Figure 2 Left fundus, showing 25° excyclotorsion.

effects resulting from the association of blind spot displacement and bundle defects, when using computer programs excluding the testing of the presumed area of the blind spot, and letting the clinician believe that the blind spot is located within the unchecked surface of the visual field.

Examination of the fundus provides valuable information about the degree of cyclotorsion. Doubtful perimetric findings should be checked, using either manual kinetic perimetry or program options for automated perimetry which are specifically designed for evaluation of the blind spot area.^{2,4}

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IgG-κ extramedullary plasmacytoma of the conjunctiva and orbit

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A rare case of primary extramedullary plasmacytoma of the conjunctiva and orbit is reported. The patient was a 78-year-old woman who presented with tumour of subconjunctiva and orbit in the left eye.

Case report

A 78-year-old woman was referred on 22 October 1990 with a hard mass of the left lower conjunctiva, first noticed 2 months previously and gradually increasing in size. No history of

Figure 1 Facial view showing left-sided conjunctival tumour.

Figure 2 Computed tomography scan: horizontal section through orbit revealing a high density area in the left orbit.

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trauma could be elicited and there was no general complaint. Her family had no ocular problems. On admission, an elastic hard tumour was found in the lower conjunctiva measuring 36×15 mm (Fig 1). Its surface was smooth and there was no adhesion between the conjunctiva. No signs of inflammation were found in the skin overlying the lesion. There was no proptosis and no limitation of eye movements. Her best corrected visual acuity was 20/20 in both eyes. The intra-ocular pressures were normal. The anterior and posterior segments in both eyes were unremarkable except for cataract. Computed tomography of the brain revealed a high density area in the left orbit (Fig 2). There were no abnormal findings on blood examination, and tumour markers such as α fetoprotein, squamous cell carcinoma-related antigen (SCC), carbohydrate antigen 19-9 (CA19-9), carcinoembryonic antigen (CEA) were all within normal limits.

The left orbit was explored through a trans-conjunctival approach. The tumour was found to extend to the deep orbit and was excised as much as possible. Its content was friable and could not be excised totally.

Histopathological examination showed a nodular proliferation of plasma cells surrounded by adipose and connective tissue. The tumour was composed of a mixture of mature lymphocytes and plasma cells. Some of plasma cells demonstrated mitotic figures and binucleated plasma cells were also seen. Immunoperoxidase staining for κ light chain and also for IgG was positive and it was negative for IgM, A, λ light chain (Figs 3, 4). From these findings, we diagnosed this tumour as an IgG- κ plasmacytoma.

Ear, nose, and throat examination revealed no clue. The general physical examination of the patient was unremarkable, and all laboratory tests, including serum immunoelectrophoresis and urinalysis for such as Bence-Jones protein, were normal. Computed tomography of the skull, chest, and vertebrae was normal. Skull, spine, and long bone x rays and liver, spleen, and bone scans were all normal. Bone marrow aspiration was normal. The blood viscosity was slightly decreased. This case was finally diagnosed as primary extramedullary plasmacytoma of the conjunctiva and orbit.

After the subtotal resection of the tumour, the patient received systemic chemotherapy with endoxan and prednisolone. This had an apparent effect on the residual tumour and subsequently the tumour disappeared. The patient has been followed up for 18 months and still has exhibited no lesion elsewhere in the body.

Comment

Extramedullary plasmacytoma (EMP) is a solitary plasma cell tumour of bone or soft tissue and it is classified as a malignant lesion. However, from its benign behaviour, EMP is considered a separate disease from multiple myeloma. EMP represents only 3% of plasma cell tumours^{1,2} and approximately 80% of EMPs develop in the walls of the upper respiratory tract or nasopharynx. Involvement of the gastrointestinal tract or lymph nodes follows infrequently. Orbital mani-

Fig 3A

Fig 3B

Figure 3 Photomicrograph of histological appearance of the section showing plasmacytic lymphoproliferation (A, haematoxylin and eosin, $\times 25$). Abundance of mature looking plasma cells and mitotic figures (arrow) are seen (B, haematoxylin and eosin, $\times 100$)

Figure 4 Immunoperoxidase staining of the specimen for κ light chain ($\times 170$).

festation of EMP is exceedingly rare. To our knowledge, it has been reported in only 10 cases in the world literature,³⁻⁸ including three Japanese cases. In this report, we have described a new case of orbital EMP, with immunocytochemical results.

However, rarely, multiple myeloma also involves the orbit. Rodman and Font⁷ were able to identify 31 cases. The mean survival of EMP is 8.3 years compared with a mean survival of 20 months for multiple myeloma.⁹ To establish that the orbital involvement is not an early manifestation of multiple myeloma, long term follow up, including systemic evaluation, is required. In

this sense, the exact incidence of orbital EMP is difficult to determine.

Although orbital lesions characteristically present with proptosis, our patient showed no proptosis, only presenting lower lid retraction owing to mass effect. Computed tomography demonstrated that the tumour involved the left lower orbit.

The nature of six recently reported cases of orbital EMP was disclosed by immunocytochemical staining, with one case of IgG- κ ,¹⁰ one case of IgD- λ ,¹¹ one case of IgM- κ ,⁵ one case of biclonal light chain,¹² and two cases of IgG- λ .^{6,13} Our specimen demonstrated a monoclonal proliferation of IgG- κ .

As in other neoplastic processes, EMPs are thought to arise from a single clone; hence the immunocytochemistry shows monoclonality or a biclonal light chain. In contrast, hyperplastic disorders demonstrate polyclonality.¹⁴ Thus immunocytochemical staining may help to confirm the diagnosis when the histopathological appearance is ambiguous.

The treatment of plasmacytoma consists initially of radiotherapy with surgery and chemotherapy. Our patient was treated by local excision of the tumour followed by systemic chemotherapy which led to the total regression of the residual tumour. Because informed consent was not obtained from the patient, radiotherapy was not indicated. She was anxious about irradiation.

In managing a patient with an orbital tumour, long term careful follow up, including systemic

evaluation, is essential and any sign of recurrence and/or progression should be treated promptly.

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