A RARE CASE OF KAPOSI SARCOMA

Rebecca Billingham

Classic Kaposi sarcoma (KS) is a rare type of cancer that develops in endothelial cells in the skin. This unusual neoplastic disorder was first described by Moritz Kaposi in 1872. It is predominantly found in men between the ages of 50 and 80 who are of Mediterranean ancestry. The incidence of classic Kaposi sarcoma in men in northern Italy has been estimated at 2.5 per 100,000 (Ascoli et al, 2001).

Aetiology and presentation

The aetiology of Kaposi sarcoma depends upon the type. There are four types; classic KS, endemic/African KS, transplant or immuno-suppressed patients and acquired immunodeficiency syndrome (AIDS)-related KS. The usual presentation for all types is widespread slow-growing multiple lesions and widely dilated vascular spaces, which usually occur in the lower extremities together with lymphoplasmacellular infiltrate, extravasated erythrocytes and haemosiderin deposits (Cheung and Rockson, 2005). Generally, the lesions are painless, flat, do not itch and vary in size and colour. Over time these can progress into confluent plaque, nodules and/or tumours affecting distant skin sites (i.e. those removed from the primary tumour), lymph nodes and other organs, e.g. lungs.

Classic Kaposi sarcoma most commonly runs a relatively benign, indolent course for 10 to 15 years or more, with a slow enlargement of the original tumours and the gradual development of additional lesions. Brenner et al (2002) studied 248 patients with classic Kaposi sarcoma over 20 months and showed 39% had progression and 1.6% had died due to the disease.

Kaposi sarcoma is often concurrent with lymphoedema. This is due to impairment of lymphatic and/or venous drainage. In one small study of 13 patients (10 with AIDS-associated KS, two with classic KS and one immuno-suppressed patient) by Takizawa et al (1996), lymphoscintigraphy showed lymphatic disorder in all cases even in the absence of clinical signs of lymphoedema.

Radiotherapy seems to offer the best management of the skin lesions, but the literature details the use of chemotherapy if the Kaposi sarcoma has spread to internal organs (Fardet et al, 2006).

Case report

The patient was a 69-year-old man of Italian descent who had moved to the UK from Italy at the age of 19. He was referred to the lymphoedema clinic in April 2005, as a case of atypical lymphoedema of the left arm following a deep vein thrombosis (DVT) in the left subclavian vein and a subclavian artery occlusion. The occlusion had resolved for 12 months and there was good circulation throughout the left arm but significant venous oedema of the hand. We were asked to review the patient to see if a compression garment would help.

On initial assessment the patient gave a history of the swelling which had started throughout the left hand and forearm about two years before. At the time he believed it to be a frozen shoulder, as he had experienced this about 17 years before. He was seen at a local hospital and diagnosed as having no brachial pulse and was urgently referred for a vascular review. He was diagnosed with DVT. The swelling reduced overnight and with rest but was made worse by long periods of activity. Oedema was present throughout the fingers, hand, forearm and slightly in the upper arm. Numbness was noted throughout the fingers of both hands with occasional shooting pains and pins and needles. The skin was intact, the tissues were soft and non-pitting and a positive Stemmer's sign was noted. Measurements showed the left arm to be 32ml larger than the right. A treatment plan of skin care, exercise, strong compression hosiery and simple lymphatic drainage was prescribed with a review after two months.

At the review in August 2005, the patient said that the compression hosiery had not been comfortable and had increased the pins and needles and had therefore been removed. There had been little change and a different garment was prescribed and an appointment made for three months later.

