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.¹ 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 lymphomas 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. Adjuvant 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.²

Histological examination of the excised conjunctival lesion confirmed the tumour to be a diffuse low grade B cell lymphocytic lymphoma (Figs 1A 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 recur-

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

rence; 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 presentation⁵; 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.⁶ 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.⁴ 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.

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Optic neuropathy associated with hypertrophic cranial pachymeningitis

EDITOR,—Hypertrophic pachymeningitis is a rare chronic inflammatory process involving the dura mater, tentorium, and the falx cerebri.^{1,2} Although several causative agents have been implicated, the origin of this disorder is often obscure.^{3,4} Intracranial involvement is very rare, and to the best of our knowledge there is only one previously reported case involving complete visual loss.⁵ 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 abnormalities 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 methylprednisolone). 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 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 ($\times 115$). (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 ($\times 135$).

reduced pupillary response to light in the right eye, as well as a pale optic disc in the right fundus. The visual field testing of the right eye demonstrated barring of the blind spot of Mariotte. Visual evoked response testing revealed the slight elongation of P100 latency. Fluorescein angiography revealed late staining of the right disc without other specific abnormalities. Exophthalmometric readings were 24 mm in the right eye and 20 mm in the left eye with a base of 111 mm. The orbital CT scan revealed severe hypertrophy of the extraocular muscles and orbital fat. Laboratory results showed an increase in thyrotropin binding inhibiting immunoglobulin (TB II) (23.7%) and triglyceride (188 mg/dl). Serum and cerebrospinal fluid tests for syphilis were negative. The antinuclear antibodies, anti-Ro (SS-A) antibodies, anti-La (SS-B) antibodies, rheumatoid factor, anticardiolipin antibodies were negative. After testing the patient was diagnosed as having endocrine ophthalmopathy. Radiation (20 Gy) was administered and the steroid therapy was continued. Although TBII became normal (5.5%), there was no beneficial effect. Sixteen months after the first attack, the colour of the optic disc was completely pale and visual acuity was zero in the right eye.

Six months later, the patient complained of a decrease in visual acuity in the left eye. Examination revealed the best corrected visual acuity to be 20/50 in the left eye. The visual field testing of the left eye demonstrated central scotoma within 10 degrees. A CT scan disclosed a thickening and enhancement of the dura mater, extending to the cavernous sinuses including the area of the optic foramina which appeared to be constricting the optic nerves. Severe hypertrophy of the extraocular muscles was unchanged (Fig 1). The chest radiograph was normal. Laboratory results showed an increase in C reactive protein (8.4 mg/dl); however, serum angiotensin II converting enzyme level was normal. An open biopsy revealed marked thickening of the temporal dura mater due to severe chronic inflammation, as well as fibrosis with small necrotic foci and perivasculitis with early granulomatous reaction. The reactive cells

Figure 2 Marked dense fibrosis of the temporal dura mater with inflammatory reaction containing perivasculitis (arrow) and a small necrotic foci with destructions of the collagen fibres (arrowhead). The reactive cells were mainly lymphocytes with some plasma cells and histiocytes (haematoxylin and eosin, $\times 30$).

were mainly lymphocytes with some plasma cells and histiocytes. No evidence of tuberculosis or fungal infection was found (Fig 2). At this point, the diagnosis of idiopathic hypertrophic pachymeningitis was established. The patient experienced a complete loss of vision for 1 month.

COMMENT

We believe this case to be the first in the ophthalmic literature, presenting bilateral optic neuropathy associated with idiopathic hypertrophic pachymeningitis. Different pathological entities which can produce thickening of the dura mater, sarcoidosis, late syphilis, rheumatoid arthritis, tuberculosis, intracranial fibromatosis, fungal infection, and so on should be considered in the differential diagnosis.⁴ In addition, Adler *et al*⁵ described a case of spinal aural pachymeningitis associated with pulmonary nodule, and concluded that this entity may be part of a systemic inflammation, like an autoimmune or connective tissue disorder. However, both clinical and laboratory findings were not suggestive for the existence of such conditions in the present case.

Initially, this case was treated as idiopathic optic neuritis and the steroid treatment was effective; however, owing to the development of hypertrophic pachymeningitis, the patient suffered a complete loss of vision. CT scans and magnetic resonance imaging of the brain and orbit revealed no abnormalities at first, but subsequent neuroimaging enabled us to establish the diagnosis accurately. Although cases of hypertrophic cranial pachymeningitis are very rare, it may be added to the differential diagnosis of optic neuropathy.

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A case of traumatic retinal avulsion at the optic nerve head

EDITOR—We present an unusual case of blunt trauma to the optic nerve head. Following trauma the patient developed a retinal avulsion at the optic nerve head with subsequent total retinal detachment which resulted in a rubeotic eye.

CASE REPORT

A 37-year-old woman fell down a staircase and injured the left side of her face and brow directly against the edge of a radiator. She lost consciousness briefly and was admitted to the local district general hospital. A brow laceration was sutured and she was referred to the ophthalmic department because of reduced acuity.

Examination of the right eye was normal. There was no perception of light in the left eye. There was a left anterior uveitis: 2+ cells, 3+ flare. There was a left afferent pupillary defect present. Funduscopy findings showed a ring of haemorrhage at the optic nerve head and a pale swollen retina with a cherry red spot at the macula (Fig 1). A provisional diagnosis of anterior traumatic optic neuropathy was made. A B-scan ultrasound revealed some

Figure 1 Contrast computed tomographic scan revealing granulomatous lesion involving the right orbit, bilateral cavernous sinuses (large arrows), the dura mater in the right middle fossa (small arrows), and the anterior wall of the pituitary fossa. Also, severe hypertrophy of the extraocular muscles are shown.