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Journal

Dermatology Online Journal, 22(12)

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Publication Date

2016

DOI

10.5070/D32212033378

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Peer reviewed

Kaposi's sarcoma in an HIV-negative patient

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Abstract

We report an HIV-negative, 55-year-old man with recurrent Kaposi's sarcoma (KS) of the lower extremities, who does not fit into any of the four previously described variants of KS: classic KS, AIDS-related KS, iatrogenic KS, and African-endemic KS. There are reports in the literature of childhood-onset KS, which is thought to be due to an inherited immune deficiency that confers a higher susceptibility to human Herpesvirus-8 (HHV-8), which is the virus that is known to cause KS. Our patient may be affected with an inherited immune deficiency that has predisposed him to KS, and this mutation also may account for his prostate and bladder cancer.

Case Presentation

PATIENT: 55-year-old-man

DURATION: Seven months

DISTRIBUTION: Lower legs and feet

HISTORY: A 55-year-old man presented to the Dermatology Clinic at the Veterans Affairs New York Harbor Hospital for evaluation of a progressive eruption on his lower legs. He had a history of recurrent, classic Kaposi's sarcoma (CKS), which had resolved with chemotherapy and radiation. Approximately one year prior, the patient developed pain of the lower legs and feet. Several months after the onset of pain, a slowly-enlarging, violaceous plaque developed on the plantar aspects of both feet. More recently, he had developed several, asymptomatic, red-brown papules on his lower legs.

Past medical history included Kaposi's sarcoma. His wife died of complications from acquired immune deficiency syndrome in 1988, but multiple tests for the human immunodeficiency virus in the patient were negative, most recently one month



Figure 1. Hyperpigmented patches on bilateral lower legs. Admixed within the hyperpigmented patches were several, small, red-brown papules.



Figure 2. Violaceous micronodules that coalesced into a plaque were present on the medial plantar aspect of the left foot.

prior to presentation. His medical history included prostate cancer and bladder cancer, both of which had been treated surgically and were in remission. He denied any Mediterranean or Ashkenazi Jewish ancestry. He denied any systemic symptoms, which included weight loss, fevers, chills, fatigue, and gastrointestinal symptoms. A punch biopsy was obtained from a violaceous lesion on the medial aspect of the left foot. A punch biopsy also was obtained from a red-brown papule on the left lower leg.

PHYSICAL EXAMINATION: Hyperpigmented patches were present on both lower legs (**Figure 1**). Violaceous micronodules that coalesced into a plaque were present on the medial plantar aspect of the left foot (**Figure 2**). On the plantar aspect of the right foot, there was a small focus of violaceous papules. Admixed within the hyperpigmented patches were several, small, red-brown papules.

LABORATORY DATA: An ELISA test for human immunodeficiency virus was negative.

HISTOPATHOLOGY: In the superficial-to-mid dermis, there is a proliferation of blood vessels with slit-like spaces, some of which are present around pre-existing vascular plexuses. A sparse, perivascular, lymphocytic infiltrate with rare plasma cells is noted. The overlying epidermis is unremarkable (**Figure 3**).

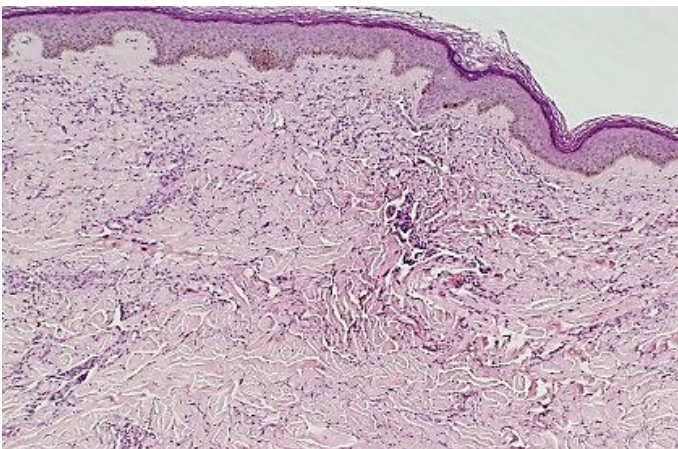


Figure 3. In the superficial-to-mid dermis, there is a proliferation of blood vessels with slit-like spaces, some of which are present around pre-existing vascular plexuses.

