# KAPOSI'S SARCOMA. A CASE REPORT

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## SUMMARY

**Aim:** The aim of this case report is to present the case of a patient with iatrogenic Kaposi's sarcoma afflicting several organs, ocular manifestation.
**Case report:** In a 74-year-old kidney transplant patient receiving immunosuppressive therapy, iatrogenic Kaposi's sarcoma (KS) developed in both lower eyelids. Subsequently, KS was confirmed in the region of the left forearm, with suspicion of lesions in the lungs. The ocular tumor was surgically removed with negative margins, requiring no further therapy. The lesion on the left forearm was completely excised. The patient underwent radiotherapy for the lung lesions, and immunosuppressive therapy was reduced.
**Conclusion:** The case highlights the importance of early identification of KS, its histological verification, radical resection, and multidisciplinary collaboration. Knowledge of the epidemiology of this condition is a key factor in determining the correct diagnosis.
**Key words:** Kaposi's sarcoma, radical resection, immunosuppression, multidisciplinary collaboration

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## INTRODUCTION

Kaposi's sarcoma (KS) is a malignant vascular tumor associated with the Human Herpes Virus 8 (HHV-8) [1,2]. The virus is transmitted predominantly in childhood through saliva, other vehicles of transmission may be blood, sperm or a transplanted organ [3]. This pathology primarily affects immunocompromised individuals [4]. We distinguish between 4 basic forms of KS – classic, endemic, iatrogenic and epidemic [5]. In previous years, before the spread of acquired immune deficiency syndrome (AIDS), it was considered a rare tumor, now its incidence is more frequent [6].

The tumor predominantly affects the skin and mucosa, but may occur in any organ [5]. In ocular localization it is generally situated on the eyelids or in the eye socket, though most often on the bulbar conjunctiva in the inner corner. It has the form of vascular lesions, and tortuosity of blood vessels and subconjunctival hemorrhages tend to be present in the surrounding area [7]. In ocular localization the main therapeutic modality is excision with negative margins, cryotherapy and radiation therapy. In all patients it is necessary also to consider the general condition of the immune system – to exclude HIV infection, and in the case of immunosuppressive therapy to consider its reduction [7–9].

## CASE REPORT

A seventy-four-year-old male patient was referred by a regional eye center to the Department of Ophthalmology at the University Hospital in Ostrava (UHO), with a 3-month growing lesion on the lower left eyelid, on which a biopsy was performed with a finding of Kaposi's sarcoma.

From the patient's personal medical history it was of fundamental significance for us that the patient had previously undergone a kidney transplant and had been treated with a combination of immunosuppressive drugs – mycophenolate, prednisone and tacrolimus. The patient was then monitored by internal medicine specialists for secondary arterial hypertension, and was a non-smoker. Other data from the patient's medical history were of less significance from an ocular perspective. Natural visual acuity in the right eye was 5/5 weak, in the left eye

5/5. Values of intraocular pressure were 19 mmHg in the right eye, 22 mmHg in the left eye. Upon examination of the patient, a lesion was found on the lower left eyelid corresponding to Kaposi's sarcoma with a size of 10 x 5 mm, on the lower right eyelid further new small reddish lesions of less than 5 mm in diameter, which were not present according to the report from the referring center, otherwise the anterior segment was without any remarkable features. Fig. 1.

The patient was referred to the Department of Oncology at UHO for completion of a basic examination with regard to generalization, and infection with the HIV virus and AIDS (Acquired Immune Deficiency Syndrome) were excluded. At the same time total excision of Kaposi's sarcoma with a safety margin was planned on the left eyelid, followed by reconstruction in a second session, and probatory excision of the lesion on the right eyelid. Fig 2.

The procedure was performed under local anesthesia according to the plan, Kaposi's sarcoma was histologically verified affecting the lower right eyelid, on the left also Kaposi's sarcoma, though the tumor here did not reach the margins of excision. A further suspected lesion was discovered during the examination on the upper left eyelid. In the operating theater a biopsy of the upper left eyelid was performed, with resection of the tumor on the lower right eyelid and reconstruction in a single session. With regard to the scope of the lesion on the lower left eyelid, the Hughes procedure was used as the most appropriate reconstruction technique [10]. Fig. 3.

