

AIDS-related-Kaposi's sarcoma: A case report.

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ABSTRACT :

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Kaposi's sarcoma (KS), the most common neoplasm in patients with AIDS, is classified into four clinico-epidemiological types. HHV-8 infection precedes the development of all variants of KS. AIDS-related KS is one of these variants. The clinical presentations typically begin as purplish macular and papular lesions that progress to plaque-like or nodular tumors. Lesions usually occur on the head, face and upper trunk. Skin biopsy is crucial to make a correct diagnosis of KS.

We report a 30 year-old man with violaceous plaques on the face. The histology revealed numerous dilated and congested blood vessels with some slit-like vascular channels infiltrating among the collagen bundles. On the immunohistochemical examination, CD 34 staining was observed in a very large number of vessels.

Key words: AIDS-related Kaposi's sarcoma, HHV-8, CD 34

บทคัดย่อ:

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**ภาควิชาพยาธิวิทยา มหาวิทยาลัยสงขลานครินทร์

Kaposi's sarcoma (KS) ซึ่งเป็นมะเร็งที่พบบ่อยที่สุดในผู้ป่วยเอดส์แบ่งตามลักษณะทางคลินิกและระบาดวิทยาเป็น 4 กลุ่มย่อยพบว่า HHV-8 เป็นสาเหตุของโรคนี้ KSที่เกี่ยวข้องกับโรคเอดส์จัดเป็นกลุ่มย่อยหนึ่งลักษณะทางคลินิกจะเริ่มต้นด้วย เป็นผื่นราบหรือตุ่มนูนสีม่วงซึ่งต่อมาจะเจริญขึ้นเป็นผื่นนูนหรือก้อนนูนออก มักพบบริเวณใบหน้า ศีรษะและลำตัวส่วนบน การตรวจลักษณะทางพยาธิวิทยาของผิวหนังมีความสำคัญในการวินิจฉัยโรค

ผู้เขียนรายงานผู้ป่วยชายอายุ 30 ปี มีผื่นสีม่วงที่ใบหน้า ลักษณะทางจุลพยาธิวิทยาเข้าได้กับโรคKS การตรวจทางอิมมูโนฮิสโตเคมี พบมีการย้อมติดซีสต์สามสิบสี่ที่ผนังของหลอดเลือด

คำสำคัญ : Kaposi's sarcomaภาวะเอดส์, ซีสต์สามสิบสี่

Introduction

Kaposi's sarcoma is the most common neoplasm encountered in HIV patients and disproportionately involved men who have sex with men. KS is classified into four clinico-epidemiological types. Human herpesvirus 8 (HHV-8) is present in all KS variants. Patients with AIDS-related KS often have multifocal cutaneous diseases. Lesions may arise quickly in a few days, beginning as macules, papules, plaques and tumors. KS course ranges from indolent with only skin manifestations to rapidly progressive with visceral diseases. Though its dermatological manifestations can be alarming, the involvement of visceral organs is more commonly life-threatening.

Case report

A 30 year-old man, recently diagnosed with HIV infection, was admitted to Hadyai hospital because of

progressive dyspnea. He had an occasional productive cough. He did not complain of chest pain, hemoptysis or abdominal pain. For 1 week, he had noticed a purplish rashes growing on his face. He is a heterosexual man and denied using illicit drugs or receiving blood transfusion.

On admission, he was conscious. The temperature was 38 c°, the blood pressure 100/60 mmHg, the pulse 95 beats per minute, and the respiratory rate of 30 breaths per minute. The pupil were 3mm in diameter and reactive bilaterally. Oral cavity examination revealed oral thrush coating the posterior pharynx. No lymphadenopathy was detected. Chest auscultation showed fine crepitation of both lungs. Dermatological examination revealed multiple well- defined violaceous plaques in various sizes, ranging from 1 cm to 8 cm, on the face (Fig.1). Histopathological examination of a skin

biopsy taken from a nose lesion revealed numerous dilated and congested blood vessels with some slit-like vascular channels infiltrating among the collagen bundles of the upper and lower dermis (Fig.2A,2B). On immunohistochemical examination, CD 34 positive staining was observed in the wall of a very large number of blood vessels (Fig.3). His chest X-ray showed a “ground glass” appearance. Computed tomography (CT) of the brain was done due to decreased level of conscious. The result was normal. After 3 days of admission, his condition deteriorated rapidly and he expired from respiratory failure. His family denied an autopsy.

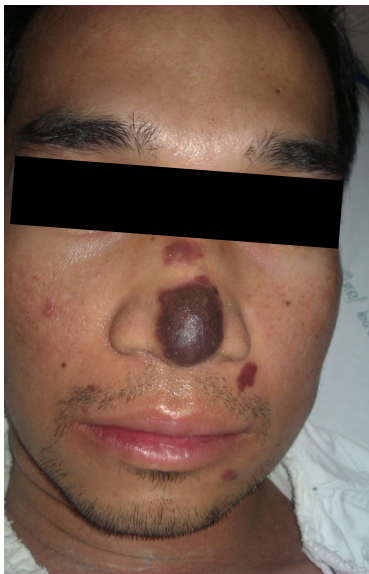


Figure 1 Physical examination show multiple well-defined violaceous plaques in various sizes ranging from 1 cm to 8 cm on face.

