

Pyogenic granuloma-like Kaposi sarcoma: Report of two cases

Piyojenik granülom benzeri Kaposi sarkomu: İki olgu sunumu

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SUMMARY

Kaposi sarcoma is a vascular tumor associated with Human Herpes Virus-8 infections. Lesions predominantly present at mucocutaneous sites but may involve all organs. There are many kinds of Kaposi sarcoma variants. Pyogenic granuloma like Kaposi sarcoma is a new variant which protrudes from skin and resembles a pyogenic granuloma. We decided to report two cases of Kaposi sarcoma because there are only few case reports about pyogenic granuloma like Kaposi sarcoma in literature, which is usually misdiagnosed as pyogenic granuloma.

Keywords: Human Herpes Virus-8, kaposi sarcoma, pyogenic granuloma

ÖZET

Kaposi sarkomu Human Herpes Virüsü-8 enfeksiyonu ile ilişkili vasküler bir tümördür. Lezyonlar genellikle mukokutanöz bölgelere yerleşir. Ancak tüm organlarda görülebilir. Kaposi sarkomunun birçok varyantı vardır. Piyojenik granülom benzeri Kaposi sarkomu deriden kabarık olan ve piyojenik granüloma benzeyen yeni bir varyanttır. Literatürde, piyojenik granülom benzeri Kaposi sarkomu ile ilgili sadece birkaç olgu sunulduğu ve genellikle piyojenik granülom yanlış tanısı aldığı için Kaposi sarkomlu iki olguyu sunmaya karar verdik.

Anahtar sözcükler: Human Herpes Virüsü-8, kaposi sarkomu, piyojenik granülom

INTRODUCTION

Kaposi sarcoma (KS) is a low grade vascular tumor associated with Human Herpes Virus (HHV)-8 infections. KS lesions predominantly present at mucocutaneous sites but may involve all organs and anatomic locations¹. KS was first described by dermatologist Moritz Kaposi more than a century ago². Human herpes virus 8 (HHV8) was identified in KS lesions by Chang et al³ in 1994. Investigations have been focused on oncogenesis and cancer immunology up to now after study of Chang. KS is the most prevalent malignancy among patients with AIDS¹.

CASE 1

31-year-old man presented with a 1-year history of a purple colored lesion on his right foot. With the exception of recurrent bleeding with minimal trauma, he was asymptomatic. He was healthy. Laboratory tests were normal. Anti HIV was negative. Physical examination disclosed a 0.5x0.5 cm non-tender, skin colored exophytic nodule in the middle of 4x8cm purple colored patch (Figure 1). Biopsy was taken from nodule. Histopathological examinations showed increased vessels, mitotic figures in dermis, extravasated erythrocytes and oval atypical spindle shaped

neoplastic cells which has enlarged nucleus and PAS stained cytoplasm (Figure 2). HHV-8 (LNA-1), CD31, CD34, FACTOR-8 and PAS were positive for immunohistochemical staining (Figure 3).

CASE 2

81-year-old women presented with a 2-month history of a purple colored lesion on his right foot. With the exception of recurrent bleeding with minimal trauma, she was asymptomatic. She was healthy. Laboratory tests were normal. Anti HIV was negative. Physical examination disclosed two 0.5x0.5 cm non-tender, skin colored exophytic nodule on medial side of left leg (Figure 4). Biopsy was taken from nodule. Histopathological examinations showed increased vessels, mitotic figures in dermis, extravasated erythrocytes and oval atypical spindle shaped neoplastic cells which has enlarged nucleus and PAS stained cytoplasm (Figure 5). HHV-8 (LNA-1), CD31, CD34, FACTOR-8 and PAS were positive for immunohistochemical staining (Figure 6).



Figure 1: There is a 0.5 cm x 0.5 cm non-tender, skin colored exophytic nodule in the middle of 4x8 cm purple colored patch in right side of leg.

