รายงานผู้ป่วยโรคมะเร็งคาโปซิซาร์โคมา

A Case Report: Disseminated AIDs-related Kaposi Sarcoma

สุทธิกิต พิกุลผล

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บทคัดย่อ

ภูมิหลัง: โรคมะเร็งคาโปซิซาร์โคมา (Kaposi Sarcoma, KS) เป็นเนื้องอกของเส้นเลือดที่เกี่ยวข้องกับเชื้อไวรัส Human Herpesvirus-8 (HHV-8) และพบได้บ่อยในผู้ป่วยที่ติดเชื้อเอชไอวีขั้นรุนแรง โดยเฉพาะในกลุ่มที่มีจำนวนเซลล์เม็ดเลือดขาวชนิด CD4 ต่ำ มะเร็งชนิดนี้สามารถเกิดขึ้นได้ที่ผิวหนัง เยื่อบุช่องปาก ปอด และระบบทางเดินอาหาร ซึ่งมีผลต่ออัตราการเสียชีวิตของผู้ป่วย

วัตถุประสงค์การวิจัย:เพื่อรายงานกรณีศึกษาผู้ป่วยเอชไอวีที่ได้รับการวินิจฉัยว่าเป็นมะเร็งคาโปซิซาร์โคมาแบบแพร่กระจาย และเน้นย้ำถึงความสำคัญของการวินิจฉัยโรคผ่านการตรวจรอยโรคที่ผิวหนังเพื่อป้องกันการแพร่กระจายของโรค

ระเบียบวิธีการวิจัย:นำเสนอกรณีศึกษาของผู้ป่วยชายอายุ 25 ปี ซึ่งมีอาการหายใจลำบากเรื้อรังเป็นเวลา 1 สัปดาห์ และพบรอยโรคที่เป็นก้อนในช่องปากร่วมกับผื่นสีม่วงบนผิวหนังเป็นระยะเวลา 1 เดือน การวินิจฉัยยืนยันโดยผลชิ้นเนื้อจากรอยโรคที่ผิวหนังและช่องปาก รวมถึงผลตรวจทางพยาธิวิทยาที่แสดงลักษณะเฉพาะของมะเร็งคาโปซิซาร์โคมา

ผลการวิจัย:รายงานผู้ป่วยเล่มนี้ เป็นรายงานผู้ป่วยเรื่องมะเร็งคาโปซิซาร์โคมาระยะแพร่กระจายในผู้ป่วยเอชไอวีจากโรงพยาบาลศูนย์ ในประเทศไทย ผู้ป่วยชายอายุ 25 ปี เป็นผู้ป่วยรักร่วมเพศ มาโรงพยาบาลด้วยอาการหายใจเหนื่อยมา 1 สัปดาห์ และมีก้อนบริเวณเหงือก ร่วมกับผื่นสีม่วงเป็นระยะเวลา 1 เดือน โดยผู้ป่วยรายนี้ถูกวินิจฉัยว่าเป็นมะเร็งคาโปซิซาร์โคมาระยะแพร่กระจาย หลังจากนั้นผู้ป่วยเสียชีวิตจากภาวะแทรกซ้อนที่เกิดขึ้นจากการเริ่มยาต้านไวรัสและการได้รับเคมีบำบัดที่โรงพยาบาลสงขลานครินทร์ โดยวัตถุประสงค์ของการเขียนเรื่องนี้เพื่อเป็นการคำนึงถึงการวินิจฉัยในการตรวจร่างกายทางด้านผิวหนัง เป็นส่วนช่วยในการวินิจฉัยถึงตัวโรคและป้องกันการนำไปสู่ระยะแพร่กระจายภายหลังได้หากวินิจฉัยได้เร็ว

สรุปผล:การตรวจพบรอยโรคที่ผิวหนังตั้งแต่ระยะแรกมีความสำคัญต่อการวินิจฉัยและป้องกันการแพร่กระจายของโรคมะเร็งคาโปซิซาร์โคมาในผู้ป่วยเอชไอวี นอกจากนี้ การเริ่มต้นการรักษาด้วยยาต้านไวรัสโดยเร็วมีบทบาทสำคัญในการลดอัตราการเสียชีวิตของผู้ป่วย

คำสำคัญ: มะเร็งคาโปซิซาร์โคมา, เอชไอวี, ผื่นสีม่วง, ยาต้านไวรัส, Kaposi Sarcoma

Abstract

Background: Kaposi sarcoma (KS) is a vascular tumor associated with the Human Herpesvirus-8 (HHV-8) virus and is common in patients with advanced HIV infection, especially those with low CD4 lymphocyte counts. It can occur in the skin, oral mucosa, lungs, and gastrointestinal tract, which has a high mortality rate.

