LETTERS TO THE EDITOR

Metachronous lymphoma of the breast and conjunctiva

EDITOR,—Primary lymphoma of the breast or ocular adnexae (orbit, conjunctiva, and eyelids) is rare, each accounts for 2% of all extra-nodal lymphomas. The most frequently encountered type of lymphoma at either site has been likened to the MALTomas (lymphoma of mucosa associated lymphoid tissue) arising in the gastrointestinal tract, although no direct association between the occurrence of lymphoma at these sites has been reported. We now describe a case of metachronous breast and conjunctival lymphomas with a 14 year interval.

CASE REPORT

A woman patient first presented in 1978 aged 55 with a left sided breast lump which was excised and histologically confirmed to be a primary breast lymphoma of low grade B cell lymphocytic type. Adjunctive postoperative radiotherapy was given and the patient has remained disease free since. In 1992 she was referred with a 2 year history of irritation and discomfort in the medial aspect of the left eye. On examination a flesh-coloured swelling measuring 1 by 0.5 cm was noted in the corresponding area of the bulbar conjunctiva which was thought to be a lymphoma.1

Histological examination of the excised conjunctival lesion confirmed the tumour to be a diffuse low grade B cell lymphocytic lymphoma (Figs A and B). Extensive investigations including a bone scan failed to demonstrate evidence of dissemination and the tumour was treated with a course of localised radiotherapy. In 1994 an abnormal area was noted at the site of the previous biopsy (Fig 2) which was thought to represent local recurrence; re-biopsy of this area demonstrated the presence of chronic inflammation but no evidence of tumour. To date there has been no recurrence of the conjunctival lymphoma.

COMMENT

Although up to 1-5% of all patients with extranodal lymphoma may develop involvement of the orbit or conjunctiva, primary lymphoma of the ocular adnexae is rare and constitutes 10% of all ocular tumours.2 Breast lymphoma accounts for 0-1-0-5% of all primary breast tumours. Most primary conjunctival3 and breast lymphomas belong to the MALToma group of extranodal lymphomas. These tumours are characterised by a long history of localised disease, a tendency to involve other mucosal sites in metastatic spread and a characteristic cytology in which follicle centre cells, centrocyte-like cells, and plasma cells are usually present, often with evidence of epithelial infiltration, the so called ‘lymphoepithelial’ lesion.4 Favourable prognostic features of ocular adnexal lymphomas are conjunctival origin, ‘low grade’ histology (lymphocytic or centrocytic morphology), and localised disease at presentation; all three features were present in this case.

MALT is not found in normal human conjunctiva but is acquired during life in a proportion of apparently asymptomatic individuals.5 Most MALTomas arise in lymphoid tissue that has been acquired as a result of some pre-existing inflammatory disorder for example, gastric MALToma associated with Helicobacter pylori infection and the MALTomas of the thyroid gland which arise against the background of autoimmune thyroiditis.6 We could find no abnormal antigenic stimuli to account for the development of the MALTomas which affected our patient. The occurrence of both breast and conjunctival lymphomas affecting the same patient is, we believe, unique. Both tumours exhibited similar histological appearances and similar immunophenotypes. Poor preservation of the archival material from the breast lesion has meant that it has not been possible to perform retrospective studies of immunoglobulin gene rearrangements that would be necessary to prove that the breast and conjunctival lesions were representative of the same neoplastic clone. There was no evidence of an underlying immune deficient state or an increased genetic susceptibility to neoplasia.

In conclusion, as a period of 14 years separated the appearance of the two lesions it seems most likely that the conjunctival lymphoma represents a second primary extranodal lymphoma of MALT type rather than a metastatic lesion from the breast neoplasm. This unusual occurrence may reflect an innate predisposition in our patient to develop lymphoma at various sites in the body, although no family history of lymphoma existed in this case.

Correspondence to: Dr M Wright, Princess Alexandra Eye Pavilion, Chalmers Street, Edinburgh EH3 9HA.

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Optic neuropathy associated with hypertrophic cranial pachymentignitis

EDITOR,—Hypertrophic pachymeningitis is a rare chronic inflammatory process involving the dura mater, tentorium, and the falx cerebri.7 Although causative agents have been implicated, the origin of this disorder is often obscure.8 Intracranial involvement is very rare, and to the best of our knowledge there is only one previously reported case involving complete visual loss.9 We present such a case.

CASE REPORT

A 72-year-old woman experienced a long history of headaches associated with a slow growing soft tissue mass in her right eyelid. In addition, the patient complained of a decrease in visual acuity in the right eye. Her vision was determined to be 20/200; however, no abnor- malities were found in the fundus. Computed tomographic (CT) scans and magnetic resonance imaging of the brain revealed no abnormalities. The patient was diagnosed as having retrobulbar optic neuritis, and underwent steroid treatment (1000 mg methylprednisolo- ne). Although the treatment was effective, two attacks subsequently occurred, and she was thus referred to our clinic.

Our initial examination revealed the best corrected visual acuity to be 20/40 in the right eye and 20/20 in the left eye. There was a

Figure 2 Scarring at the previous biopsy site with a birefringent pale pink elevation at its inferior border.

Figure 1 (A) The conjunctival lymphoma is characterised by numerous centrocyte-like cells with fewer plasma cells in a diffuse pattern, and an occasional prominent blood vessel. Mitotic activity is not a conspicuous feature of this lesion. Haematoxylin and eosin (×115).

(B) Immunocytochemistry for CD20 shows a strong positive reaction around the neoplastic cells (arrowed), confirming a B cell lineage. Staining for CD45 was also positive, but no reaction was observed with a panel of antibodies to T lymphocytes (×135).