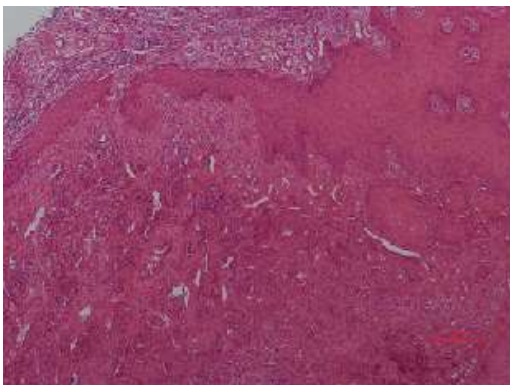


Figure 2: Histopathological examinations showed increased vessels, mitotic figures in dermis, extravasated erythrocytes and oval atypical spindle shaped neoplastic cells which has enlarged nucleus and PAS stained cytoplasm.

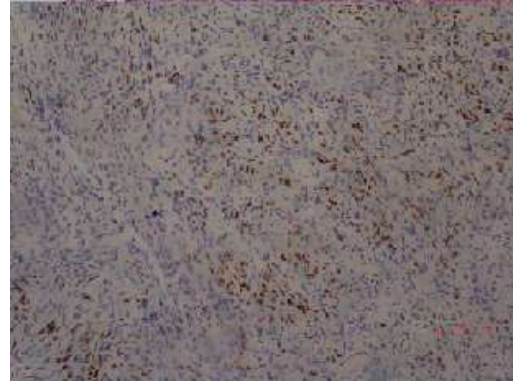


Figure 3: HHV-8 (LNA-1) was positive for immunohistochemical staining.



Figure 4: There are two 0.5 cm x 0.5 cm non-tender, skin colored exophytic nodules on medial side of left leg.

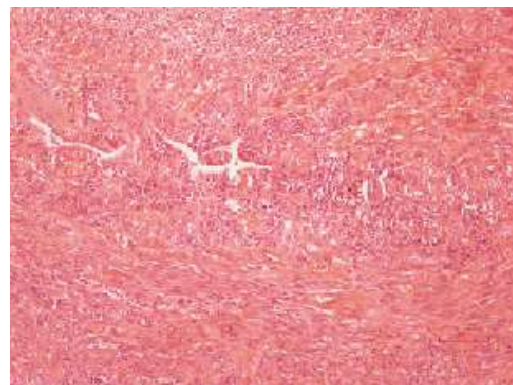


Figure 5: Histopathological examinations showed increased vessels, mitotic figures in dermis, extravasated erythrocytes and oval atypical spindle shaped neoplastic cells which has enlarged nucleus and PAS stained cytoplasm.

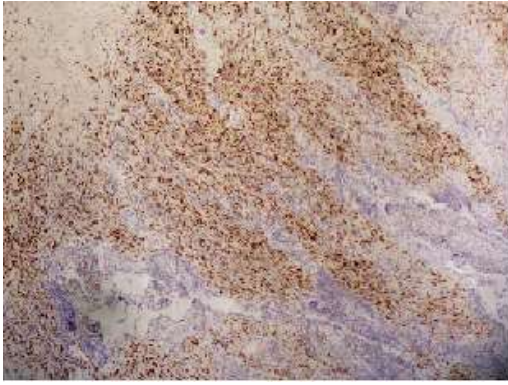


Figure 6: HHV-8 (LNA-1) was positive for immunohistochemical staining.

DISCUSSION

Proliferating KS spindle tumor cells are essential in pathogenesis and these cells are endothelial origin. Infection with HHV8 reprograms blood endothelial cells so that they resemble lymphatic endothelium and uncontrolled cells proliferate. HHV8 infection is essential but it alone appears to be insufficient for development of KS. KS growth involves up regulation of HHV8 gene products especially latency associated nuclear antigen (LNA-1)¹. HHV8 is detectable in more than 95 percent of lesions in KS⁴. KS is a low grade vascular malignancy that has been categorized into 4 epidemiological forms as classic, endemic (African), epidemic (AIDS associated) and iatrogenic (transplant associated)⁵. Classical KS is seen in mainly 40-70 years aged males of Mediterranean, Jewish and European origin. It predominantly involves in skin of lower extremities. African KS affects middle aged black adults and children from equatorial Africa. It is usually multifocal in skin and lymphadenopathic form is progressive. AIDS associated KS mainly affects homosexuals and IV drug abusers. It may disseminate to mucocutaneous sites and viscera. Lesions may regress with antiretroviral therapy. Iatrogenic KS affects immunosuppressed persons of any age from autoimmune disease, drugs or transplantation. It may regress after immunosuppression is discontinued¹. Early patch stage KS is characterized by abnormal vessels lined by thin endothelial cells in dermis. Proliferating vessels often surround larger ectatic vessels and skin adnexa and this is called promontory sign.

