MRI findings of AIDS-related giant facial Kaposi’s sarcoma

A case report

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Abstract

Rationale: Kaposi’s sarcoma (KS) is the most common malignant tumor in HIV-infected people and occurs mainly in the skin, mucous membranes, and lymph nodes. Approximately 33% of the initial skin manifestations of AIDS and approximately 35% to 79% of KS occur during disease progression. Otherwise, AIDS-related facial KS that was simultaneously examined by magnetic resonance imaging (MRI) is rare.

Patient concerns: This case was a 30-year-old male homosexual, with left facial nodule for 14 months, and HIV infection was diagnosed 1 month previously. The patient was admitted to hospital because the nodule gradually expanded from 0.2 to 10.0 cm in diameter. Ultrasound examination showed edema of the subcutaneous tissue of the left facial mass, and the boundary was not clear between lesion and normal tissues. Magnetic resonance imaging (MRI) indicated that the left facial mass showed low signal intensity on T1-weighted imaging (T1WI) and T2-weighted imaging (T2WI), and a small amount of high signal intensity was seen in it. Diffusion-weighted imaging (DWI) and susceptibility-weighted imaging (SWI) showed low signal intensity. After enhanced scan, the tumor showed uneven enhancement.

Diagnoses: The pathological biopsy indicated KS.

Interventions: The patient began chemotherapy with the intravenous drip infusion of Doxorubicin Hydrochloride Liposome.

Outcomes: The facial KS decreased and the facial swelling was relieved.

Lessons: MRI could not only provide the diagnostic basis of KS for the therapy, but also could accurately determine the scope of the disease.

Abbreviations: AIDS = Acquired Immune Deficiency Syndrome, DWI = diffusion-weighted imaging, FLAIR = fluid-attenuated inversion recovery, HHV-8 = human herpesvirus 8, HIV = Human Immunodeficiency Virus, KS = Kaposi’s sarcoma, MRI = magnetic resonance imaging, SWI = susceptibility-weighted imaging, T1WI = T1-weighted imaging, T2WI = T2-weighted imaging.

Keywords: HIV, Kaposi’s sarcoma, magnetic resonance

1. Introduction

Kaposi’s sarcoma (KS) was first reported by Hungarian dermatologist Moritz Kaposi in 1872.\textsuperscript{1–4} KS is the most common malignant tumor in HIV-infected people, and the incidence of AIDS patients suffering from KS is 20,000 times that of the general population with normal immune status.\textsuperscript{15} It is commonly seen in the advanced stage of AIDS, more often in individuals with CD4+ T lymphocyte count <200/\mu L, and occasionally appears in the course of HIV antiretroviral therapy.\textsuperscript{16} Starting with antiviral therapy, the incidence of KS can be decreased significantly.\textsuperscript{17} The AIDS-related KS is associated with younger age, rapid progress, and high mortality. KS mostly originates from the skin, followed by mucosa, lymph nodes, and viscera. KS accounts for approximately 33% of the initial skin manifestations of AIDS and approximately 35% to 79% of KS occur during disease progression.\textsuperscript{18,9}

What we are providing is a rare case of a giant AIDS-related facial KS that was examined by magnetic resonance imaging (MRI).
small nodule in the left facial area with a diameter of about 0.2 cm that was initially red in color, then dark purple. Eight months ago, the nodule gradually enlarged and evolved into a purple-black nodule with a size of about 4.0 cm × 1.0 cm, with hard texture and no pain. Four months ago, the mass further enlarged to a size of about 10.0 cm × 10.0 cm, and the left eye could not be opened. Three months ago, the mass did not decrease by the anti-inflammatory treatment in another hospital. Two months ago, HIV preliminary screening antibody positive was found in other hospital. One month ago, the Center for Disease Control confirmed that the test was positive, with CD4 cells at 14 cells/μL. During this period, skin biopsy pathology in the stated hospital showed neoplasm, suspected to be angiosarcoma or KS. Twenty days ago, highly active anti-retroviral therapy (HAART) was initiated, and levofloxacin, vancomycin, and other anti-infective treatment were given, but the facial mass of the patient still enlarged. He had a history of homosexuality, and denied the history of blood transfusion, intravenous drug abuse, hypertension, coronary heart disease and diabetes, other infectious diseases, food and drug allergy, and surgical trauma as well as genetic diseases.