Objective: To report a case study of an HIV patient diagnosed with invasive Kaposi sarcoma and emphasize the importance of diagnosis through skin lesion examination to prevent disease spread.

Method: We present a case study of a 25-year-old male patient who presented with chronic shortness of breath for 1 week and a purplish lesion in the oral cavity with skin rash for 1 month. The diagnosis was confirmed by biopsy results of skin and oral lesions and pathological examinations that showed characteristics of Kaposi sarcoma.

Results: This case report is a case report of invasive Kaposi sarcoma in an HIV patient from a regional hospital in Thailand. The patient was a 25-year-old homosexual male. A patient came to the hospital with shortness of breath for 1 week and a lump on the gums, along with a purple rash for 1 month. This patient was diagnosed with metastatic Kaposi's sarcoma. Later, the patient died from complications resulting from starting antiretroviral therapy and receiving chemotherapy at Songklanagarind Hospital. The purpose of writing this article is to consider the diagnosis in skin examination as a part of helping in diagnosing the disease and preventing it from spreading later if diagnosed early.

Conclusion: Early detection of skin lesions is important for diagnosing and preventing the spread of Kaposi's sarcoma in HIV patients. In addition, early initiation of antiretroviral therapy plays an important role in reducing the mortality rate of patients.

Keywords: Kaposi's Sarcoma, HIV, Purple rash, Antiretroviral drugs, Kaposi Sarcoma

Introduction

Whereas Kaposi sarcoma (KS) is a cancer that develops from vascular and lymphatic neoplasm originated by human herpesvirus-8[1]. It usually occurs as a tumor on mucocutaneous surfaces such as in the oral cavity and also develops in other parts of the body, such as lymph nodes, the lungs, or the digestive system [2]. There are several subtypes of KS, including epidemic KS or AIDS-related KS, classic/Mediterranean KS, endemic African KS, and transplant-associated KS. The most common type is epidemic KS, which affects HIV-positive patients [1]. The AIDS-related KS is an AIDS-defining malignancy and has rapidly progressive disease with early mucosal manifestation and systemic involvement respectively. The overall KS in general population was 1.53 per 100,000 persons. For HIV-infected patient's incidence was 481.54 per 100,000 person-years whearas HIV-infected men who have sex with men (MSM) had the highest incidence of KS (1397.11 per 100,000 person-year) [3]. Due to incidence of KS in Asia is very low, so early diagnosis of KS from skin lesion is considerable benefit because it leads to early treatment and decreases the mortality rate. In this report, we present a case of a 25 years old male presented with progressive dyspnea and multiple cutaneous violaceous plaques and mass at gum.

Case report

A 25-year-old male who is homosexual presented with progressive dyspnea and nonproductive cough for 1 week prior to admission at tertiary care hospital, Thailand. He had a clinical presentations of weight loss 10 kilograms within 6 month), multiple asymmetrical pigmented skin lesions involving bilateral thigh, groin, trunk and back. Moreover, he had a gingival mass for 1 month. Additionally, he had history of multiple sexual male partners.

