TUMOURS

Patients with the acquired immune deficiency syndrome (AIDS) are at increased risk of developing certain malignant tumours, in addition to their risk of opportunistic infections. Kaposi’s sarcoma and high grade malignant lymphoma occur in patients with AIDS, and the presence of either tumour in an individual who is seropositive for the human immunodeficiency virus (HIV) is now a criterion for the diagnosis of AIDS. An apparently increased incidence of oral squamous carcinoma and cloacogenic carcinoma of the rectum has also recently been seen in homosexual men, some of whom are HIV antibody positive. The relation of these squamous carcinomas to the AIDS syndrome, however, remains uncertain.

Before the recognition of AIDS early in the 1980s Kaposi’s sarcoma was seen in one of several clinical settings. The most common presentation (sometimes called the “classical” form of the disease) was as a rare multicentric vascular tumour commonly presenting on the legs of elderly patients. The sex incidence was approximately 10:1 men:women, and affected individuals were usually Jewish or of east European extraction. The course of the disease was slow and chronic, patients often dying in old age of some unrelated condition.

In Africa, particularly parts of sub-Saharan Africa, Kaposi’s sarcoma has been endemic for many years. In addition to a form of disease resembling that seen in elderly Jewish men and a form predominantly affecting the lymph nodes in infants and young children, there is a variety which affects young adults, has an aggressive course, and has many features in common with AIDS related Kaposi’s sarcoma. AIDS related Kaposi’s sarcoma itself is now not uncommon in many parts of Africa.

A third variety of non-AIDS Kaposi’s sarcoma occurs in patients receiving immunosuppressive therapy, notably renal transplant patients. In this group men and women are equally affected and the tumours may regress after the cessation of immunosuppressive therapy.

The commonest form of Kaposi’s sarcoma now occurring in Britain is that associated with AIDS, which affects particularly young male homosexuals (between a quarter and one third of all homosexual patients with AIDS). The tumour affects other groups at risk of AIDS, such as intravenous drug abusers, much less commonly, and the lesions commonly precede the development of opportunistic infections.

Clinical features

In patients with AIDS lesions of Kaposi’s sarcoma are usually multiple, often progress rapidly, and may affect practically any area of the skin as well as internal organs. The tumours often begin as small flat dusky red or violet areas of skin discoloration, progressing in weeks or months to raised painless firm nodules and plaques. Although the tumour may affect the legs, similar to the pattern of disease seen in the classical form of Kaposi’s sarcoma, lesions on the trunk, arms, and face are also very common. On the trunk tumour lesions are often elongated, following the skin cleavage lines, and lesions on the palate are often an early manifestation of the disease.
Classical nodular Lymphangioma

Kaposi's sarcoma.

Pathology

Kaposi's sarcoma is generally agreed to be a malignant tumour which arises from, or differentiates towards, vascular endothelium. It is unusual in that it appears to develop in a multifocal way, new lesions occurring at various cutaneous and internal sites which cannot be explained by lymphatic or blood spread, common in other forms of cancer.

Although Kaposi's sarcoma is a vascular tumour, the cell of origin is controversial. It was originally considered to be a tumour of blood vessels, but modern studies with monoclonal antibodies and other cell markers suggest that at least some tumours may be derived from lymphatic endothelium.

The histopathological appearances of early cutaneous lesions are subtle and may easily be missed. Elongated and irregular spaces appear in the dermis, sometimes associated with slight increase in chronic inflammatory cells (particularly plasma cells) and some haemosiderin deposition. The patch stage (flat) lesions of the Kaposi's sarcoma associated with AIDS in particular may be composed of areas of these anastomosing spaces with a jagged outline, there being very little evidence of tumour cells present (the so-called lymphangioma like type of Kaposi's sarcoma). More advanced plaque and tumour stage lesions of all types of Kaposi's sarcoma show a more diagnostic histopathological picture. In the dermis aggregates of spindle cells are seen trapping red cells between them and associated with tissue deposition of blood pigment. The degree of associated chronic inflammatory cell host response is variable, as is the degree of atypia of individual tumour cells. Pronounced anaplasia of tumour cells, with numerous and abnormal mitotic figures, is the exception rather than the rule.

The precise cause of Kaposi's sarcoma is still unknown and controversial. Although virus particles (especially cytomegalovirus) have been found in Kaposi's sarcoma tissue, the evidence for a pathogenetic role for such viruses is at best circumstantial.

