Fig. 2. Papillary tumour composed of cells with a strongly eosinophilic cytoplasm, and moderately pleiomorphic nuclei, containing nucleoli. The cells show decapitation secretion (arrows) (H&E, $\times 400$). Inset: The cells stained positive with antibody against epithelial membrane antigen (EMA) (EMA, $\times 400$).

evidence of lymphangio-invasion. The tumour cells stained positive with antibodies against epithelial membrane antigen (EMA) (Fig. 2) and S-100 and negative with prostate specific antigen (PSA), excluding metastatic prostatic carcinoma.

Comment

Only three published cases^{1,2,6} have had a clinical presentation of apocrine adenocarcinoma similar to our case (Fig. 1). Only Aurora and Luxenberg² provided a clinical photograph of this tumour resembling our lesion. In other reports, the apocrine adenocarcinoma resembled a chalazion,^{3,4,7} or was extensive, with orbital infiltration at first presentation.^{4,5}

Analysis of seven previous cases¹⁻⁶ and ours showed that 5 patients were men and 3 were women. The duration of disease at the time of diagnosis varied between 2 and 78 months. Of 6 localised apocrine sweat gland carcinomas of the eyelid that were (widely) excised, 3 recurred, eventually leading to orbital exenteration and/or lymph node dissection in 2 patients.^{1,4} Two patients with extensive lesions, involving the orbit, underwent exenteration as a primary procedure, preceded by radiotherapy in 1 patient.^{4,5} Two of the 8 patients eventually died of systemic metastases after 2 years of follow-up.^{1,4} In 3 patients the reported follow-up period was less than 2 years.

The differential diagnosis from other lid margin tumours, including sebaceous gland carcinoma, Merkel cell tumour, eccrine adenocarcinoma and Moll gland cystadenoma, melanoma, or from metastatic lid lesions, may be difficult.⁶ Our case showed definite features indicative of apocrine origin, including strongly eosinophilic cytoplasm, iron-positive intracellular pigment and decapitation secretion. The lid and prostate adenocarcinoma in our patient had markedly different histological features and we concluded they were not related.

The malignant potential of adenocarcinoma of the gland of Moll prompts radical surgical excision and regular follow-up. At presentation, regional lymph node involvement, other malignancies or metastatic disease should be ruled out.

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Sir,

Double bitemporal hemianopia

Both tilted disc syndrome and sellar tumours are well known to cause bitemporal hemianopia. The temporal field defect occurring in tilted disc syndrome is thought to be due to a combination of refractive scotoma caused by nasal fundus ectasia and segmental neuroretinal hypoplasia.^{1,2} The bitemporal hemianopia in such cases is typically incomplete and confined to the supero-temporal quadrants, has sloping margins, fails to respect the vertical meridian, and is characterised by lack of progression. In contrast, true bitemporal hemianopia resulting from chiasmal compression is usually more complete, respects the vertical meridian, and progresses as the tumour enlarges. The coincidence of sellar tumour and tilted disc syndrome occurs only very rarely.

We report the case of a 64-year-old woman with simultaneous pituitary adenoma and bilateral tilted disc syndrome who exhibited bitemporal hemianopia. When

Fig. 1. Fundus photograph of the right eye, showing extreme tilting of disc. The inferior margin of the disc can hardly be identified.

Fig. 2. Fundus photograph of the left eye, showing the obliquely oriented oval disc, large crescent and hypopigmented inferior fundus – changes characteristic of tilted disc syndrome.

a patient with tilted disc syndrome also harbours a sellar tumour, the field defects caused by these two disease entities will be superimposed, resulting in diagnostic confusion and delay.

Case report

The patient, a 64-year-old woman, had a minor cerebrovascular accident (CVA) in November 1993. She experienced transient mouth deviation to the left, and slurred speech that lasted for 10 days. A suprasellar mass was found incidentally during a neuroimaging study, and her neurologist therefore referred her to the ophthalmology department for a visual field test.

She stated that she had suffered from poor vision all her life, but had never worn spectacles. She denied any recent increase in visual blurring.