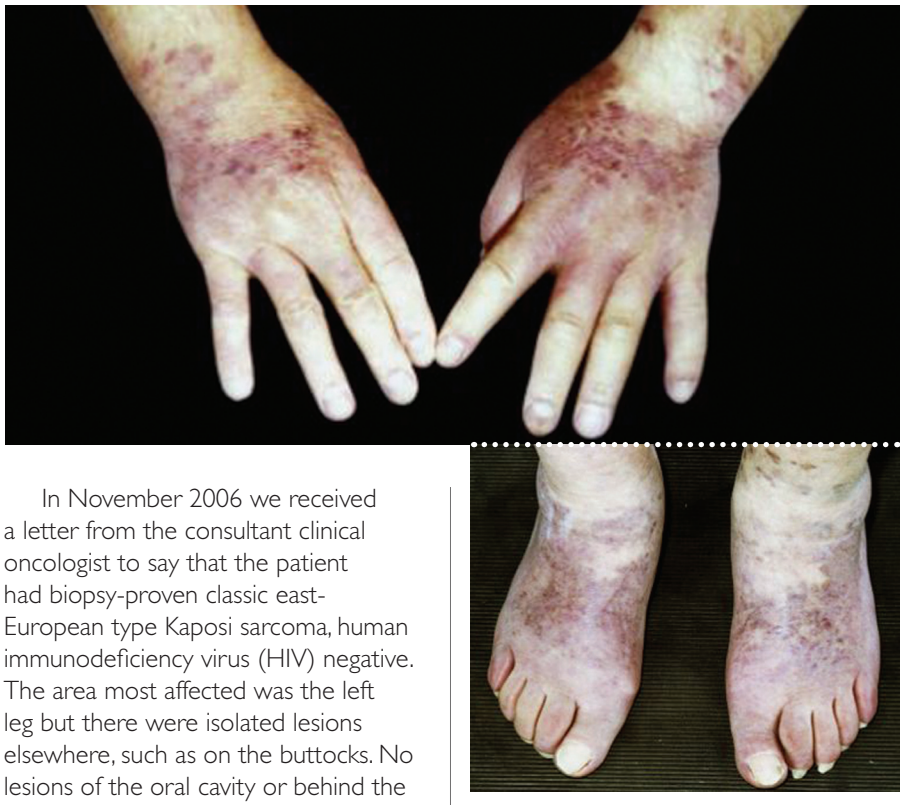
In November 2005 there was a slight reduction in the limb volume of the left arm. The shape was good, the skin was intact and in good condition and it was agreed that he would continue with skin care, exercise and compression hosiery. A small mole on the lateral aspect of the forearm just above the wrist had appeared during the previous two months. The patient was advised to seek his GP's opinion.

Rebecca Billingham is Chair of the British Lymphology Society (BLS), and Macmillan Lymphoedema Nurse Specialist at University Hospital of North Staffordshire

Three months later, in March 2006, the swelling had extended from the hand onto the wrist. The tissues had become more thickened but remained non-pitting and the limb volume had been maintained. We agreed to implement a one-week course of manual lymphatic drainage (MLD) in the hope that this would reduce the limb volume and, in turn, improve his functional use. This was to take place in May.

The patient completed a one-week course of MLD in May 2006 and the limb volume reduced, improving his functional use. On examination there was a lump to the inner aspect of the elbow which was soft and mobile in origin, and there was a query on what was causing this. After discussion with the patient and his partner and upon further examination, reddish-brown patches were found to be present on both the legs, buttocks, sacral area and very mildly to both hands. These patches were flat and did not itch, but he did complain of aching from his legs and his left hand which may have been due to his previous thrombosis on the left subclavian vein. It was agreed that he should be referred to dermatology and also back to the vascular clinic to see if there was any improvement in vascular function with lymphoedema treatment.

The patient was seen again in August 2006 when the limb volume had maintained, the tissues were thickened and non-pitting, the skin was intact and the shape was good. He had a forthcoming appointment with the vascular surgeon on the 7th September 2006. He described a swelling to the left lower leg which was thickened and pitting throughout, the toes were dusky and discoloured and warm to touch, but the skin was intact and the shape was good. The swelling had started about 6–8 weeks previously and the cause was unknown. The vascular surgeon was made aware that this required further investigation. The patient had also been seen by the dermatologist regarding the brown patches and had undergone biopsy of both areas and been photographed (*Figures 1* and *2*) and was due to be seen in three weeks time.



