The exact nature of the spread of this carcinoma is difficult to determine and it thus represents an unusual case of distant colonic carcinoma metastasis.

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Typhlitis complicating acute leukaemia in an adult

Sir,

Typhlitis is a necrotizing inflammation of the caecum recognized as a complication of chemotherapy with leukaemia in children¹ and adults.² We report a case of typhlitis in an adult with leukaemia following the neutropenic phase of chemotherapy.

A 72 year old female was diagnosed as having myelomonocytic chronic leukaemia. She underwent ambulatory therapy with hydroxyurea and etoposide when there was transformation into acute non-lymphoblastic leukaemia. She was admitted for induction therapy with cytarabine, daunorubicin and prednisone. On the seventh day after chemotherapy, she developed a fever of 38°C and ceftriaxone and amikacin was started. On the 14th day she had abdominal pain with marked tenderness in the right inferior quadrant with involuntary guarding and rebound tenderness. X-ray showed dilated small-bowel loops and air-fluid levels in the right lower quadrant. Her white cell count was $0.7 \times 10^9/l$. At laparotomy the caecum, proximal ascending colon, terminal ileum and appendix were resected, and an ileocolic anastomosis was made. Pathological examination showed an oedematous and ulcerated mucosa of the caecum. Microscopically the ileocaecal valve and the caecum had an ulcerated mucosa, marked oedema of the submucosa and multiple and small foci of ischaemic necrosis of the mucosa, some of them colonized with Gram-negative rods. Neither leukaemic infiltration nor intramural haemorrhage was identified. After laparotomy, the patient recovered well and the fever disappeared. As soon as the bone marrow recovered from

aplasia, there was evidence of relapse with 86% blasts in the peripheral blood and she subsequently died.

The pathogenesis of typhlitis remains unclear.³ A combination of factors including chemotherapy,⁴ immunosuppression,⁴ neutropenia,⁵ steroid therapy and malignant infiltration⁶ have been invoked.

Typhlitis appears to be increasing in incidence,⁵ and we believe the clinician has to consider this complication, when a neutropenic patient develops fever and abdominal pain. Early recognition and surgical intervention is crucial for the survival of the patient.

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Xeroderma pigmentosum with recurrent infiltrating ductal carcinoma of breast

Sir,

The association of malignant tumours with other diseases is always an important observation. Since xeroderma pigmentosum (XP) has already been associated with various skin and parenchymatous tumours,^{1–3} physicians should be aware of its possible connection with breast cancer as well.

A 63 year old Jewish mother of two presented in November 1992 with a one month history of two enlarging tumours: a 3×3 cm tumour of the right upper lateral breast and a 2×2 cm fungating clinical squamous cell carcinoma (SCC) over the third metacarpophalangeal region of the left hand. She was being followed by the Department of Dermatology for XP. The latter diagnosis was based on typical clinical features, histopathological data, DNA replicase deficiency in fibroblasts and familial history. In 1962 a SCC of her lower lip was excised, and in 1967 a basal cell carcinoma of her cheek was excised. In

1975 she underwent left mastectomy for T₁N₀M₀ infiltrating ductal carcinoma. In 1979 a malignant melanoma of her left upper arm was excised. She had mild bilateral deafness and a brother with XP.

Examination revealed the abovementioned tumours with right axillary lymphadenopathy and skin lesions which were typical for XP. Mammography revealed a malignant mass in the right breast, confirmed by fine needle aspiration; cytology of the left-hand tumour showed SCC. A right modified radical mastectomy was performed following frozen section confirmation of the right breast carcinoma. The SCC of the left hand was excised and the defect was reconstructed with a split-thickness skin graft. The histopathological findings showed right breast infiltrating ductal carcinoma and SCC of the left hand.

XP is a rare autosomal recessive disease, with an incidence of two per million. It is characterized by a defect in the excision repair mode of ultraviolet-damaged DNA and neoplastic changes in sunlight-exposed skin. Other cutaneous abnormalities associated with XP are abnormal reaction to sun exposure, freckling, telangiectasia, atrophy and premalignant actinic keratoses. Some patients have, in addition, ocular abnormalities and progressive neurological degeneration.¹ The presence of skin neoplasms, including SCC,^{1,2} basal cell carcinoma^{1,2} and malignant melanoma^{1,2} in patients with XP is well established in the literature. Skin tumours less frequently reported include malignant schwannoma and angiosarcoma.

Kraemer *et al.*,¹ in a survey of the medical literature (297 articles) from 1871 to 1982, abstracted descriptions of 830 affected patients. They found reports of 12 patients with primary internal neoplasms, including four with brain tumours, two with leukaemia and two with lung tumours. The same authors had previously noted an approximate 10–20-fold increase in the frequency of internal neoplasms among these case reports.³ Carcinoma of the thyroid and renal leiomyosarcoma are internal neoplasms that were also found to be associated with XP.

An impairment of cell-mediated immunity has been proposed as a cofactor in the cancer proneness of XP patients^{4,5} and impairment of the humoral immunity has been described elsewhere.¹

This case provides further evidence that patients with XP may show an increase in other organ malignancies in addition to the skin malignancies which have been well described and this should be taken into consideration during their examination. It is likely that there is an immunological defect in these patients.

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