Case report

Kaposi's sarcoma of the bowel – presenting as apparent ulcerative colitis

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SUMMARY Persistent diarrhoea with mucus production developed in a 37 year homosexual man, and an initial diagnosis of ulcerative colitis was made after barium enema examination and rectal biopsy. The patient later developed cutaneous lesions which proved to be Kaposi's sarcoma, and the bowel lesion was also subsequently shown to be Kaposi's sarcoma. This tumour occurred as a manifestation of the acquired immune deficiency syndrome (AIDS). The patient was treated with alpha interferon, with partial regression of the skin lesions, but progression of the bowel tumour. Because of severe bowel symptoms, including episodes of subacute intestinal obstruction, the localised bowel disease was treated with radiotherapy. In view of the increasing incidence of AIDS, a diagnosis of Kaposi's sarcoma must be considered in homosexual men presenting with persistent diarrhoea, for which no infectious cause can be demonstrated.

Case history

The patient, a 37 year homosexual hairdresser, was well until July 1982, when he developed diarrhoea, with frequent loose stool and a mucus rectal discharge. He was examined at a clinic for sexually transmitted diseases, and a rectal gonococcal infection was found; however, his symptoms persisted after the gonorrhoea had been eradicated. Over the following three months he passed up to 12 loose stools a day, with mucus, but no blood or pus. Repeated stool culture and microscopy were negative, but he was treated empirically with metronidazole for presumed *Entamoeba histolytica* infection. There was no clinical response, and the patient was referred to a consultant gastroenterologist for investigation. In November 1982 a barium enema was performed, which showed mucosal ulceration and oedema in the rectum and distal sigmoid colon, compatible with ulcerative colitis. Sigmoidoscopy revealed an ulcerated

mucosa, and histology of a superficial rectal biopsy specimen showed inflammatory changes suggestive of ulcerative colitis. The patient was treated with salazopyrin and hydrocortisone enemas.

The symptoms improved slightly on this treatment, with a reduction in bowel movements to 6/day. In March, 1983, however, the patient noticed purple, raised and non-tender skin lesions on the dorsum of both feet. These were not brought to medical attention. In May, 1983, the skin lesions had spread, with multiple purple plaques affecting the legs, buttocks, trunk and arms. At this time, the patient developed frank bleeding with the passage of stool, and bowel movements increased in frequency to eight per day. In June, his legs began to swell, and he was referred to a consultant dermatologist. A biopsy of a skin lesion was performed, on which a diagnosis of Kaposi's sarcoma was made; he was referred to the Praed St Clinic at St Mary's Hospital.

The patient had no previous medical history, apart from multiple episodes of sexually transmitted diseases. In 1981 he had visited San Francisco, where he had numerous anonymous sexual partners, and also acquired a tattoo.

On admission, the patient was found to have

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disseminated Kaposi's sarcoma, with multiple, discrete, purple indurated lesions all over the body, including the buccal mucosa (Figs. 1–3). The lesions on the feet closely resembled those lesions seen in 'classical' Kaposi's sarcoma, as seen in elderly men. There was generalised lymphadenopathy, and lymph node biopsy revealed Kaposi's sarcoma. Additional mucosal lesions were seen in the trachea at fibre-optic bronchoscopy. Sigmoidoscopy was performed, which revealed purple friable rectal mucosa, extending from 5 cm to above 25 cm. There was considerable oedema of the rectum, which was almost rigid. A deep rectal biopsy showed Kaposi's sarcoma infiltrating the bowel wall (Figs. 4, 5). A barium enema was performed, which showed an increased presacral space at 3 cm, with narrowing and shortening of the rectum and sigmoid with fine irregularity of the mucosal surface, consistent with fine ulceration (Fig. 6). There was a separate lesion in the caecum which appeared nodular, and narrowed the caecum circumferentially. The remainder of the colon was normal. The appearances in the rectosigmoid were compatible with inflammatory bowel disease, but the caecal lesions were atypical. A computed tomography scan of the abdomen showed para-aortic lymphadenopathy, marked thickening of the sigmoid and rectum and a reduced rectal lumen (Fig. 7). Laboratory investigations revealed a total lymphopenia, ($1.1 \cdot 10^9/l$) with T helper cell depletion, ($0.37 \cdot 10^9/l$) decreased monocyte phagocytosis (81%) and anergy to three recall antigens (candida, streptokinase/strepto-

Fig. 2 Kaposi's sarcoma – close-up of nodular lesion.

dornase and PPD 1/1000). These findings were compatible with a diagnosis of AIDS.