Chronic inflammatory cells, extravasated red blood cells and hemosiderin dyed macrophages are frequently present in lesions. Well-developed KS tumors consist of several fascicles of spindle shaped tumor cells. Eosinophilic and periodic acid Schiff (PAS) positive hyaline globules are common finding in lesions. Electron microscopy may show Weibel Palade bodies. Pleomorphic and significant number of mitotic figures are not very common¹. Histopathological investigations of nodular stage of KS shows atypical spindle cells with mitosis, slit-like vessels, diffuse extravasated red blood cells and hemosiderin. There are many kinds of KS variants. Anaplastic KS is an infiltrative, aggressive form including spindle cells without vascular spaces mainly seen in AIDS patients. Lymphangioma like KS consists of many ectatic lymphatics. Bullous KS is called so because of dermal and intradermal edema. Telangiectatic variant of KS contains large, congested ectatic vascular structures. When vascular structures are circumscribed it is called glomeruloid KS. Hyperkeratotic (verrucous) KS contains acanthosis, hyperkeratosis and fibrosis. In keloidal KS, there is dense, hyalinized collagen. In pigmented KS lesions there is increased numbers of melanin dyed dendritic cells. Pyogenic granuloma like KS is superficially located lesions that protrude from skin and resemble a pyogenic granuloma¹. Traumatized lesions may undergo ulceration and become inflamed so may misdiagnosed as pyogenic granuloma⁶. Wyatt et al⁷ described a case that was misdiagnosed as pyogenic granuloma of nasal mucosa. After further investigation they diagnosed it as pyogenic granuloma like Kaposi sarcoma. Fukunaga described a case report as Kaposi sarcoma like pyogenic granuloma. Histopathologic investigation of case showed distinctly exophytic growth, nodules, spindle cells, vascular structures, cellular atypical and mitotic activity⁸. This case resembles misdiagnosed KS. Because it consists of histopathologic features of pyogenic granuloma like KS. Scott et al⁹ described a 61 years old men presented with a protuberant growth on his foot resembled pyogenic granuloma and diagnosed as

pyogenic granuloma like Kaposi sarcoma. Ryan et al¹⁰ described 2 Kaposi's sarcoma like pyogenic granuloma which was HHV8 positive immunostaining. It is doubtful that these cases were Kaposi's sarcoma like pyogenic granuloma. The identification of HHV8 in lesions by using LNA-1 is the most diagnostic immunostaining technique in KS. It is seen as nuclear staining¹. Cheuk et al¹¹ studied 50 cases of KS and 53 cases of mimickers. %100 of KS was positive for HHV8 LNA-1 but %100 of mimickers were negative. This study showed immunostaining of LNA-1 exhibits high sensitivity and specificity for diagnosis of KS and is useful for distinguishing it from mimickers. Surgical excision, external beam radiation, laser therapy, cryotherapy, photodynamic therapy, topical tretinoin gel and intralesional vinblastine are choices of treatment. Systemic chemotherapy is used for widespread lesions. Treatment of underlying diseases affect course of KS.

KS may mimic many diseases especially pyogenic granuloma. Pyogenic granuloma like Kaposi sarcoma may misdiagnosed as pyogenic granuloma and treatment may be delayed. When lesions resemble pyogenic granuloma, pyogenic granuloma like KS must be in mind in because early diagnosis is important.

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