Well-definedhemosiderin-ladenhyper-keratinizing Physical examination showed mark pale conjunctiva and crepitation in both lungs. Cutaneous examination revealed multiple discrete violaceous macules, papule and plaque size about 1x0.5 to 2 cm over trunk, back and bilateral thigh, pruritic popular eruption at bilateral legs (Figure 1-2). Oral cavity showed well defined- purplish mass at upper gum size 2x3 cm and tiny one at soft palate (Figure 3). For laboratory test, complete blood count showed hemoglobin 5.6 g/dL, platelet 114,000 cell/uL White blood cell 9000 cells/uL, Neutrophil 79 %, Lymphocyte 15 %, CD4 count 26/mm3 (1.85%), Viral load count of 101,000 copies/mL and; radiology – bilateral reticulonodular infiltration both lung on chest x-ray (Figure 4). Histopathology for skin revealed slit-like and sieve-like vascular proliferation with bland endothelial cells, extravasated red blood cells, hemosiderin laden macrophages with few plasmas cell infiltration and positive for CD 34. Histopathology of the gingival mass biopsy revealed hyper keratinizing stratified squamous epithelium with irregular elongation of rete ridges and large areas of ulceration. Underlying fibro collagenous connective tissue demonstrate numerous dilated thin-walled vascular channels. The specimen is notable for the increased proliferation of spindle-shaped cells with slit-like vascular spaces, containing extravasated erythrocytes. Increasing mitotic activity is seen. Tumor cells extend to all margins. Tumor cells were positive for CD34 and HHV-8 immunohistochemical strain. Bone marrow immunohistochemistry studies showed CD34 positive 2%, CD138 positive 15%, kappa positive, lambda positive - plasmacytosis, no light chain restriction. A CT scan of the chest showed multiple ill-defined nodules and soft tissue thickening at peribronchovascular regions both hemithorax (mainly at superior and lateral basal segment right lower lung, size about 5.0*8.0 cm), ground glass opacity at right lower lung, small lymph nodes at right upper paratracheal, paraaortic and subcarinal regions and multiple small hypodense nodules both hepatic lobes (size about 0.3-2 cm). A bronchoscope for tissue pathological diagnosis at the right upper lung reported kaposi sarcoma.

Hospital Patient was treated by HAART; tenofovir, emtricitabine and efavirenz at tertiary care hospital and then transferred to PSU hospital. A multidisciplinary team decided to give chemotherapy AS Paclitaxel weekly. During admission in PSU hospital, he developed hospital acquired pneumonia (HAP) and then Ventilator acquired pneumonia (VAP) with septic shock respectively. Advanced care plan was discussed with the patient's family after clinical deterioration as mentioned, and the decision for palliative care was made. Consequently, the patient died after receiving HAART and 2nd dose of chemotherapy.

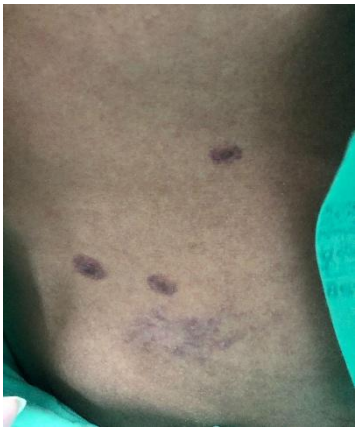
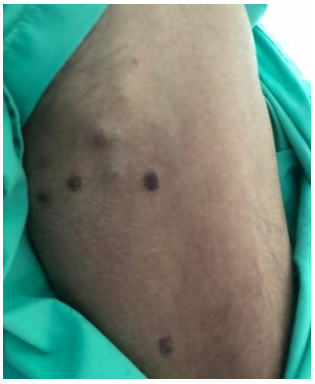


Figure 1 Violaceous macule and patch at trunk.



papulesFigure 2: Multiple discrete violaceous macules, papule, and plaque at the groin.

