CASE REPORT

Bilateral auricular classic Kaposi's sarcoma

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SUMMARY

In 2009, a 57-year-old man was referred to our maxillo facial surgery department for the appearance of a reddish-purplish swelling on his right helix. The lesion was more than 1 cm in length and did not show changes for 3 months. He was otherwise healthy, without any predisposing factors. The patient denied smoking, alcohol misuse and intravenous drug use. He had no family history of similar lesions or Kaposi's sarcoma. The lesion was completely excised and the surgical defect reconstructed with a local flap. In 2011 the same patient returned to our attention because of the appearance of a new lesion, similar to the first one on the other auricle. The lesion was completely removed. The same situation recurred in September 2012, when the patient presented a new little reddish-purplish swelling of 3 mm on the left helix. The lesion was excised. The patient has not reported further lesion at present.

BACKGROUND

Kaposi's sarcoma (KS) is related to the human herpes virus 8 infection (HHV 8) but its presentation is not always caused by immunodeficiency and HIV infection. The classic form mostly affects elderly male patients, often with Mediterranean origins, without HIV infection. Although not related to immunodeficiency, classic KS (CKS) might behave as a systemic illness, and recurrences

occur even after complete excisions of primary cutaneous lesions. The follow-up of patients with CKS is mandatory and complete physical examinations should extend to unusual and secluded areas of the skin, such as the ears. This case represents the first literature report of a bilateral auricle CKS.

CASE PRESENTATION

In 2009, a 57-year-old man was referred to our maxillo facial surgery department for the appearance of a reddish-purplish swelling on his right helix. The lesion was more than 1 cm in length and did not show any changes for 3 months (figure 1). He did not report any suspicious lesion in any other parts of his body. Either lymphadenopathy or mucosal involvement was not present. He was otherwise healthy, without any predisposing factors. His medical records reported only a mild and well-controlled arterial hypertension. The patient denied smoking, alcohol misuse and intravenous drug use. He had no family history of similar lesions or KS. The lesion was completely excised and the surgical defect reconstructed with a local flap (figure 2) with satisfactory results (figure 3).

In 2011 the same patient caught our attention because of the appearance of a new lesion, similar to the first one on the other auricle (figure 4). The lesion was completely removed. The same situation recurred in September 2012, when the patient

Figure 1 Preoperative photograph showing reddish-purplish nodules on the helix of the right ear. These three lesions represent the first occurrence of classic Kaposi's sarcoma in this patient.

Figure 2 Intraoperative photograph showing the right ear after the resection, a bipedicle retroauricular flap was used for reconstruction.

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Figure 3 Postoperative photograph showing a good aesthetic outcome, with no local recurrence.

presented a new little reddish-purplish swelling of 3 mm on the left helix. The lesion was excised.

INVESTIGATIONS

Histological and immunohistological findings were consistent with a histological diagnosis of CKS. HHV 8 immunofluorescence testing on the specimen was performed. HHV 8 was detected in the lesional skin sample. Serological analysis resulted negative for anti-HIV antibodies and anticytomegalovirus IgM and IgG while the blood count was normal.

The histological diagnosis confirmed the recurrence of CKS and HHS 8 was found in all the lesional specimens.

Figure 4 Preoperative photograph showing a reddish-purplish nodule on the helix of the left ear. This small lesion represents the recurrence of the disease and it appeared 2 years after the first occurrence of the Kaposi's sarcoma.

DIFFERENTIAL DIAGNOSIS

KS cutaneous involvement may mimic other conditions such as pyogenic granulomas and vascular anomalies or pseudo-Kaposi lesions. Pseudo-Kaposi lesions appear as popular lesions arising in patients suffering from venous failure or arteriovenous fistulae. Only the histological examinations can provide a conclusive diagnosis.

TREATMENT

The first lesion was completely excised and the surgical defect reconstructed with a local flap.

Recurrent lesions, which appeared in 2011 and 2012, were excised and the little skin defect not requiring any local reconstruction was directly closed with a cutaneous suture.

OUTCOME AND FOLLOW-UP

During a 3-year close follow-up no involvements of visceral organs or mucosal areas, no changes in his health conditions and no HIV seroconversion were detected.

DISCUSSION

KS is a vascular neoplasm originating from vascular and lymphatic endothelia, characterised by a multifocal angiogenic process, frequently presenting as multiple vascular cutaneous and mucosal nodules. Nodal and visceral manifestations usually appear in case of severe immunosuppression.¹ Moriz Kaposi first described this neoplasia in 1872.¹

KS is associated with viral infection that shows different sociodemographical, clinical and histopathological features linked to different and unique biological behaviours. Four different variants of this tumour exist: epidemic or AIDS-associated KS (the most common type), endemic or African KS, iatrogenic KS (frequently posttransplantation) and Mediterranean or classic KS.² Clinical presentation may be highly variable. Typical lesions include reddish-purplish macules, papules or nodules. CKS mostly appears in elderly men of Mediterranean origin as multiple bilateral cutaneous lesions of the lower extremities.¹ Male-to-female ratios from 10:1 to 15:1 have been reported, but a lower gender difference characterises populations with lower incidence rates.³

Advanced age, diabetes and the use of corticosteroid medication represent risk factors for CKS development. Corticosteroid medications are also associated with CKS progression.⁴

This kind of lesions may be localised without nodal or visceral involvement or they may show aggressive behaviour with severe visceral disease. In contrast to AIDS-associated KS, KS in HIV-negative patients has a relative indolent course; it is mostly limited to the skin and appears better-responsive to local or systemic therapeutic strategies.³

A standard therapeutic guideline is not available; the treatment has to be chosen depending on the subtype and stage of the neoplasm as well as on the immune status of the single patient.³

HHV 8 has been identified in AIDS-associated KS as well as in CKS. HHV 8 is endemic among Mediterranean people but it may not necessarily cause any disease in immunologically competent individuals. Thus, this herpes virus is considered necessary but not sufficient for the development of KS.⁵ Cytomegalovirus has also been identified as an aetiological factor for the development of KS.²

To our knowledge, the authors present the first case of bilateral lesions of the helix of the ear in a middle-aged

HIV-negative man, without a history of any predisposing factors.

The involvement of the head and neck have been reported in only 14% of cases of CKS. Only four cases of CKS have been reported in the helix of the ear without a history of immuno-deficiency, but this one is the first with a solitary bilateral involvement of the external auricle.¹

Learning points

- ▶ A thorough physical examination is pivotal in all kind of Kaposi's sarcoma (KS), even secluded areas rarely affected by this condition, such as the external ear, can host bilateral primitive neoplasms and local recurrences.
- ▶ Always remember that KS can also show up in HIV-negative patients, in its "classic" form.
- ▶ Owing to its viral aetiology, KS might recur locally as well as at distance even after a radical excision.

Contributors GC performed the first surgical procedure and acted as senior maxillofacial consultant. FA wrote the article. LM performed histological analyses and acted as a surgical pathology consultant. DR performed the second and third surgical procedures on the patient.

Competing interests None.

Patient consent Obtained.

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