In August 1995, an ocular examination was performed. Her corrected acuity was 20/50 in the right eye with -6.75 spherical lens and -1.5 cylinder lens, and 20/40 in the left eye with -3.75 spherical lens and -1.0 cylinder lens. Colour testing was performed using the Ishihara pseudoisochromatic colour plates. The patient could perceive 5 of 13 in the right eye, and 9 in the left eye; the colour vision was impaired in both eyes.

The fundus findings were most striking with both discs obliquely oval, and the upper discs showing relative elevation compared with the lower portion. Large crescents were found inferior to both discs (Figs. 1, 2).

Humphrey perimetry with a minus lens showed bilateral upper temporal field defects which passed over the vertical midline. The margins of the defect were irregular and sloping (Fig. 3).

Brain MRI in August 1995 disclosed a sellar mass with suprasellar extension; coronal imaging showed the chiasm was compressed upwards by the tumour underneath. Pituitary adenoma was strongly suspected (Fig. 4). MRI also demonstrated that both optic nerves entered the eyes at an extremely oblique angle.

Hormone studies were completely normal. The patient refused to have an operation until January 2000. Her visual acuity deteriorated rapidly; in May 2000 the corrected visual acuity in the right eye was counting fingers, in the left eye hand motion only, and perimetry showed that there was only a very small field remaining in the nasal upper area of the right eye. Brain MRI demonstrated that the sellar tumour had grown bigger with more extensive chiasmal compression. A transsphenoid approach tumour excision was performed

Fig. 3. Humphrey perimetric examination (30-2) showing incomplete bitemporal hemianopia with sloping margins and crossing of the vertical meridian.

Fig. 4. Brain T1-weighted MR imaging (coronal view) shows the space-occupying lesion at the suprasellar area and the chiasm being pushed upwards by the tumour underneath.

by a neurosurgeon. During the operation, a solid tumour with blood clot was found, which pathological examination showed to be a pituitary adenoma.

Comment

The visual field defects produced by tilted discs have been thought to be due to the combination of fundus ectasia and segmental neuroretinal hypoplasia. Although a tilted disc is considered as a diffusely hypoplastic optic nerve, all the retinal sectors are involved but the inferonasal sector is more affected. The field loss is more pronounced in the superotemporal quadrant, so the bitemporal field defect associated with tilted discs needs to be distinguished from the field defects of chiasmal compression. According to Brazitikos and Safran's studies, visual field alterations by means of the Octopus G1 global analysis program demonstrate that an increase in short-term fluctuation usually indicates an acquired lesion of the anterior visual pathways, while such fluctuation rarely occurs in congenital dystrophic disorders of neural pathways such as tilted disc syndrome. Thus an increase in short-term fluctuation in cooperative patients may be a clue for differentiating congenital neuroretinal dystrophy from acquired defects in anterior visual pathways.³⁻⁵

Most patients with tilted discs are not felt to require examination for suprasellar tumour. However, a few reports of disc dysplasia with anterior visual pathway tumours have been published in the literature.^{6,7} Keane⁶ reported two cases of congenital optic disc anomaly with concomitant chiasmal anomaly: one a dysgerminoma and the other a pituitary adenoma. Taylor⁷ reported 7 young patients with dysplastic discs associated with suprasellar tumours, including glioma, craniopharyngioma and fibrillary astrocytoma. In these cases, the anterior visual pathway tumours are thought to arise from developmental anomalies dating from embryonic life. Some authors have suggested that the association of anomalous optic discs with suprasellar tumour is more than incidental and may reflect a common developmental abnormality.

Pituitary adenoma, on the other hand, is not believed to be a form of developmental abnormality or hamartoma. Hence, the association of pituitary adenoma with tilted discs is thought to be coincidental and occurs only very rarely.

In the author's opinion, it is not rational to evaluate all patients with tilted disc syndrome with neuroimaging studies. Sequential examination of visual acuity and visual fields with a carefully selected program is indicated, however; any progressive change in these parameters may suggest a dual central nervous system pathology.

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