The patient came to our hospital for confirming the diagnosis and treatment. The body temperature of the patient was 37.5°C, with heart rate 90 beats/min, respiratory frequency 20 times/min, and blood pressure 115/80 mm Hg. Purple black bulge was visible in the left face, with no pain. The size of the bulge was about 10.0 cm × 10.0 cm, with the upper edge reaching the lower edge of the eyebrows, the lower edge to the lower lip level, the inner edge to the left edge of the nose, and the outer edge to the front hairline. The oppression of the skin mass made the left eyelid unable to be opened (Fig. 1). Purple nodule was seen in the right posterior neck, with a diameter of about 1.5 cm, with hard texture and no pain. Other physical examinations showed normal results. Laboratory examination showed anti-HIV antibody screening test (+), white blood cell of 3.95 × 10⁹/L, neutrophil of 40.5%, monocytes of 9.6%, eosinophils of 13.7%, C-reactive protein of 6.4 mg/L, fungi D-glucan of 26.27 pg/mL, Syphilis Toluidine Red Unhealed Serum Test response (−), CD4+ T lymphocytes of 20 cells/μL and CD4+/CD8+ ratio of 0.02. The face of the patient continued to swell, and the skin temperature increased. Staphylococcus chromogenes was produced in secretion culture. The patient was diagnosed with skin soft-tissue infections and was given norvancomycin anti-infection treatment, and the infection was gradually controlled.

**Figure 1.** The image shows a giant facial Kaposi’s sarcoma, and the oppressed eyelid cannot open.

**Figure 2.** The ultrasound examination shows the boundary of the Kaposi’s sarcoma is not clear, with the abundant arteriovenous blood flow signals (arrow).
Ultrasound examination showed edema of the subcutaneous tissue of the left facial mass. The thickest part was about 2.2 cm, without envelope. The boundary was not clear between the lesion and the surrounding normal tissues. Opaque dark area of fluid was not observed (Fig. 2). Color Doppler flow imaging presented the abundant arteriovenous blood flow signals in it. The left facial mass was suspected to be of inflammatory origin.

MRI showed the giant, irregular, abnormal signals in the left face with unclear boundary. The range was about 15.2 cm × 13.0 cm × 2.8 cm, with the upper margin of the mass to the frontal part, the lower margin to the mandibular angle level, the left margin to the outer ear and the right margin to the right side of the nose, and there was no invasion in the orbit. T1-weighted imaging (T1WI) and T2-weighted imaging (T2WI) of the tumor showed low signals, where a small amount of high signals could be seen. Both diffusion-weighted imaging (DWI) and susceptibility-weighted imaging (SWI) showed low signals, mixed with a small amount of high signals. Fluid-attenuated inversion recovery (FLAIR) showed no high signal. Edema signal could be seen at the edge of the tumor, where T1WI showed low signal intensity and T2WI showed high signal intensity. After enhanced scan, the tumor showed uneven enhancement, with unclear boundary. In brief, the sequences of T1WI, T2WI, SWI, and FLAIR except DWI could clearly exhibit the tumor size and the scope of invasion (Fig. 3). We made a diagnosis of the left facial KS.

Puncture biopsy of the facial swelling was performed. The pathological results showed the areatus spindle cell proliferation with vascular fissure formation in the dermis, red blood cell overflow, and hemosiderin deposition, with infiltration of a few inflammatory cells, which accorded with KS. Immunohistochemical results were Platelet endothelial cell adhesion molecule-1 (+), endothelial cell marker (+), human herpesvirus 8 (HHV-8) (+),

Figure 3. (A) T1WI; (b) axial T2WI; (c) sagittal T2WI; (d) fluid-attenuated inversion recovery; (e) diffusion-weighted imaging; (f) susceptibility-weighted imaging; (g) enhanced T1WI. Magnetic resonance imaging shows T1WI, T2WI, diffusion-weighted imaging and with susceptibility weighted imaging with low signal intensity (arrow) and a small amount of high signal intensity (arrowhead). Fluid-attenuated inversion recovery shows no high signal intensity. Edema signal intensity can be seen at the edge of the tumor (hollow arrow). The enhanced T1WI shows uneven enhancement (star), with unclear boundary.
And then, the patient began chemotherapy with the intravenous drip infusion of Doxorubicin Hydrochloride Liposome. The facial KS slightly decreased. One month later, the patient was hospitalized again for chemotherapy, without adverse reaction. The KS decreased significantly, the swelling was relieved, and the left eyelid could be opened (Fig. 5). The left eye movement was normal, the corneal reflex existed, and the left pupil was sensitive to light reflex.

3. Discussion
KS is a multiple idiopathic hemorrhagic sarcoma. The etiology of the disease is unknown and linked to genetic susceptibility, viral infections such as HHV-8, angiogenic cell differentiation, immune deficiency such as AIDS, immune status, racial inheritance, geographical location, and so on. KS is divided into 4 types: classical type, African local type, immunosuppressive type, and AIDS-related type. The classical KS progresses slowly, with a long survival period and

nuclear-associated antigen (about 1%+) and Factor VIII (+) (Fig. 4).