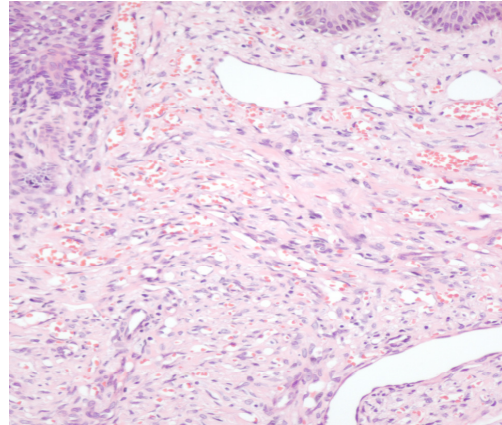
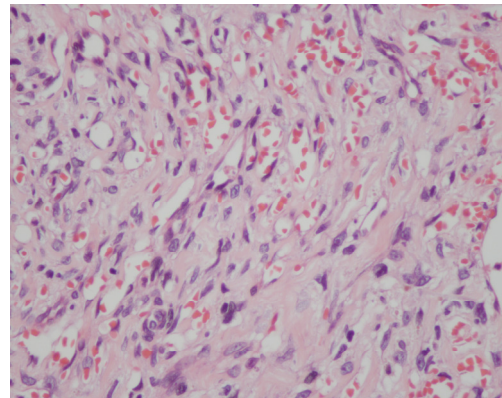


Figure2A, 2B Numerous dilated and congested blood vessels with some slit like vascular channels among the collagen bundles. (H&E10x,20x)

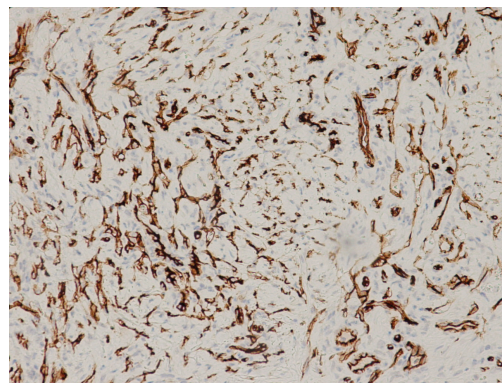


Figure3 Positive staining for CD34 was observed in the wall of very large number of blood vessels.

Discussion

Kaposi's sarcoma (KS) is the most common vascular neoplasm affecting people with HIV infection. The skin is the most common site of involvement. However, KS can also involve visceral organs such as the lungs, leading to severe morbidity and contributing to death in almost 30% of patients with the acquired immunodeficiency syndrome (AIDS)¹.

In 1994, a novel virus was discovered and termed human herpesvirus 8 (HHV-8), which can be found in all types of KS. The mode of transmission of HHV-8 remains unclear. Possible routes might be via sexual, horizontal, and parenteral transmission. In the USA, unsafe sex among homosexual men may be an important route of transmission². The presence of HHV-8 antibodies in infants suggests the transmission from mother-to-child³. The oropharynx is the site of strongest viral replication and a high level of HHV-8 copies was detected in the saliva. Before the HAART era, oral KS was the first clinical manifestation in about one quarter of AIDS patients⁴.

KS is classified into four clinico-epidemiological types: classic KS, African-endemic KS, immunosuppressive drug-related KS and AIDS-related KS^{5,6}.

Mucocutaneous lesions initially presented as macular and papular lesions and progress to plaques or nodules. AIDS-related KS differs from classic KS by its more rapid course and rapid multifocal dissemination. It seems to have a predilection for the upper body, head,

and neck and tends to flare after intercurrent illness (eg. opportunistic infections). AIDS-related KS causes significant morbidity due to dysfunction of visceral organs such as the gastrointestinal, pulmonary, and lymphatic system. Brain and intraocular lesions are extremely rare, probably because of the lack of lymphatics⁷. Post-mortem studies suggested that more than 25% of patients with AIDS-related KS also have visceral lesions. The most commonly involved sites are the stomach, bowel, liver, spleen, and lungs⁸. Common causes of death are intercurrent illness and systemic involvement especially of the pulmonary system⁹. Pulmonary KS is rapidly fatal if left untreated. Presenting symptoms include dyspnea without fever and occasionally hemoptysis. Chest radiographs typically demonstrate diffuse, reticulonodular infiltrates and sometimes pleural effusions¹⁰. Sequential thallium and gallium scans help to differentiate KS from other pulmonary lesions in AIDS patients. Positive thallium and negative gallium uptake were highly sensitive and specific findings in pulmonary KS¹⁰. However, bronchoscopy with transbronchial biopsy is the most appropriate method for the diagnosis of pulmonary KS.

The histologic spectrum can be divided into stages corresponding to the clinical findings. In patch stage KS, a proliferation of small, irregular and jagged endothelium-lined spaces surrounding normal dermal vessels is characteristic¹¹. Plaque stage KS represents the expansion of a spindle cell vascular process throughout the entire dermis. The vessels show variable

morphology, some as canalized cords and some showing lymphatic-like features. In nodular stage KS, characteristic feature is a honeycomb-like network of blood –fill spaces¹². The histopathological examination of our patient revealed numerous dilated and congested blood vessels with some slit-like vascular channels infiltrating among the collagen bundles, compatible with plaque stage KS. Most of the spindle cells in KS lesions express endothelial markers including CD34¹³ similar to those found in our patient.

In AIDS-related KS, HAART, in concurrent with or without chemotherapeutic agents, was reported to induce partial and complete remission in 16% and 46% of patients respectively¹⁴. Co-administration of HAART and pegylated liposomal doxorubicin was considered as the first line of therapy in one report¹⁵. Paclitaxel was an alternative agent that can induce remission in persons who relapsed after pegylated liposomal doxorubicin therapy. Our patient was dead from respiratory failure. Unfortunately, bronchoscopy and transbronchial biopsy were not done before and his family denied the autopsy.

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