Treatment

The patient's bowel symptoms and lymphoedema were disabling, and therefore it was decided to treat the Kaposi's sarcoma with recombinant human alpha interferon (Roche). He received 36 mega-units daily intra-muscularly for three months, planning to reduce to a maintenance schedule of three times/week for nine months, depending on response. This therapy was well tolerated apart from

Fig. 1 Kaposi's sarcoma on the foot.

Fig. 3 *Kaposi's sarcoma lesions on the hard palate.*

Fig. 4 *Photomicrograph of the rectal biopsy showing destruction of the muscularis mucosae and replacement of the submucosa by Kaposi's sarcoma. (H & E, original magnification $\times 50$).*

Fig. 5 Photomicrograph of Kaposi's sarcoma involving the large bowel; slit like spaces lined by plump pleomorphic spindle cells are present. (H & E, original magnification $\times 500$).

early fevers (which resolved within one week), and persistent anorexia and malaise.

The skin lesions showed a substantial regression, with loss of induration, leaving residual skin staining; during three months of therapy no new skin lesions developed. The lymphadenopathy and splenomegaly regressed clinically. The Kaposi's sarcoma lesions in the rectum showed progression, however, with extensive narrowing of the rectal lumen by tumour. By this stage, passage of stool was extremely difficult, and associated with severe colicky pains. It was decided to start local radiotherapy to the pelvis to alleviate these distressing symptoms. Initial response to this therapy was satisfactory, with improved bowel habit. The reduction of the interferon to maintenance regime resulted in relapse of the skin lesions, the original lesions becoming indurated again, and many new lesions appearing. The induction regime was therefore reinstituted. Despite

this treatment the patient sustained large bowel perforation and died 20 months after the onset of symptoms.

Discussion

In 1981, several cases of Kaposi's sarcoma in homosexual men with unexplained immunodeficiency were reported from New York and San Francisco in the USA.¹⁻³ It is now clear that these cases represented the beginnings of a new well-defined syndrome, the acquired immune deficiency syndrome (AIDS), and by January 1984, over 3200 cases of AIDS had been reported to the Centres for Disease Control, Atlanta.⁴ Of these, 858 cases have presented with Kaposi's sarcoma alone; the overall mortality of this group is 23%,⁴ reflecting their apparently less severe immunodeficiency in comparison with patients with opportunist infection. Kaposi's sarcoma is seen predominantly in the

Fig. 6 Double contrast barium enema, showing ulceration of the rectum and sigmoid colon.

homosexual males with AIDS, and is uncommon in patients from the other risk groups (intravenous drug addicts, haemophiliacs, Haitians, etc).⁵

The course of AIDS Kaposi's sarcoma has recently been extensively reviewed.^{6,7} The frequency of bowel involvement by Kaposi's sarcoma

in AIDS is extremely high, (over 50% of cases) and bowel involvement has been reported to occur before the development of skin lesions.⁶ The bowel lesions are generally asymptomatic and at endoscopy are seen as nodules on the bowel mucosa.⁸⁻¹⁰ At necropsy, extensive nodular disease has been reported, as well as some cases of more diffuse involvement.^{9,10} Diffuse symptomatic bowel involvement resembling ulcerative colitis, however, has not previously been reported. The initial diagnosis of ulcerative colitis in this case was supported by the radiological findings, and by the superficial rectal biopsy; the histological changes characteristic of Kaposi's sarcoma are most readily seen deep to the submucosa (see Fig. 5). This case is also unusual in that symptomatic Kaposi's sarcoma occurred five months before the development of skin lesions.

Infectious disease of the bowel is a common cause of proctitis and diarrhoea in homosexual men.^{11,12} Diarrhoea is frequently caused by sexually acquired parasites, especially *Entamoeba histolytica* and *Giardia lamblia*; bacterial infections, notably *Salmonella*, *Shigella* and *Campylobacter*, may also be sexually transmitted by anal intercourse, or by oro-anal contact. Proctitis, with a purulent rectal discharge and rectal ulceration, may be caused by *Neisseria gonorrhoeae*, *Chlamydia trachomatis* and *Herpes simplex*. In general, homosexual men presenting with bowel disturbances should have a stool examination and culture for pathogens, and a rectal swab for direct microscopy by Gram stain and culture for the gonococcus.

Kaposi's sarcoma must now be considered as an important differential diagnosis in homosexual men

Fig. 7 Abdominal CT scan, showing thickening of the rectum and sigmoid colon, and a greatly reduced rectal lumen.

with prolonged persistent diarrhoea, in whom no infectious agent is identified. The frequent involvement of the oral cavity and buccal mucosa with Kaposi's sarcoma (Fig. 3), in the presence of gastro intestinal lesions, provides a useful clinical clue, and these lesions should always be sought.⁹

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