Figures 1 and 2. Patient's hands and feet when initially seen by the dermatologist.

In November 2006 we received a letter from the consultant clinical oncologist to say that the patient had biopsy-proven classic east-European type Kaposi sarcoma, human immunodeficiency virus (HIV) negative. The area most affected was the left leg but there were isolated lesions elsewhere, such as on the buttocks. No lesions of the oral cavity or behind the ears were present, but he was found to have Kaposi sarcoma on his penis, predominantly on the left side. Given the extensive nature of the cutaneous disease, blood tests and a computed tomography (CT) scan were undertaken. No palpable nodes or abdominal masses were found to be present. The patient was informed that the disease was not a life-threatening condition, but could cause significant morbidity. Radiotherapy was planned.

He was seen again in the lymphoedema clinic on 15th December 2006. The swelling had been maintained, he was undergoing radiotherapy to the left medial thigh and had developed swelling throughout the left leg, especially below the knee to the foot and ankle. A light compression stocking was fitted which was to be removed at night and he was instructed to commence moisturising daily once radiotherapy had finished and to take gentle exercise. Kinesio tape was employed for the hand swelling.

In February 2007 the swelling started to become more troublesome at night and there had been no noticeable benefit from the tape so this was discontinued. The left leg had improved with strong compression hosiery, the tissues had softened above the knee and the skin was very dry. The importance of skin care was reiterated. The patient complained of some pain on walking any distance, possibly due to a suspected long-standing clot in his left leg, but otherwise he had good movement and function.

The November 2006 CT scan had been clear and the cutaneous lesions continued to be treated with radiotherapy. The worse discomfort was in the left leg and the large nodules on the medial aspect were to be treated initially. Later, it was agreed to treat some individual lesions in the left posterior thigh. It was hoped that at the end of this treatment it would be possible to assess the progress and that other sites would then be treated. His lower legs, particularly the feet and ankles, had fairly confluent disease.

During February and March 2007 further radiotherapy was given to the legs to soften the lesions. The patient began to feel much better and it was

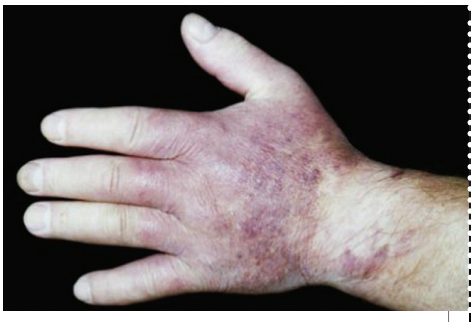


Figure 3. Close-up of the patient's hand showing the exacerbation of lymphoedema in February 2007.

planned to gradually treat the lesions on the lower left leg. During treatment it became apparent that other spots were appearing on the left arm and hand (*Figure 3*). These sites would also require radiotherapy.

In April 2007 treatment on his leg was quite successful and the swelling markedly reduced (*Figures 4* and *5*). The lesions were less visible and treatment was started to the left arm, extending from the elbow to the dorsum of the hand.

In May 2007 further radiotherapy was planned to the inner side of his hand and wrist which were getting worse. The dorsum of the hand also needed to be treated to prevent the swelling from increasing.

