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Enteropathy type T cell lymphoma with an unusually late relapse: a case report

Enteropathy type T cell lymphoma (ETTL) is a rare subtype of T cell lymphoma that is strongly associated with coeliac disease.¹ The overall prognosis is poor. In one study, 15 of 19 (79%) patients relapsed within 60 months after the initial diagnosis (median, 6 months), and the overall survival rate at 5 years was 24%.² We describe a case of ETTL with an unusually late relapse. The patient was a 50-year-old woman who developed an ulcerated gastric tumour in 1992, with no associated lymphadenopathy or hepatosplenomegaly. A partial gastrectomy was performed, and at the time of surgery, the tumour was found grossly infiltrating into the peritoneum, posterior rectus sheath and the liver. The pathological diagnosis was diffuse large cell lymphoma of mature T cell lineage (fig 1). On immunohistochemical analysis, the neoplastic cells were found to be CD3+, CD4–, CD5–, CD7+/– and CD8–, and positive for CD56, TIA 1 and granzyme B. She received a full course of VACOP-B (etoposide, doxorubicin, cyclophosphamide, vincristine, prednisone,

bleomycin). Shortly after the diagnosis of T cell lymphoma was made, she was diagnosed with coeliac disease. In view of this finding, the T cell lymphoma was reclassified as ETTL. She was in complete remission and her gastrointestinal symptoms were under control with a gluten-free diet. In 2003, she experienced symptoms of progressive nasal obstruction. Excisional biopsy of a left nasal mass was performed and the pathological diagnosis was diffuse large cell lymphoma of mature T cell phenotype. The immunohistochemical results from the tumour of 1992 and those of 2003 were identical. Molecular analysis of the T cell receptor γ gene also showed an identical rearrangement between these two neoplasms (fig 1), supporting the fact that they were derived from the same neoplastic clone. She was treated with eight cycles of CHOP+R (cyclophosphamide, vincristine, doxorubicin, prednisone, rituxan), followed by additional surgical excision, radiation therapy to the nasal cavity (3600 cGy over 20 fractions) and three cycles of prophylactic intrathecal chemotherapy (methotrexate and cytarabine). After a short remission, she presented with swelling of her right nipple in 2005, which was confirmed to be relapsed T cell lymphoma. She received two cycles of intensive chemotherapy and autologous stem cell transplantation. After a brief remission, she developed jaundice and thrombocytopenia. Circulating malignant cells were found in the peripheral blood (fig 1). She was placed under home palliative care. In summary, our patient represents a highly unusual case of ETTL that relapsed after a long latency time period.

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Anaplastic large cell lymphoma presenting with paraneoplastic pemphigus

Paraneoplastic pemphigus is an autoimmune acantholytic mucocutaneous disorder that rarely accompanies an overt or occult neoplasm. Typical features include painful mucosal erosive lesions and pruritic papulosquamous eruptions that often progress to blisters. The antibody is a unique immunoglobulin G that recognises epidermal proteins. We discuss the case of a 23-year-old woman who presented with anaplastic large cell lymphoma complicated by paraneoplastic pemphigus, rapidly resulting in fatal multiorgan failure.

A 23-year-old woman presented with 1 week of fever and a disseminated erythrodermic rash with desquamation of the hands and feet, and erosions in the mouth. A local doctor started treatment with ciprofloxacin and prednisolone.

A diagnosis of toxic epidermal necrolysis was made. On day 4, she required intubation, ventilation and transfer to a tertiary referral centre. A skin punch biopsy showed features of Staphylococcal toxic shock syndrome.

By day 9, multiorgan failure developed and a laparotomy was performed for acute abdominal distension. Retroperitoneal lymphadenopathy, hepatomegaly and splenomegaly were noted, and splenectomy and lymph node biopsy performed.

The histological examination of the spleen and splenic hilar lymph node showed replacement by a nodular infiltrate of small lymphoid cells with vesicular nuclei, multiple nucleoli and a moderate amount of eosinophilic

Figure 1 Left upper panel: gastric enteropathy type T cell lymphoma diagnosed in 1992, composed of a monotonous population of lymphoid cells with a medium cell size, a moderate amount of pale-staining cytoplasm and a round vesicular nucleus. Left lower panel: a circulating large lymphoma cell with a few azurophilic granules in the cytoplasm. Right panel: molecular studies showed that both the gastric and the relapsed nasal tumour had an identical T cell receptor gene arrangement, TCR γ 2, detectable using an automated gene sequencer (AB13100, Applied Biosystems, Foster City, California, USA).