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內文：

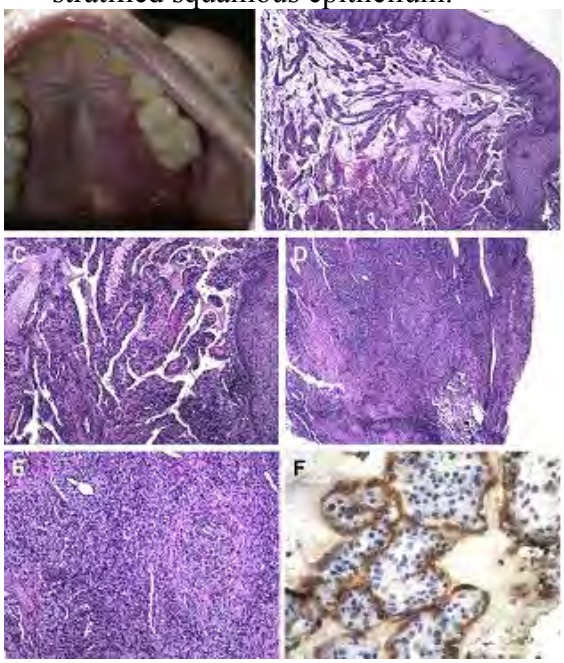
Introduction

- Lymphangioma-like KS (LLKS) is a rare histological variant of KS occurring in skin.
- Four different clinical forms of KS have been described.
- The classic form occurs more commonly in Poland, Russia, and Italy in lower extremities of elderly males.
- An endemic African form of KS affects children and young adult males in Africa.
- Another form of KS occurs in renal transplant patients, known as iatrogenic (or transplant-associated) KS.
- AIDS-associated form of KS has also been described, most commonly affecting human immunodeficiency virus (HIV)-infected individuals.
- All 4 clinical forms of KS, the lesions go through similar histological evolvement. The lesion progressively evolves from patch to plaque to nodular stages.
- The patch stage, normally seen in early developing KS,presents with flat macules. At this stage, the lesion histologically shows proliferation of new small blood vessels around larger dilated vascular spaces.
- In more established plaque lesions, the vascular proliferation
  Involves the dermis almost completely with a spindle cell proliferation limited to areas around proliferating vessels, resulting in a slightly elevated skin lesion.
- The nodular stage presents as a spindle cell lesion with slit-like vascular spaces.
- classic KS, who later developed fluid-containing bullae on their lower extremities. This presentation was later described as “bullous lesions” and was believed to be characteristic of lymphangioma-like
  KS (LLKS), it often presented as multiple bullae-like lesions on the skin.
- With the absence of a unique clinical presentation and a prog-nostic difference, LLKS is described as a histological variant, rather than a clinicopathologic entity.
- LLKS in the skin has been reported most frequently in older men between 59 and 80 years of age and most often in the skin of the lower extremity and more frequently in females
- The histological features of LLKS vary considerably from the traditional KS
- They consist predominantly of dilated vascular spaces, dissecting the dermal fibrous connective tissue stroma with a delicate strand-like papillary architecture
- The endothelial cells lining the vascular spaces are bland, imparting a lymphangioma-like appearance
- LLKS may share histological features with other vascular tumors including lymphangioendothelioma, hemangioendothelioma, and low-grade angiosarcoma.
- Similar to conventional KS, the identification of HHV8 in lesional tissue is diagnostic for the LLKS variant as well.
- Both endothelial markers (CD31 and CD34) and lymphatic markers (D2-40) are

expressed in KS and its histological variants like LLKS.

Case1

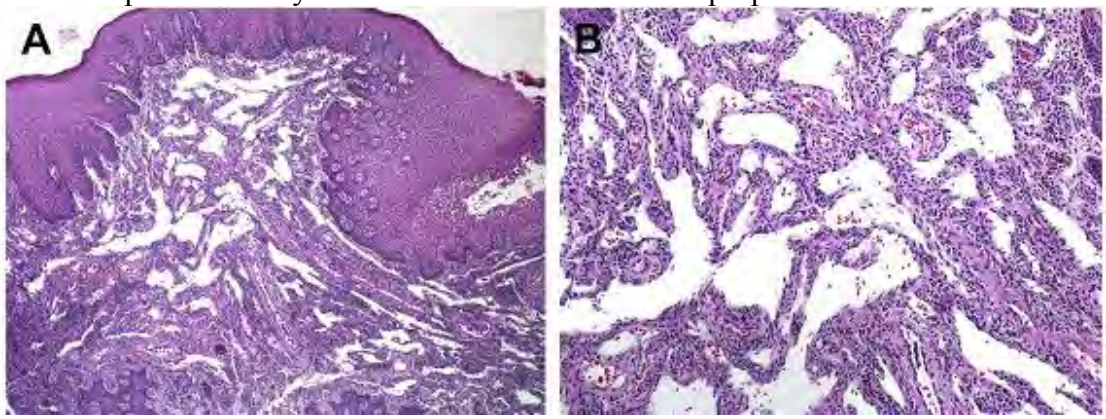
- A 45-year-old male patient presented with a diffuse erythematous swelling of the right palate and tuberosity area that felt 'boggy' on palpation
- A biopsy of the erythematous area showed mucosa lined by parakeratinized stratified squamous epithelium.