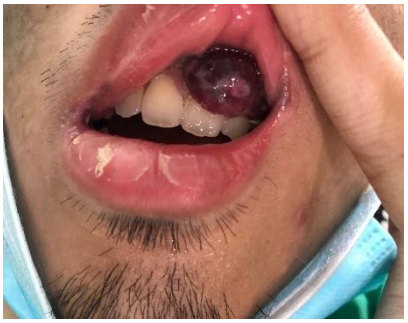


Figure 3: Well, defined- purplish mass at upper gum

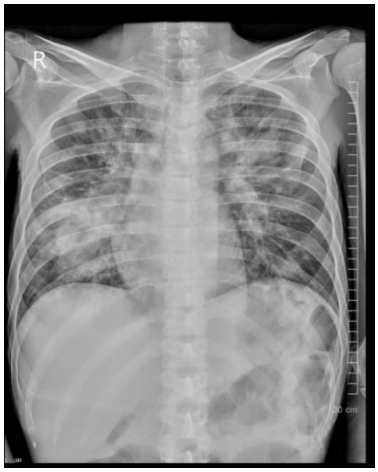


Figure 4: Bilateral reticulonodular infiltration both lung

Discussion

Kaposi sarcoma was first described in 1872 by Pathologist, Moritz Kaposi [4]. It's associated in HIV patient which CD4 count ≤ 100 cell/mm3 and high viral load count > 10,000 copies/mL [5]. Kaposi sarcoma is varied in clinical presentations; mucocutaneous or visceral organ, especially in AIDS-related KS typically presents as more widely distributed lesion (e.g. lung, digestive system). The skin lesion can appear most often at lower extremities, face, oral mucosa and genitalia. Extracutaneous spreading is also common, 15% of early presentation of extracutaneous is oral cavity affected at palate and gingiva respectively. Digestive system presented 40 % of initial diagnosis and followed by pulmonary involvement quite common [6]. The diagnosis of KS is confirmed by a skin biopsy of a suspected lesion [7].

Treatment of AIDS-KS is early initiate combination antiretroviral (cART) or HAART for allowing the immune system to recover and reducing HHV-8. Whearas early starting antiretroviral is goal of treatment, but it can increase occurring immune reconstitution syndrome (IRIS) and infection after that. In case of aggressive/disseminated KS have a complex treatment between HAART and divided to local and systematic treatments, such as chemotherapy and/or immunotherapy. Liposomal anthracyclines and paclitaxel have been approved by FDA as first line and second line mono-therapy, respectively. Interferon-alpha (INF-) is the only immunomodulant agent to have shown a therapeutic effect. Chemotherapy and/or immunotherapy dose not cure. Only for regression of disease and rapid response especially in visceral kaposi sarcoma, used Liposomal anthracyclines and paclitaxel have been approved by FDA as first line and second line mono-therapy, respectively [8,9].

AIDS-KSs achieve suppressing the immune system and HIV replication with HAART for first line treatment among KS lesion were found. In some case has a progression after HAART treatment, we termed KS immune reconstitution syndrome (KS-IRIS) [10]. Disease can progress in lesion and wide spread to the visceral organ too. Then disease will slowly regress. HAART was started and immune suppressed with low viral count. In KS-IRIS who commence ART but still progress disease, have report short term chemotherapy (Doxorubicin) co-treatment with ART and short course of corticosteroid to treat IRIS with successful response [11]. After treatment of HAART 6.6 % of patient developed progressive KS and also occurs in patient with higher CD4 count. However, patient who developed IRIS associate KS should get successful HAART [12].

Response for early treatment kaposi sarcoma associated with reduction of incidence rate finding's kaposi sarcoma in patients, especially in Visceral-KS (>50%) and nonVisceral-KS [>30%] respectively [13]. So, we should early initiate antiretroviral drug if there is no contraindication in every patient with HIV infection to prevent the Kaposi sarcoma progress and maintain high CD4 count for essential decline of disease [14].

Conclusion

Disseminated Kaposi sarcoma is high mortality rate disease. Nevertheless, diagnostic delay can lead to more spreading, which can increase risk of developing IRIS and others infection, although treatment of KS is early HAART and considered chemotherapy and radiation in any advanced case which can lead to IRIS and infection but there is more benefit than delay HAART. One clue of diagnosis HIV-KS is cutaneous lesion (Violaceous patch/plaque on body) is early detectable and important to considered to be diagnosed of KS in HIV- infected patient who may be presented with others symptoms.

complications In summary, we present a 25 years old man with HIV infection with low CD4 count and high viral load was diagnosed with disseminated kaposi sarcoma with cutaneous lesion, pulmonary and digestive system involvement that mean poor outcome. Complication after treatment occurred in this patient and resulting death. However, patient should get HAART and chemotherapy to reduce risk of mortality, then beware about KS-IRIS and complication.

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