Treatment

In most patients Kaposi's sarcoma never becomes life threatening, and the prognosis is dictated by the underlying immune deficiency, to which the patients generally succumb. Exceptionally, however, patients may develop massive intra-abdominal and pulmonary disease, when haemorrhage from Kaposi's sarcoma may be life threatening. The treatment should be tailored to the form of the disease. Localised nodules of Kaposi's sarcoma increasing in size and becoming embarrassing and cosmetically distressing may be
Malignant lymphomas

Shortly after the acquired immune deficiency syndrome was described an increased incidence of B cell lymphoma, usually of high grade malignancy, began to be recognised in male homosexuals. Many of these patients showed serological evidence of HIV infection, and, after Kaposi's sarcoma, malignant lymphoma is now the commonest malignant tumour affecting patients with AIDS. Like Kaposi's sarcoma, this tumour affects male homosexuals much more commonly than other risk groups, such as haemophiliacs and intravenous drug abusers.

Clinical features—Young homosexual men (mean age 37) present with lymphomatous infiltration of extranodal areas, particularly the central nervous system, bone marrow, gastrointestinal tract (including the rectum), and mucocutaneous sites. Disease confined to lymph nodes occurs but is uncommon. The lymphoma is often at an advanced stage at presentation, and “B symptoms” of fever and weight loss are frequent. About half the patients with this tumour already have a history of previous opportunistic infection. The differential diagnosis of malignant lymphoma in these patients depends largely on the site affected. Lymph node disease must be distinguished from the lymphadenopathy of the persistent generalised lymphadenopathy syndrome, and lymphoma of the central nervous system must be carefully separated from the many infections, including AIDS related dementia itself, which may present with neurological signs. The prognosis of AIDS related malignant lymphoma is poor, relapse rates following chemotherapy are high, and mean survival is less than a year.

Pathology.—Although various types of malignant lymphoma (including Hodgkin's disease) have been described in patients positive for HIV, most AIDS related lymphomas are extranodal high-grade B cell lymphomas. The exact histological subtype may be difficult to classify but is usually reported as immunoblastic lymphoma, centroblastic or centrocytic/centroblastic lymphoma, lymphoblastic lymphoma, or Burkitt like lymphoma. Some of the Burkitt like lymphomas have shown chromosome translocations on cytogenetic studies. The relation of the lymphadenopathy of persistent generalised lymphadenopathy to AIDS related lymphoma is unclear, but only a small proportion of patients with generalised lymphadenopathy progress to lymphoma, which, as already mentioned, chiefly affects extranodal sites.

Treatment

Because of its association with immune deficiency and the frequent occurrence of opportunistic infections, treatment of AIDS related
Epidemiology

Rubella susceptibility and the continuing risk of infection in pregnancy

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In November 1983 the Department of Health and Social Security launched a campaign to increase, over three years, the uptake of rubella vaccination in schoolgirls and women. Part of this initiative the Public Health Laboratory Service, with funding from the Office of the Chief Scientist, set up a study to monitor rubella susceptibility and infection in pregnancy and to determine vaccine uptake in non-immune women. The study began in 1984 in eight public health laboratories (Ashford, Bristol, Gloucester, Hereford, Leeds, Luton, Manchester, and Reading) which together screen over 100000 pregnant women each year, about one sixth of the annual antenatal population of England and Wales.

Methods

In the eight laboratories the request form for antenatal rubella screening was changed to include a question on parity, a microcomputer was installed for entry of all rubella data, and a nurse was appointed to inquire into the vaccination of non-immune women. Data, including the results of diagnostic screening, were analysed at the Communicable Disease Surveillance Centre of the Public Health Laboratory Service from discs sent from each laboratory. Each case of rubella infection in pregnancy reported to the Communicable Disease Surveillance Centre from laboratories in England and Wales was followed up through the microbiologist, obstetrician, and general practitioner.

Results

ANTENATAL SUSCEPTIBILITY

The results for Manchester showed that in the annual antenatal population of around 40000 the proportion susceptible to rubella fell from 6.7% in 1979 to 2.7% in 1984; susceptibility was lower in parous than in nulliparous women. Continuing surveillance of this antenatal population has shown no further decline in susceptibility over the two and a half years since April 1984 (fig 1).

Results from five of the other seven study laboratories for which data were available from January 1985 showed an overall decline in susceptibility to rubella in both nulliparous and parous women over the period (table). The downward trend was significant for the five laboratories combined (p<0.05) but was not observed in two individual laboratories (Ashford and Reading).

VACCINATION OF NON-IMMUNE WOMEN

Initially the study nurses found that local postpartum vaccination policies and their implementation were variable. For instance, in the population served by one laboratory only four out of 81 (5%) non-immune women who delivered between January 1984 and May 1985 had been vaccinated. After action by the nurse, in the first six months of 1986 41 out of 49 (84%) non-

![Graph showing susceptibility to rubella among women tested antenatally in Manchester April 1984-December 1986.](http://www.bmj.com)

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