- Immediately underneath the epithelium, there were numerous dilated, anastomosing vascular spaces arranged in an edematous fluid-filled background in the lamina propria
- The vascular spaces were lined by endothelial cells with vesicular nuclei.
- Immunohistochemical studies for HHV8 and D2-40 were performed and the results revealed positive staining for HHV8 and D2-40
- A diagnosis of KS was therefore rendered on the basis of the positive HHV8 immunostaining in the nuclei of the lesional cells

Case 2

- A 45-year-old white male patient, who was HIV sero-positive, had periodontal disease and a nodular mass of the maxillary alveolar ridge and palate.
- Histopathological analysis of the biopsy specimen revealed dilated anastomosing vascular spaces lined by endothelial cells in the lamina propria

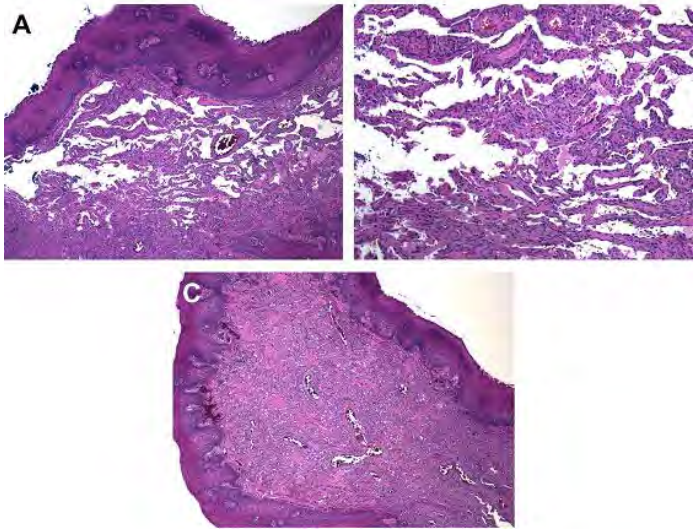


- The smaller vessels had a papillary architecture. Spindled cells, mitotic activity,and extravasation of erythrocytes were not seen.

- Immunohistochemistry showed positive staining for HHV8 in the lesional cells, and a diagnosis of LLKS was established.
- The positive immunostaining for D2-40 was observed only in the endothelial cells lining the lymphatic spaces and was less intense than in the other cases.

Case3

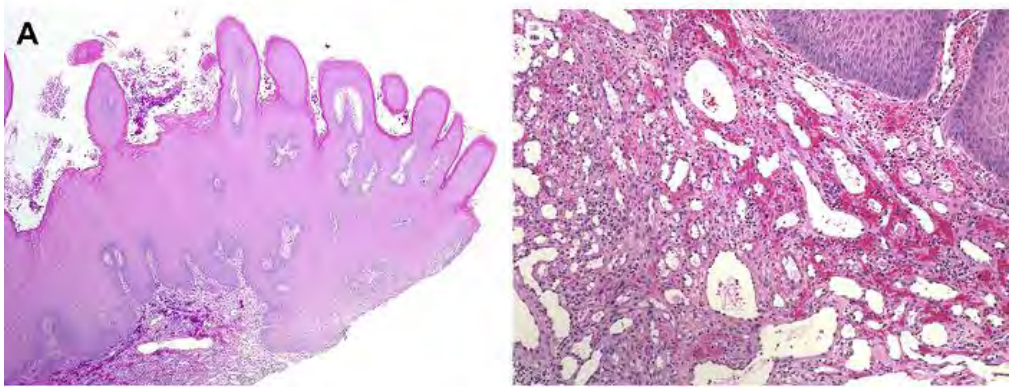
- A 37-year-old African American male presented with “granulation tissue” of the right and left retromolar areas.
- Panoramic radiograph demonstrated mandibular bone loss beneath the soft tissue swellings.
- there were areas showing large anastomosing vascular channels lined by endothelial cells with a bland cytomorphology



- There was also a focal area of spindle cells, which had occasional mitoses. A plasmacytic infiltrate was noted.
- An immunohistochemical stain for HHV8 was performed and it showed positivenuclear staining which confirmed the diagnosis of KS.
- D2-40 immunostaining also showed the presence of the protein in the endothelial cells lining the vascular spaces.

