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

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

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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.

Figure 1 (a) Conjunctival mass at presentation. (b) Haematoxylin and eosin stain demonstrating non-Hodgkin’s lymphoma (magnification × 400).

Figure 2 (a) Computed tomographic scan of thorax demonstrating nodular lesions. (b) Haematoxylin and eosin stain of thoracoscopic lung biopsy specimen showing granuloma (magnification × 160).
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Discussion

Eye lesions and pulmonary lesions are seen in patients with sarcoidosis and also in patients with lymphoma, but the presenting features of the lesions in this patient were not typical of either disease. The unexpected lung biopsy result raised the possibility of dual pathology. A review of papers by Brennan has suggested that cases reporting sarcoidosis and lymphoma should only be accepted if biopsy specimens are reported independently from unrelated anatomical sites and appropriate clinical, radiographic, and biochemical features of each disease are present.\(^4\) It was also suggested that sarcoidosis should be demonstrated in two or more systems. Lymphoma may induce a sarcoid-like granulomatous reaction, although this is usually found at a site adjacent to the lymphoma.\(^2\) In previous studies this type of reaction has caused some confusion leading to over reporting of sarcoidosis in cases of lymphoma. In our patient the tissues from which biopsy specimens were taken were anatomically unrelated and a subsequent positive Kveim skin test confirmed sarcoidosis. The converse scenario where sarcoidosis affecting the eye could be mistaken for lymphoma seems unlikely. Both specimens were reported independently by pathologists expert in their respective fields and histological slides have subsequently been exchanged with agreement on both sides. Furthermore, immunological cell surface marker analysis of the eye lesion has confirmed a non-Hodgkin's lymphoma.

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.\(^1\) The association was noted by Brincker and Wilbeck who found 48 cases of cancer in 2544 Danish patients reported to have sarcoidosis.\(^4\) 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.\(^5,6\) 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.\(^7\) The main therapeutic approach to ocular adnexal lymphomas is low dose radiotherapy\(^8\) which achieves good local control at the cost of lens opacity in approximately 10%.\(^9\)

 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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