DIAGNOSIS: Kaposi's sarcoma

Discussion

Kaposi's sarcoma (KS) is an uncommon, vascular malignant condition that is characterized by a proliferation of aberrant endothelial cells. KS is a virally-induced disease that is caused by human herpesvirus-8 (HHV-8), which is also known as Kaposi-sarcoma-associated herpesvirus (KSAH). There are four major clinical variants: classic KS (CKS) that affects elderly men of Ashkenazi Jewish and Mediterranean origin, African-endemic KS, iatrogenic KS that arises in immunosuppressed patients, and acquired immune deficiency syndrome (AIDS)-related KS [1]. Although infection with HHV-8 is necessary for the development of all types of KS, the virus is not sufficient to cause disease. Seroprevalence studies have demonstrated HHV-8 infection in healthy individuals, and the reason that only a small portion of individuals infected with HHV-8 develop KS has eluded researchers for years.

First reported by Mortiz Kaposi in 1872, KS was originally described as an indolent disease of elderly men and was named idiopathic multiple pigmented sarcoma in the skin [2]. The disease did not garner much attention until the AIDS epidemic, when Alvin Freidman- Kien first reported disseminated Kaposi's sarcoma in young homosexual men in 1982 [3]. KS now is recognized as a common AIDS-defining malignant condition, and, although the incidence has declined with the introduction of highly active retroviral therapy (HAART), AIDS-related KS is the most common KS variant in the United States [4].

On histopathologic examination, KS shows a vascular dermal proliferation that is comprised of neoplastic blood vessels and spindle cells. Common features include the promontory sign, which describes a pattern of neoplastic vessels that preferentially form around pre-existing vessels. Immunostaining for HHV-8 is positive.

Each of the four clinical variants of KS are associated with a different disease course, prognosis, and site of predilection [5]. Classic KS is the most indolent form of the disease and typically is characterized by slow-growing, violaceous plaques on the lower extremities of elderly men from the Mediterranean. It overwhelmingly affects men, with a male-to-female ratio of 17:1 [6]. Systemic involvement

is rare, and therefore aggressive therapy is not necessary [5].

AIDS-related KS is characterized by an aggressive course [7]. It typically presents as rapidly-expanding, violaceous nodules that favor the extremities, face, and oral cavity [5]. Over one-half of the affected individuals will develop internal organ involvement; the gastrointestinal tract and lungs are the two most common sites [7]. HAART therapy represents the first-line of therapy, and chemotherapy is added in cases of particularly aggressive disease or if there is visceral involvement [7].

Our patient, who first developed KS at the age of 34 and whose ancestry does not contain known Mediterrean or Jewish ancestry, does not fit any of the previously described four variants of KS. He is human immunodeficiency virus (HIV)-negative, with no known immunosuppression and was relatively young at the time of disease onset. The death of his wife to AIDS several years prior to his developing KS raises more questions about the puzzling etiology and transmission of HHV-8. His wife, who likely contracted HIV from her first husband, was not known to have KS.

Other instances of Kaposi's sarcoma in HIV-negative individuals have been reported, which includes a 2008 retrospective case series of 28 HIV-negative homosexual and bisexual men with KS [8]. Their clinical course resembled classical KS with a few exceptions: a younger mean age of disease onset and a possibly increased rate of developing a lymphoproliferative disorder [8]. Our patient has never engaged in homosexual or bisexual behavior, which distinguishes him from this cohort. Also, he differs from this cohort because of his extremely young age of presentation and his subsequent solid organ cancers (prostate and bladder).

There is another subset of classic KS that is characterized by childhood onset, which is believed to result from an inherited immune deficiency of a specific co-stimulatory receptor on activated T-cells that confers an high susceptibility to HHV-8 infection [7]. Therefore, it may be possible, that our HIV-negative and relatively young patient, who has suffered with recurrent KS for 20 years, may have an

inherited susceptibility to HHV-8, which accounts for his disease course. It may be that this unidentified inherited immune deficiency also accounts for his history of two other solid organ malignant conditions.

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