Unusual association of diseases/symptoms

Primary cutaneous marginal zone B cell lymphoma in monozygotic twins

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The authors present the first reported case of primary cutaneous marginal zone B cell lymphoma (PCMZL) in monozygotic twins. The occurrence of PCMZL in monozygotic twins is likely to be due to a combination of shared genetic and environmental factors. A different treatment modality was used to treat each patient but both achieved a complete clinical response.

BACKGROUND

Primary cutaneous marginal zone B cell lymphoma (PCMZL) accounts for approximately 7% of primary cutaneous lymphomas,¹ with a disease-specific 5-year survival of approximately 100%.² Here, we present the first reported case of PCMZL in monozygotic twins.

CASE PRESENTATION

A 45-year-old man, patient 1, presented to the dermatology department with lesions on his back and left arm. These had been present for over a year. The patient had no medical history of note and he was not on medication

at the time of diagnosis. The lesions were biopsied and the patient was referred to an oncologist. When first examined in the oncology department, the skin lesions had completely resolved; there was no lymphadenopathy or organomegaly. The patient returned to the clinic for review 3 months later and there was evidence of recurrence at the site of his previous biopsies. The two lesions measured 15×10 mm and 10×10 mm (figure 1).

Figure 1 A lesion on the skin of patient 1 at recurrence.

Figure 2 A lesion on the skin of patient 2 at presentation.

Figure 3 Histology from patient 1 showing cutaneous marginal zone B cell lymphoma.

Figure 4 Histology from patient 2 showing cutaneous marginal zone B cell lymphoma.

Patient 1 stated that his identical twin brother (patient 2) had noticed similar flat erythematous lesions on his back. Patient 2 was concerned that he may have a similar malignant condition to his twin brother and sought medical attention. He was reviewed by the same clinical oncologist who elicited a 15-year history of intermittent skin lesions

affecting numerous parts of his body, predominately his back. The burden of lesions was unpredictable; sometimes there were crops of lesions and other times there were none. At presentation, there were multiple erythematous lesions on his back (figure 2), but there was no lymphadenopathy or organomegaly. A biopsy was performed. Patient

2 was asthmatic and used fluticasone and salbutamol inhalers.

INVESTIGATIONS

Biopsies of two lesions on the back of patient 1 were taken. Both lesions demonstrated PCMZL (figure 3). The dermis showed a nodular lymphoid infiltrate extending into the deep dermis. The cells were monocytoid with a rim of pale cytoplasm dissecting through collagen bundles. Immunohistochemistry showed the cells to be predominantly B cells expressing CD20, CD79a and BCL2. Immunoglobulin heavy (IgH) chain gene analysis confirmed IgH (V-J) rearrangement and a clonal population.

A biopsy of one of patient 2's skin lesions showed similar histological features and confirmed cutaneous marginal zone B cell lymphoma (figure 4).

TREATMENT

Patient 1 was initially kept under review because at presentation there were no active skin lesions. When his cutaneous lymphoma recurred, patient 1 was treated with megavoltage electrons. Each lesion was treated with a separate field and received a dose of 30 G in 15 fractions over 3 weeks using 4 MeV electrons.

Treatment options were discussed with patient 2. Due to the large area of skin affected on his back, it was felt that treatment with megavoltage electrons would be associated with significant toxicity without the guarantee of complete response. Instead, patient 2 received four cycles of systemic chemotherapy using R-CVP (rituximab 375 mg/m², cyclophosphamide 750 mg/m², vincristine 2 mg and prednisolone 40 mg/m² for 5 days).

OUTCOME AND FOLLOW-UP

Patient 1 had a complete clinical remission following radiotherapy and no new lesions have developed.

Patient 2 had a complete clinical remission following chemotherapy. He remains well and has not developed any further lesions.

DISCUSSION

The increased incidence of systemic lymphoma in monozygotic twins is well established. For example, a monozygotic twin has a 99-fold higher risk of developing lymphoma if the other twin has Hodgkin's disease.³ Examples of cutaneous lymphoma in monozygotic twins are much rarer. There are two reported cases of mycosis fungoides in monozygotic twins.^{3 4} This case report is the first to describe PCMZL in monozygotic twins. The clinical presentation of both patients was typical of PCMZL;² in both patients, the trunk was the main site of involvement, there was spontaneous appearance and resolution of skin lesions and the lesions were not ulcerated.

The occurrence of PCMZL in monozygotic twins is likely to be caused by a combination of shared genetic and environmental factors. Specific genetic abnormalities such as T(14;18)(q32;q21), involving the IgH chain locus and the MALT1 gene, have been identified in some patients with PCMZL.⁵ As the patients described here are monozygotic, both individuals will carry the same genetic predisposition to the development of cutaneous lymphoma. The twins described had a very similar childhood and will have been exposed to similar environmental agents. Infectious agents have been linked to the development of lymphoma; viruses have been linked with the development of non-Hodgkin's lymphoma, such as Epstein-Barr virus in Burkitt's lymphoma and human herpes virus 8 in primary effusion lymphoma. The spirochete *Borrelia burgdorferi* has previously been linked to PCMZL,⁶ but there was no history of this infection in the patients described here.

This case report also demonstrates how two different treatment modalities can achieve a complete clinical remission in PCMZL. The choice of treatment modality depends on the site, distribution and burden of disease. Patient preference and the toxicity associated with treatment must also be taken into consideration.

Learning points

- ▶ This is the first reported case of PCMZL in monozygotic twins.
- ▶ The development of PCMZL in monozygotic twins could help to generate new hypotheses regarding the pathogenesis of PCMZL.
- ▶ PCMZL can be successfully treated by chemotherapy or radiotherapy.

Competing interests None.

Patient consent Obtained.

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