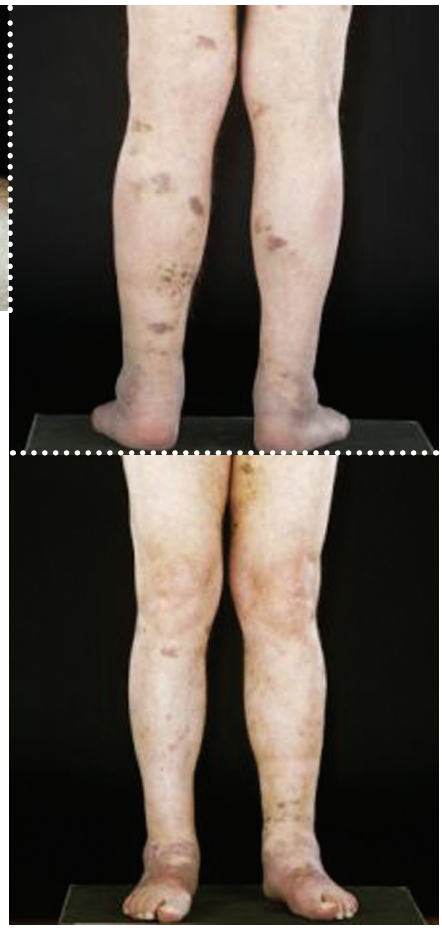
In June 2007, following radiotherapy to the dorsum of the hand, the patient stated that he felt very well, the staining had most definitely been reduced and the radiotherapy was generally considered to have been successful. The patient is aware that he may need to undergo further radiotherapy in the future when necessary.

Discussion

During his radiotherapy the patient was able to continue his lymphoedema management and the lymphoedema team and oncologists worked closely together so that it was possible to keep swelling to a minimum.

Conclusion

Classic Kaposi sarcoma is a rare and slow-growing disease which does not usually cause any problems, apart from the appearance of lesions. In the early stages it does not usually require treatment but, with time, radiotherapy may be indicated. Due to the strong association with lymphoedema, as illustrated by this case, it is important that all limbs are examined upon assessment regardless of the area denoted by the initial referral. If these seemingly innocent patches are found they should be fully investigated. The disease can progress with the number and size of the lesions increasing to cover more surface area of the skin. Lymphoedema can arise, if it has not already done so, and can become more complex to manage. There is no definitive evidence to suggest that the disease might spread to the internal organs if it is not treated, but it seems likely that this may be the case. If the patient is well and the lesions are extensive, radiotherapy is the treatment of choice. JL



Figures 4 and 5. Anterior and posterior views of both legs in April 2007 after several sessions of radiotherapy. Lymphoedema is present in both legs.

Key points

- Classic Kaposi sarcoma (KS) is a rare type of cancer that develops in endothelial cells in the skin
- The aetiology of KS depends upon the type. There are four types; classic KS, endemic/African KS, transplant or immuno-suppressed patients and acquired immunodeficiency syndrome (AIDS)-related KS.
- Radiotherapy seems to offer the best management of the skin lesions, but the literature details the use of chemotherapy if the KS has spread to internal organs (Fardet et al, 2006).
- Due to the strong association with lymphoedema, as illustrated by this case, it is important that all limbs are examined upon assessment, regardless of the area denoted by the initial referral.

References

Ascoli V, Belli S, Benedetti M, Trinca S, Ricci P, Comba P (2001) High incidence of classic KS in Mantua, Po valley, Northern Italy (1989–1998). *Br J Cancer* **85(3)**: 379–82

Brenner B, Weissmann-Brenner A, Rakowsky E, et al (2002) Classical Kaposi sarcoma: prognostic factor analysis of 248 patients. *Cancer* **95(9)**: 1982–7

Cheung L, Rockson SG (2005) The lymphatic biology of Kaposi's sarcoma. *Lymphat Res Biol* **3(1)**: 25–35

Fardet L, Stoebner P, Bachelez H, et al (2006) Treatment with taxanes of refractory or life-threatening Kaposi sarcoma not associated with human immunodeficiency virus infection. *Cancer* **106(8)**: 1785–9

Takizawa C, Cestari S, Petri V, Nishio C, Nishio P (1996) *Lymphatic study of lower limbs in epidemic, classic and iatrogenic Kaposi sarcoma.* Proceedings of the 11th International Conference on AIDS. July 7–12: 168