Figure 4. (HE × 200) The pathologic examination shows that the areatus spindle cell proliferation with vascular fissure formation, red blood cell overflow, and hemosiderin deposition.
relatively benign. Otherwise, the AIDS-related KS progresses rapidly, and has a high mortality. The lesions of classical KS are often composed of patches, plaques, and nodules, mainly at the end of the extremities (dorsum of hand and dorsum of foot and arch). The most common damage manifestations of AIDS-related KS are plaques and mild infiltrating plaques, mainly in a patient’s trunk and head and face.\(^\text{[10,20]}\) Children and young people are the groups at particular risk of the African-type KS, which often involved the internal organs with skin damage not commonly seen and is mainly manifested as the involvement of the lymph nodes.\(^\text{[10,20]}\)

Skin is an important organ involved in KS, with early lesions manifested as small papules and gradually formation of patches or nodules of different sizes, with light red color, which gradually evolves to purplish red and purple black.\(^\text{[21]}\) The patches, plaques, and nodules of KS mainly affect the lower extremities, the face, the trunk, the genital, and the mucous membrane of the oropharynx.\(^\text{[21]}\) Compared with HIV-negative patients, dermal outer layers or visceral KS lesions are more common in HIV-infected people.\(^\text{[22]}\) KS has been reported in almost all anatomical sites, and with the emergence of HIV. AIDS-related KS in the head and neck has become one of the most common manifestations of AIDS.\(^\text{[23]}\) However, reports on the giant facial KS are rare.

The pathological basis of KS is very complex, but it is generally believed that the tumor originates from endothelial cells, most probably from blood vessels. In different pathological stages of the disease, the manifestations are also different. The early manifestations are chronic inflammation or granulomatous inflammation with formation of new vessels and lymphatic vessels, and expansion, edema and bleeding with the lymphocytes, plasma cells and mast cells infiltrating around the blood vessels.\(^\text{[24–25]}\) The late manifestations are presented with the significant proliferation of endothelial cells, dermal spindle cells diffuse hyperplasia, extensive vasodilatation congestion. And the sieve-like structures are formed, red blood cells overflow, and hemosiderin deposition. The nuclear mitotic figures can be seen. Inflammatory cells infiltrate around them, with necrosis and fibrosis seen.\(^\text{[24–25]}\)

The imaging findings of facial KS in literatures are relatively rare. However, it has been reported in literatures that the KS distributes along the vessels of the infiltrated organs in the digestive system, with involved local lymph node swelling and uniformly increasing in density. The intestinal tract KS generally manifests as periodic and diffuse intestinal wall thickening with nodular enhancement, mesenteric lymph nodes enlargement and enhancement, suggesting that possible dissemination of KS.\(^\text{[26–27]}\)

MRI of liver KS shows high signal lesions infiltrate around the portal vein on T2WI, which, in the shape of grape clusters, are relatively characteristic. Liver and spleen enlarge, and the bigger lymph nodes are seen around the hepatic hilum and the celiac trunk.\(^\text{[28]}\) Pulmonary KS involves bronchovascular bundles, interlobular septa, pleura, and other interstitial tissues. Infiltration of adjacent lung parenchyma results in the formation of consolidation area of nodules or with indistinct boundary. CT shows the distribution along the bronchovascular bundles, most of which appear in bilateral symmetry, irregular flame-like nodule edges, and thickened ant leg-like interlobular septa. It may be accompanied with the masses enlarging and consolidation of the lungs, whereas the consolidation can be caused by tumor invasion, bleeding, or obstructive pneumonia.\(^\text{[29,30]}\)

In this case, the pathological results showed that the areatous spindle cell proliferation with vascular fissure formation in the dermis, red blood cell overflow, and hemosiderin deposition, which presented as low signal intensity with a small amount of high signals in T1WI, T2WI, DWI, and SWI of MRI. Moreover, the MRI could provide the images of multiplane, and the invasion scope of the KS. In this case, the scope of the KS showed by MRI was larger than appearance seen by eyes. Therefore, the MRI could provide the accurate scope of the KS for the better diagnosis. It was regrettable that the follow-up MRI had not been done. But we observed that the MRI was sensitive to the facial KS, so we inferred that MRI may be helpful to the KS for follow-ups.

In conclusion, the case of the giant facial KS being simultaneously carried out by MRI is rare although skin is the common organ involved in KS. Moreover, for the better therapy and follow-up, MRI can not only make a diagnosis of KS, but also can accurately show the invasion range of tumor.

**Author contributions**

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