The histopathological examination of the biopsy sample from the upper left eyelid did not demonstrate malignant structures, and it was further confirmed that Kaposi's sarcoma was removed from the lower right eyelid, with a negative safety margin. A division of the Hughes flap was planned on the left side. The planned division of the Hughes flap on the left eye took place entirely without complications, the oncologists did not recommend adjuvant therapy in the surrounding area of the eyes due to the locality and the performed R0 resection. At the final follow-up at the Department of Ophthalmology of UHO the anterior segment was pacific and the patient was referred to his local ophthalmologist for observation. Fig. 4.

At the oncological screening, a red lesion with a size of

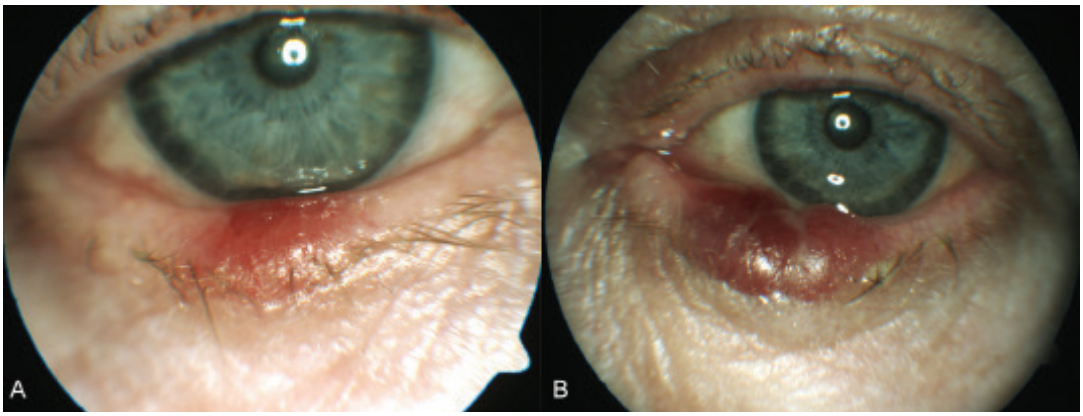


**Figure 1.** Findings on the eyelids of the right (**A**) and of the left (**B**) eye during the first examination at our clinic

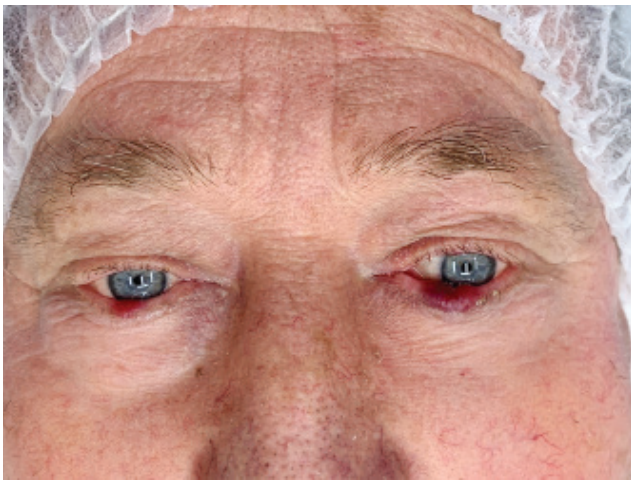

**Figure 2.** Photograph of the eyelids before performing total excision with a safety margin of Kaposi's sarcoma on the left and on the right before probing excision of the lesion

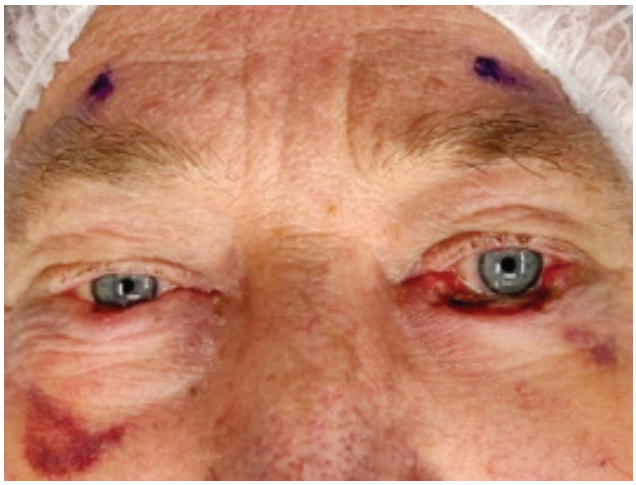

**Figure 3.** Condition after performing total excision with a safety margin of Kaposi's sarcoma on the left and on the right after probing excision of the lesion