Case 4

- A 54-year-old male presented with a purple mass extending to involve the posterior hard palate, soft palate, and oropharynx.
- An incisional biopsy of the palatal lesion showed numerous dilated vascular spaces

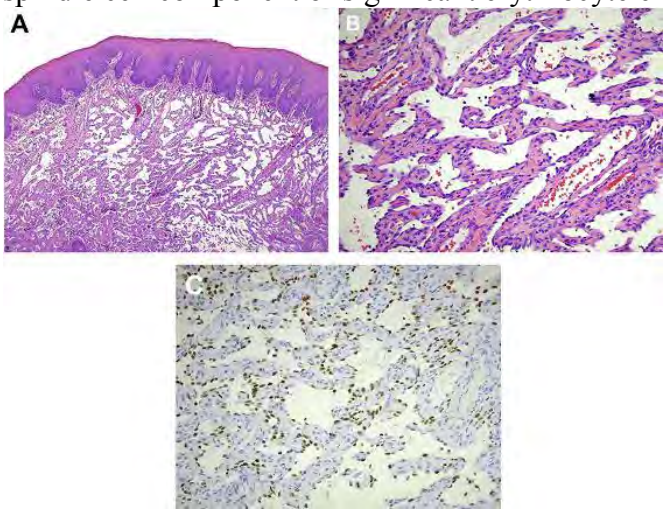


- This specimen demonstrated a prominent papillary epithelial hyperplasia.
- An immunohistochemical study for HHV8 showed positive nuclear staining in the lesional cells and confirmed the diagnosis of KS. Immunohistochemical results for

D2-40 also demonstrated positive staining in the endothelial cells.

Case 5

- A 35-year-old Caucasian male patient , presented with bilateral purplish nodular lesions in the palate
- Histological analysis revealed anastomosing dilated vascular spaces without a spindle cell component or significant erythrocyte extravasation



- Mitotic activity was sparse and no significant inflammatory component was seen
- A positive immunohistochemical reaction in the nuclei of the endothelial cells for HHV8 established the diagnosis of KS
- D2-40 immunostaining was also performed and observed to be positive in the endothelial cells lining the lymphatic spaces

Discussion

- LLKS has been described as bullous or cystic lesions as well as red or purple patches, plaques, or nodules on the skin
- The unusual histological pattern in the LLKS variant typically presents as ectatic, irregularly shaped vascular spaces that are present in the lamina propria right underneath the epithelium.
- The characteristic presentation is the anastomosing, interconnecting vessels that sometimes project like papillary tufts.
- The vascular structures are lined by banal endothelial cells, which have either plump and round or flat and elongated nuclei.
- The location of the lesion right underneath the epithelium, the absence of erythrocytes in many vessels, and the papillary strand-like architecture imparts a lymphatic vessel appearance to the lesion.
- A predominantly lymphoplasmacytic infiltrate has often been described in these lesions and is also seen in our intra-oral cases 1, 2, and 3.

Conclusion

- LLKS is a rare histological variant of KS that occurs most often in the skin of the extremities.
- We report 5 intra-oral cases demonstrating this histological pattern in which the typical KS features (spindle cell proliferation with erythrocyte extravasation) are not evident.
- It is important for pathologists to recognize this unusual variant of KS, especially when the characteristic spindle cell component of KS is not present or the HIV status of the patient is not known.

題號	題目
1	卡波西氏肉瘤（Kaposi sarcoma）與下列何種病毒的相關性最高？ (A)巨細胞病毒（cytomegalovirus） (B)單純疱疹病毒（herpes simplex virus） (C)人類乳頭瘤病毒（human papillomavirus） (D)人類疱疹病毒第 8 型（human herpes virus-8）
答案(D)	出處：Neville, Oral and Maxillofacial Pathology, 3rd ed, P. 557
2	Kaposi's sarcoma-associated herpesvirus 與下列何病毒在分類上最接近？ (A) Herpes simplex virus 2 (B) Epstein-Barr virus (C) Cytomegalovirus (D) Human herpesvirus 6
答案 (B)	出處：Neville, Oral and Maxillofacial Pathology, 3rd ed, P. 270