Infections are a major cause of morbidity and mortality in patients with chronic lymphocytic leukaemia (CLL). Predisposition to infection in CLL is mediated through various abnormalities including both the impairment of humoral and cellular immunity and further immunosuppression related to the therapy of CLL. Among the infections in CLL patients, pulmonary manifestations are very common and are often difficult to distinguish from other pulmonary disorders on clinical grounds. Opportunistic infections such as Pneumocystis carinii, fungi, viruses, and mycobacteria are seen in patients with CLL, most commonly in those being treated with corticosteroids and/or purine analogues. A 52 year old woman is reported who was on long term treatment with chlorambucil and taking a short course of prednisone for familial CLL before she developed progressive dyspnoea, and P. carinii pneumonia was diagnosed in bronchoalveolar lavage fluid. Despite treatment with high dose co-trimoxazole the patient died.

We describe a patient with familial CLL and a very low CD4 count who developed a P. carinii pneumonia infection in the absence of fludarabine therapy.

CASE REPORT

A 52 year old woman with a 10 year history of familial CLL was treated with chlorambucil (pulsed treatment with chlorambucil at a daily dose of 10 mg for three consecutive days every four to eight weeks). This regimen led to reversion of her anaemia. One of her brothers also had CLL as had her sister. Treatment with fluudarabine and steroids is rare, although clinically relevant CD4 T-cell depletion can occur in longstanding CLL without prior treatment with purine analogues. A 52 year old woman was reported who was on long term treatment with chlorambucil and taking a short course of prednisone for familial CLL before she developed progressive dyspnoea, and P. carinii pneumonia was diagnosed in bronchoalveolar lavage fluid. Despite treatment with high dose co-trimoxazole the patient died.

Pneumocystis carinii pneumonia in chronic lymphocytic leukaemia

S R Vavricka, J Halter, L Hechelhammer, A Himmelmann


Pneumocystis carinii pneumonia in patients with chronic lymphocytic leukaemia (CLL) who have not been treated with fludarabine are rare, although clinically relevant CD4 T-cell depletion can occur in longstanding CLL without prior treatment with purine analogues. A 52 year old woman is reported who was on long term treatment with chlorambucil and taking a short course of prednisone for familial CLL before she developed progressive dyspnoea, and P. carinii pneumonia was diagnosed in bronchoalveolar lavage fluid. Despite treatment with high dose co-trimoxazole the patient died.

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patients not treated with purine analogues P carinii pneumonia is usually associated with steroid therapy over a long period of time, often in combination with cyclophosphamide.3 5–8 In one large series the occurrence of P carinii pneumonia in four patients with CLL is described but no clinical details regarding treatment or CD4 counts are provided.3

Hypogammaglobulinaemia is probably the most important immune defect in terms of risk of severe bacterial infections in CLL patients. In patients with CLL, hypogammaglobulinaemia shows a prevalence from 10% to 100% and is correlated with the duration of the disease and with the stage of CLL. However in the patient presented here several immunoglobulin measurements were within the normal range.

A T-cell defect has also been described in CLL patients. In many patients with CLL the number of CD4 cells is actually increased but decreases as the disease progresses. More consistently, the CD4/CD8 ratio appears to decrease with advanced CLL, but such a low ratio as seen in our patient has never been reported.9 10 In all CLL patients suffering from P carinii pneumonia not treated with purine analogues, the number of CD4 cells was reported to be normal or only slightly reduced (range 300–370).9 Also the CD4/CD8 ratio was only slightly reduced. In contrast, in our patient the number of CD4 cells and the CD4/CD8 ratio were severely depressed. We could only find one other patient suffering from CLL with such low CD4 numbers; this patient developed a mycobacterial infection.11 As in our patient this patient had longstanding disease that lasted approximately 10 years and was treated with chlorambucil and intermittently with prednisone. Therefore longstanding CLL might lead to depletion of CD4 T-cells and clinicians might therefore consider a prophylactic antibiotic in these patients.

Surprisingly the necropsy showed perforation of the colon caused by a blastic, angiocentric, Epstein-Barr virus positive B-cell infiltration. Uncontrolled sepsis originating from the perforation was probably the ultimate cause of death in this patient. The course of the disease in the present patient demonstrates that clinically relevant CD4 T-cell depletion can occur in longstanding CLL in the absence of prior treatment with purine analogues.

Learning points

- Opportunistic infections such as Pneumocystis carinii, fungi, viruses, and mycobacteria can occur in patients with chronic lymphocytic leukaemia (CLL) having treatment with long term corticosteroids and/or purine analogues.
- A clinically relevant CD4 T-cell depletion and/or a hypogammaglobulinaemia can occur in longstanding CLL without prior treatment with purine analogues.
- Therefore, patients with long lasting CLL without prior purine analogue treatment might be considered for a prophylaxis similar to those patients with CLL treated with purine analogues.
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