Case reports

A commentary on the following two case reports appears on pp 448–9.

Co-existing conjunctival non-Hodgkin's lymphoma and pulmonary sarcoidosis

PJ Ryan, P Stableforth, DT McLeod

Abstract

A 55 year old woman with a conjunctival non-Hodgkin's lymphoma was found to have pulmonary nodules on a thoracic computed tomographic scan which were initially thought to be lymphomatous deposits. A subsequent biopsy specimen demonstrated granulomas consistent with sarcoidosis. The relationship between sarcoidosis and malignancy, in particular lymphoma, is discussed.

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Keywords: non-Hodgkin's lymphoma, sarcoidosis, orbit.

Case report

A 55 year old woman presented with a five year history of a subconjunctival swelling at the outer canthus of her right eye. The lesion was not painful and there was no history of a change in size. There were no other complaints except for occasional exertional dyspnoea on walking up hills. On examination there was a pink fleshy mass under the lateral bulbar mucosa, the posterior edge of which was not seen (fig 1a). There were no other ocular abnormalities and the general examination was normal. Biopsy specimens of the mass demonstrated a low grade non-Hodgkin's lymphoma of the B cell type (fig 1b). Subsequent investigation for evidence of systemic dissemination demonstrated normal results for full blood count, erythrocyte sedimentation rate, direct Coomb's test, lymphocyte blood markers, liver function tests, lactate dehydrogenase, uric acid, and immunoglobulins. A computed tomographic (CT) scan of the pelvis was also normal, as were pulmonary function tests including spirometry, lung volumes, and carbon monoxide transfer factor. A chest radiograph showed some vague soft nodular shadows and a thoracic CT scan revealed bilateral upper zone nodules (fig 2a). These were initially thought to be lymphomatous deposits but histological examination of thoracoscopic lung biopsy specimens dem-
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There has been much discussion as to whether, in sarcoidosis, there is an increased risk of primary neoplasms in general and of lymphomas in particular. The association was noted by Brincker and Wilbeck who found 48 cases of cancer in 2544 Danish patients reported to have sarcoidosis. Malignant lymphomas occurred 11 times and lung cancer three times more frequently than expected. However, these findings were later challenged when, on re-examination, 14 of the cases were excluded on the grounds of inaccurate diagnosis or sarcoid-like reactions.

Sarcoidosis and lymphoma both show granulomas on histological examination and both are associated with abnormalities of the immune system. The concept of T cell dysfunction causing both sarcoidosis and reduced immunological surveillance leading to lymphoma is attractive but as yet unproven. The prolonged use of corticosteroids in patients with sarcoidosis has been suggested as a cause of increased incidence of lymphoma but is unlikely to explain the preponderance of Hodgkin's disease.

Ocular adnexal lymphomas are rare and often occur as secondaries. When they occur as primaries they tend to be confined to the orbit for a long time, as in this case. Delayed metastasis may be explained by them arising from reactive lesions before malignant change occurs, the absence of lymphatic drainage and, as has been found in other extranodal lymphomas, by their cells tending to home back to their original site if they enter the general circulation. The main therapeutic approach to ocular adnexal lymphomas is low dose radiotherapy which achieves good local control at the cost of lens opacity in approximately 10%.

Conjunctival non-Hodgkin's lymphoma and pulmonary sarcoidosis occurring in the same patient have not been previously reported. The combination presented an interesting diagnostic challenge that was resolved by assimilating all the clinical and histological information available. This case illustrated the need to investigate assumed lymphomatous deposits if possible, especially where the overall management would be radically altered. Whether the association between sarcoidosis and lymphoma is more than a coincidence remains unclear.

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