Kaposi's sarcoma in a renal allograft recipient with cytomegalovirus infection

A case report

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Summary

There are increasing reports of Kaposi's sarcoma arising in immunosuppressed patients, including renal allograft recipients. Furthermore, evidence is accumulating that cytomegalovirus infection may be an aetiological factor in Kaposi's sarcoma. We report an additional case in a renal allograft recipient treated with corticosteroids, azathioprine and niridazole, who also had active cytomegalovirus infection.

In 1872 Moritz Kaposi described the entity now bearing his name under the title 'Idiopathic multiple pigmented sarcomata of the skin'.

Kaposi's sarcoma was originally regarded as a disease of the skin with a predilection for the lower limbs. Skin lesions are characterized by the appearance of multiple small reddish-blue macules which tend to coalesce and develop into plaques or nodules which may ulcerate. Ulcerated nodules may mimic infected granulomas which fail to heal. Some nodules may regress while others develop. Similar lesions may occur in the mucous membranes. Kaposi's sarcoma is now regarded as a multicentric malignant haemangiosarcoma which can affect the skin, extracutaneous tissues and viscera, but as yet consensus has not been reached on its cellular origin. Occasionally the disease may present in a visceral form with no initial involvement of the skin. With lesions limited to the skin the patient may live for 10-25 years, but with visceral involvement life expectancy may be only 1-2 years. The association of Kaposi's sarcoma with other primary malignant lesions and with immunosuppression has been described. Evidence is accumulating that cytomegalovirus may act as an aetiological factor in Kaposi's sarcoma. At least 20 cases of Kaposi's sarcoma in renal transplant recipients have been reported. We wish to report another case.

Case report

The patient, a 29-year-old White man, presented in 1969 with asymptomatic proteinuria. Renal biopsy showed focal and segmental sclerosis. Antihtypertensive treatment was commenced in 1974. He was admitted to hospital in November 1976 with renal failure, and renal dialysis was started. In September 1977 a cadaver renal transplant was performed; the kidney functioned well until February 1978, when the patient developed progressive signs of rejection despite immunosuppressive treatment with steroids, azathioprine and at times niridazole. He developed persistent thrombocytopenia and gastro-intestinal bleeding occurred. On 25 April the transplanted kidney was removed. It was found to be swollen and...
tense and showed signs of cellular rejection on histological examination. Immunosuppressive therapy was withdrawn, apart from low-dose maintenance corticosteroids. The patient subsequently developed progressive jaundice and diffuse pulmonary infiltration with low-grade fever, while the thrombocytopenia persisted. Massive gastro-intestinal haemorrhage responded dramatically to vasopressin infusion, but he died of respiratory failure on 8 May despite intensive supportive haemodialysis and ventilation.

At autopsy the skin was deeply jaundiced and there were numerous petechiae, ecchymoses and a few subcutaneous nodules on the lower limbs and abdomen. There was herpes-like ulceration of the lips and mouth, and a hard submucosal nodule of tumour tissue 1 cm in diameter was found in the lower trachea. Widespread nodular masses were present throughout both lungs. The right lung had a mass of 650 g and the left lung 930 g. The mass of the heart was 310 g and there was moderate left ventricular hypertrophy. There were numerous raised nodules in the stomach, the largest 3.5 cm in diameter, each with central umbication. These nodules extended throughout the small intestine but were most numerous in the proximal portion, gradually diminishing in number distally until only an occasional lesion could be seen in the large intestine. The liver (2210 g) was dark green in colour. Numerous reddish-brown specks, later shown to be tumour, could be seen in the regions of the portal tracts. The original kidneys were contracted and granular. The mass of the right kidney was 45 g and that of the left kidney 55 g. At the site of the transplanted kidney there was a large organizing haematoma.

On histological examination the features of Kaposi's sarcoma were seen in the tumour masses removed from the gastro-intestinal tract, liver, lungs, trachea and mediastinal and paratracheal lymph nodes (Fig. 1). The liver showed numerous bile lakes, bile in the canaliculi and acute-on-chronic cholangitis, with tumour tissue infiltrating the portal tracts. There were three parathyroid glands, which were enlarged and hyperplastic. The lungs showed cytomegalovirus infection (Fig. 2) and metastatic calcifications were noted in the alveolar membranes. Cytomegalovirus was cultured from the kidney removed on 25 April, from a throat swab taken on 8 May and from the lung, liver and lymph node specimens removed at autopsy on 19 May. No cytomegalovirus was cultured from spleen and brain specimens or from blood taken post mortem. Herpesvirus hominis had previously been isolated from a throat swab taken on 3 February 1978.

**Discussion**

The patient's gastro-intestinal bleeding, for which blood transfusion was required, could be adequately explained by the ulceration of tumour nodules. Kaposi's sarcoma may present with gastro-intestinal symptoms, and when these are found in renal allograft patients this diagnosis should be considered because patients may respond favourably to withdrawal of immunosuppressive therapy. An additional predisposing factor in our patient was the persistent thrombocytopenia which is one of the effects of cytomegalovirus infection.

The patient died of Kaposi's sarcoma and a concomitant cytomegalovirus infection. Heavy immunosuppression could not control rejection of his renal allograft. This report adds to the growing number of cases of Kaposi's sarcoma seen in association with other primary tumours, or with immunosuppression either for renal transplants or for other indications. Cytomegalovirus infection is also a complication of immunosuppression, especially in association with renal transplantation; like reactivated H. hominis infection, which also occurs with immunosuppression, the association may be fortuitous. However, there is increasing suspicion that cytomegalovirus may be one of the aetiological factors in the development of Kaposi's sarcoma, to which it may be related in a similar way to Epstein-Barr virus and Burkitt's lymphoma.

**REFERENCES**