Kaposi’s Sarcoma in a Human Immunodeficiency Virus-negative Patient Treated with Corticosteroid for Idiopathic Thrombocytopenic Purpura

Key words: Kaposi’s sarcoma, immunosuppressive therapy, corticosteroid, idiopathic thrombocytopenic purpura

Kaposi’s sarcoma (KS) is a multifocal vascular neoplasm most commonly seen in association with human immunodeficiency virus (HIV) infection (1, 2). However, KS has been also reported in HIV-negative patients treated with immunosuppressive drugs for several clinical entities (3–5). We report a case with idiopathic thrombocytopenic purpura (ITP) that developed KS during corticosteroid therapy.

In June 1999, a 68-year-old Japanese man, who was a native of Miyako island of Okinawa, was admitted to Miyako Prefectural Hospital because of gingival bleeding. The primary condition was diagnosed as ITP. The patient was treated with intravenous methylprednisolone (1,000 mg/day for 3 days) followed by oral prednisolone (60 mg/day). After five months, the prednisolone dose was gradually tapered to 15 mg/day. The effect of corticosteroid was transient and the platelet count gradually decreased despite the use of a higher dose of oral prednisolone (60 mg/day for two weeks and thereafter tapered). The patient reported the appearance of dark-red nodules in the extremities in April 2000. He was referred to the Second Department of Internal Medicine, Faculty of Medicine, University of Ryukyus Hospital for splenectomy and examination of skin lesions. On admission, physical examination revealed several dark papules and nodules measuring 0.5 to 2.0 cm in diameter, on the hands, arms and legs. Laboratory tests revealed thrombocytopenia with platelet count of 22×10^3/μl, white blood cell count 5,300/μl with 1,130/μl lymphocytes and CD4/CD8 ratio of 0.46. Antibodies against both human T-lymphotropic virus type I and HIV were negative. Histopathological analysis of biopsied skin specimens showed proliferation of spindle-shaped cells with dilated vascular spaces, which were lined with plump endothelial cells. Immunohistochemically, staining for CD34 was positive in many endothelial cells and some of spindle-shaped cells. HHV8 DNA sequence was detected in the skin tumors by polymerase chain reaction using specific primers (2) (Fig. 1). Serum HHV8 antibody was also detected by indirect immunofluorescence test. Based on the above findings, the case was diagnosed as KS. Laparoscopic splenectomy was carried out without any complications in May 2000. Subsequently, the platelet count remained above 200×10^3/μl. Soon after discontinuation of corticosteroid therapy, the skin tumors regressed without the use of any specific treatment for KS. No recurrence of KS was observed during the 30-month follow-up period.

KS has been well described in a number of renal and other organ transplant recipients during long-term immunosuppression (1). Less frequently, appearance of KS during immunosuppressive therapy has been reported in various clinical entities such as rheumatoid arthritis, autoimmune hemolytic anemia, bullous pemphigoid, bronchial asthma, Sjögren’s syndrome, chronic lymphocytic leukemia, pemphigus vulgaris, Sézary syndrome, chronic obstructive lung disease, Wegener’s granulomatosis, systemic lupus erythematosus, polymyositis and ITP (3–5). It is known that HHV8 is the causative agent of KS (1, 2) and can be transmitted by sexual contact, maternal-infant pathway, and other means. The seroprevalence of HHV8 among blood donors ranges from 0.2 percent in Japan, to up to 10 percent in the United States, and to more than 50 percent in many African countries (1).

Although KS is rare in Japan, some cases of KS have been diagnosed in Okinawa (6). Therefore, Okinawa is suspected to be one of the endemic areas of KS. Okinawans are genetically similar to main island Japanese. On the other hand,
Okinawa is located 600 km from the southernmost part of main island Japan in the subtropical zone and it is also known to be an endemic area of HTLV-I. Since there is no significant difference in the seropositivity rates of anti-HHV8 antibodies between the Okinawa population and general Japanese population (7), environmental factors might be important in the development of KS in Okinawa. Thus, it can be speculated that HHV-8 infection and steroid-induced immunosuppression, as well as environmental factors, played a role in the development of KS in the present patient.

The use of immunosuppressive therapy has increased greatly in recent years. It should be kept in mind that KS could develop during immunosuppressive treatment even in HIV-negative patients. As we reported here, immunosuppression-associated or iatrogenic KS regressed with the cessation, reduction, or modification of immunosuppressive therapy (3, 4).

References