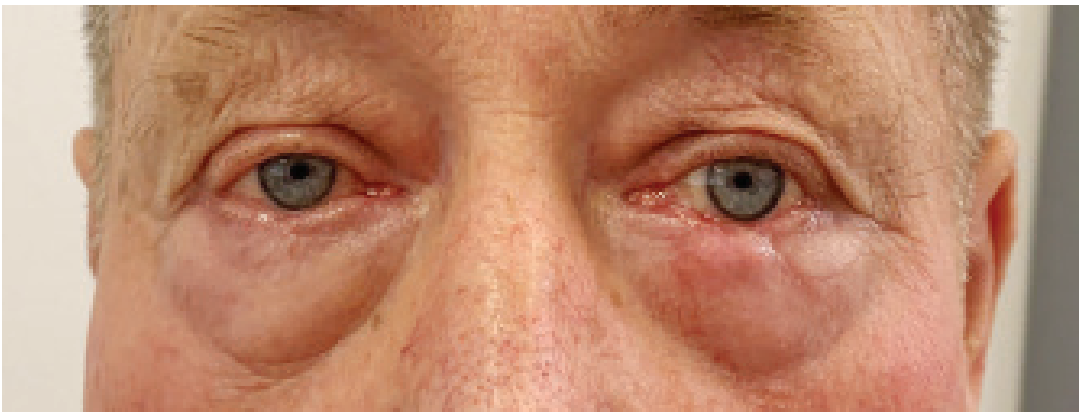

**Figure 4.** Findings at the last check-up – state after division of the Hughes flap on the left, on the right after resection of the Kaposi´s sarcoma with reconstruction at the same time

1 cm, which according to the patient was becoming progressively paler, was discovered as part of the physical examination. The client was referred to the Department of Dermatology of UHO, where excision was planned. An X-ray of the chest detected a suspected lesion in the upper field of the left lung, and a date was arranged for CT verification of this finding. At the same time the patient's general medication was adjusted, the immunosuppressants mycophenolate and prednisone were discontinued, and the patient was left on tacrolimus (mTOR inhibitor), the dose of which is regularly adjusted according to the patient's serum concentration. The patient then underwent a CT examination of the chest, which confirmed a lesion in the upper left field of the lungs, and histological verification was recommended, which the patient refused. The oncologists concluded that the patient was not a suitable candidate for general therapy, though navigated stereotactic radiation therapy of the lesions in the lungs with the aid of CyberKnife was planned. The dermatologists then performed excision of a lesion from the patient's left forearm, in which Kaposi's sarcoma was histopathologically confirmed. The patient is undergoing therapy with the aid of CyberKnife, follow-up CT is now planned in order to evaluate the result, so far the patient is without any finding of recurrence.

## DISCUSSION

Kaposi's sarcoma was first described in 1872 by Moritz Kaposi, an Austrian dermatologist of Hungarian-Jewish origin who defined it as an "idiopathic multiple pigmented sarcoma of the skin" [11]. For a long time this tumor was considered a relatively rare, slowly growing malignancy, appearing most commonly in men of middle and advanced age. This did not change until 1981, when a relationship was described between HIV infection and this sarcoma [12]. At the beginning of the 1990s a new virus was isolated from a Kaposi's sarcoma lesion, which was named as the Human Herpes Virus 8 (HHV-8) [2].

HHV-8 belongs to the Gammaherpesvirinae family. It is an oncogenic virus, and in addition to Kaposi's sarcoma may also cause Castleman disease or primary effusion lymphoma (both rare B-cell malignancies) [3]. The highest rate of seroprevalence is found in the "Kaposi's sarcoma belt" in sub-Saharan Africa, where it reaches as high as 86%. Europe manifests medium seroprevalence with 10–30%, in northern Europe and the USA seropositivity is low, around 6%. In these regions with low incidence of the virus, the highest values are recorded in HIV-positive men who have sex with men (MSM) and HIV-negative MSM [2]. In the regular population the virus is transmitted predominantly in childhood through saliva, transmissions have also been described within the family, most frequently from mother to child. Other vehicles of transmission may be blood, sperm or vaginal secretion. Last but not least a transplanted organ may be the cause of transmission; in these cases the risk of onset of Kaposi's sarcoma is significantly increased due to immunosuppressive therapy [3,13,14].

