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Editorial

Lymphomatoid papulosis: an enigmatic condition

Lymphomatoid papulosis (LyP) is rare condition characterised by recurrent papulonodular lesions which have a malignant histological appearance. However, despite the reported incidence of 10-20% turning into lymphoma, the clinical course is benign in the majority of patients. The question is, which patients will turn into lymphoma? Are there any clinical features or histological subtypes that predict the risk of malignant transformation? The other question is, were any of the cases of malignant transformation actually cases of cutaneous lymphoma?

Over the past few decades, our knowledge of lymphomatoid papulosis has expanded. There are now five histological subtypes of LyP, type A (histiocytic), type B (mycosis fungoides like) and type C (anaplastic large cell lymphoma like) and two new recently described histological variants, type D (which mimicks an aggressive epidermotropic cytotoxic lymphoma with $CD8^+$ $CD30^+$ atypical T cells infiltrating the epidermis) and type E (an angioinvasive type). Occasionally, more than one histological subtype may be present in one patient. Given this appearance, mycosis fungoides, Hodgkin disease or large-cell $CD30^+$ lymphoma, cutaneous γ/δ T-cell lymphoma and extranodal NK/T-cell lymphoma, nasal type and pagetoid reticulosis are just some of the malignant conditions that need to be considered in the histopathological differential diagnosis.

Do we have any information on predictors of lymphomatous transformation? There have been mixed reports on the role of age. Old age was reported to be a risk factor for lymphomatous transformation in LyP in one study.[1] However, age did not have an effect in a later study.[2] Recently, older age and presence of a T-cell clone in LyP lesions have been reported as risk factors for transformation to lymphomas.[3]

Histologically, it has been suggested that type C has been associated with an increased risk, while type B is protective.[1] However, other reports have proposed that the mixed histological type that is protective.[4] Given these conflicting reports, it is obvious that more research on this entity is required. This is not easy as LyP is a rare condition such that recruitment of cases for research will be a slow process. In this issue, Cheng provides much needed data on the LyP cases in Hong Kong, further adding to our knowledge of LyP in Chinese. It is only through this type of information that we can truly determine whether this condition exhibits any variation between different ethnic groups.

Given the alarming histological appearance, the only clinical feature distinguishing LyP from ALCL is the relapsing nature of the skin lesions. Yet, although reassuring to the clinician, this is understandably worrying to the patient. Faced with the prospect of no permanent cure of these lesions that come and go, (for example, in type D, patients can present with rapidly progressive ulcerative papulonodular lesions), it not surprising that patients would prefer better treatment and prognostic information on their condition. At present, treatment options include phototherapy, methotrexate, as well as isolated reports of modalities such as topical carmustine, nitrogen mustard being effective.[5]

The aetiology of LyP remains unknown, although virus particles have been reported

in LyP lesions.[6] There are differing views on whether LyP is a benign chronic disorder of T cells or a T cell cutaneous malignant condition. To answer these questions and to further develop our knowledge of this enigmatic entity, continued research and accumulation of cases is required. Only then can we provide better information on the prognosis and maybe new treatments to patients with this condition. However, due to the rarity of this entity and the long interval to lymphomatous transformation, this can be a slow process. For now, regular monitoring for transformation remains an essential part of treatment.

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