitis (10).

KS is an opportunistic tumor associated with HHV-8 and linked to CD4-cell depletion. Louthrenoo et al. (2) reported a low CD4/CD8 lymphocyte ratio in three of the six patients with KS and rheumatic disease. In total lymphocyte numbers and absolute/relative numbers of CD4 T-helper cells are low in patients with GPA compared with controls, irrespective of the disease activity (11). Therefore, low CD4-T helper state may play an important role in the pathogenesis of KS development.

The coincidence of the two diseases may be unexpected for clinicians but must be considered despite its rare occurrence. Therefore, the physician must be aware of the risk of cancer in patients with rheumatism.

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