We distinguish between 4 basic forms of this pathology – classic, endemic, iatrogenic and epidemic. The classic form most commonly affects older men in the Mediterranean region or men of Jewish origin [2]. The course is usually indolent and the pathology is limited to lesions in the region of the lower limbs, while affliction of the mucosa or internal organs is rare. An interesting fact is that the risk of occurrence of the classic form of KS is 4x lower among cigarette smokers, though no causality has been confirmed. The endemic variant is generally described in sub-Saharan Africa, predominantly in children. This is a highly aggressive pathology with an unfavorable prognosis. The iatrogenic form occurs in patients following an allogeneic transplant, most often of the kidneys and in patients undergoing immunosuppressive therapy. Tumor lesions usually regress after reduction of the dose of immunosuppression. The epidemic variant of this pathology appears in HIV-positive patients, most commonly in homosexually active men, and is one of the three "AIDS-defining cancers" [2,5,8,15–18].

The tumor predominantly affects the skin and mucosa, though may occur in any organ [5]. In ocular localization it is generally situated on the eyelids or in the eye socket, but most often on the bulbar conjunctiva in

the inner corner. [7]. It is clinically manifested as a vascularized nodule of red, purple or brown color, and may be confused with a hematoma or nevus [6,7]. Sometimes, in the later phases, large exophytic, ulcerating and hemorrhaging nodules may occur, often in connection with painful edema. Its diagnosis is confirmed by biopsy [8]. Histologically the tumor is characterized by neoangiogenesis, spindle cells and inflammatory infiltrate [19]. In the disseminated variant of KS, the lungs, lymph nodes, gastrointestinal tract and nasal cavity are most frequently affected [20]. Simultaneous affliction of the lungs and skin with KS has already been described [21].

The treatment depends on the actual form of the pathology and its scope [2]. In ocular localization the main therapeutic modality is excision with negative margins, cryotherapy and radiation [7,9]. In patients with immune suppression the first step is to strengthen the immune system [8]. As regards patients with the epidemic variant (AIDS), it is essential to ensure optimization of antiretroviral therapy, in the case of dissemination then in combination with chemotherapy [2]. Patients with iatrogenic form may be treated by merely reducing immunosuppressive therapy, although this presents a higher risk of rejection of the transplant. The other therapeutic procedure for these patients is the conversion of the existing immunosuppressive therapy to mTOR inhibitors (e.g. sirolimus, tacrolimus), in which their antiangiogenic effect is utilized [8,22].

The case we describe here corresponds with the specialist literature. A patient following kidney transplantation, treated with immunosuppressants, developed KS affecting multiple organs, in which KS was detected on both lower eyelids, although the most frequent localization tends to be the inner corner or bulbar conjunctiva. We can only speculate as to whether the transplanted kidney was the vehicle of transmission of the Human Herpes Virus 8, since this transplant is not ordinarily examined for the presence of HHV-8. The patient reported to our clinic already with KS of the lower left eyelid confirmed by biopsy, and was immediately referred to the Department of Oncology with regard to the need to complete the examination for generalization of the pathology, and we excluded Acquired Immune Deficiency Syndrome. With regard to the fact that the treatment was being treated with a combination of immunosuppressants, from an epidemiological perspective we can state that in our client this concerned the iatrogenic form of Kaposi's sarcoma.

In the ocular localization adequate therapy was chosen, namely excision with histologically negative margins, which was performed promptly without waiting for the result of the oncological screening. Adjuvant radiotherapy was not recommended by the oncologist in this case. From the ophthalmological perspective this case can now be considered closed. In our patient a further spread of KS occurred also on the left forearm, this lesion was removed by dermatologists. The nephrologists reduced the patient's immunosuppressive therapy, discontinuing mycophenolate and prednisone, while leaving the patient on tacrolimus (mTOR inhibitor). With the aid of computer tomography, the oncological screening detected a lesion on the lungs, which was removed by stereotactic radiation therapy.

Our therapeutic procedure in this presented case report fully followed the recommendations laid down for the treatment of iatrogenic KS. From a general perspective our patient remains under outpatient observation, and is stable.

## CONCLUSION

In this case report we presented the case of a patient with iatrogenic Kaposi's sarcoma of the lower eyelids on both eyes, in whom the disease spread to the left forearm, with suspicion of spreading also to the lungs. The case highlights the importance of timely identification of the tumor, verification by biopsy, the performance of rapid oncological screening, radical resection and multidisciplinary cooperation. Knowledge of the epidemiology of this condition is a key factor in determining the correct diagnosis.

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