plexiform neuromas an attempt is made to look for sphenoid dysplasia, in the absence of ocular symptoms and signs it is probably reasonable to adopt the wait-and-watch policy.

References

1. Sakai A, Suzuki K. Von Recklinghausen's disease and its pathogenesis. Nippon Rinsho. Jpn J Clin Med 1995;53:2688–90.
2. Ahn MS, Jackler RK, Lustig LR. The early history of neurofibromatosis: evolution of the concept of neurofibromatosis type 2. Arch Otolaryngol Head Neck Surg 1996;122:1240–9.
3. Friedman JM. Epidemiology of neurofibromatosis type 1. Am J Med Genet 1999;89:1–6.
4. Neurofibromatosis conference statement (National Institutes of Health consensus development conference). Arch Neurol 1998;45:575–8.
5. DeBella K, Szudek J, Friedman JM. Use of National Institutes of Health criteria for diagnosis of NF1 in children. Paediatrics 2000;105:608–14.
6. Khairallah M, Messaoud R, Ladjimi A, *et al*. Association of spheno-orbital dysplasia with plexiform neuroma in von Recklinghausen's neurofibromatosis. J Fr Ophtalmol 1999;22:975–8.
7. Abassi-Bakir D, Graiess-Tlili K, Turky A, *et al*. Cranio-facial bone abnormalities in von Recklinghausen neurofibromatosis. Ann Radiol 1995;38:139–44.
8. Kaste SC, Pivnick EK. Bony orbital morphology in neurofibromatosis type 1 (NF1). J Med Genet 1998;35:628–31.
9. Macfarlane R, Levin AV, Weksberg R, *et al*. Absence of the greater sphenoid wing in neurofibromatosis type 1: congenital or acquired. Case report. Neurosurgery 1995;37:129–33.
10. Harkens K, Dolan KD. Correlative imaging of sphenoid dysplasia accompanying neurofibromatosis. Ann Otol Rhinol Laryngol 1990;99:137–41.
11. Havlik RJ, Boaz J. Cranio-orbital-temporal neurofibromatosis: are we treating the whole problem? J Craniofac Surg 1998;9:529–35.
12. Krastinova-Lolov D, Hamza F. The surgical management of cranio-orbital neurofibromatosis. Ann Plast Surg 1996;36:263–9.
13. Snyder BJ, Hanieh A, Trott JA, *et al*. Transcranial correction of orbital neurofibromatosis. Plast Reconstr Surg 1998;102:633–42.

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Sir,

Apocrine sweat gland carcinoma

Apocrine sweat gland carinoma (Moll adenocarcinoma) of the eyelid is extremely rare. The first case report was by Stout and Cooley in 1951.[1] Since then, only seven published cases[2–7] have been consistent with the histopathological criteria for these lid lesions,[2] of which three were illustrated with clinical photographs.[2,5,7] In these reports, the clinical presentation varied between a chalazion-like lid lesion and a blue-red raised nodular mass at the eyelid margin. Apocrine sweat gland carcinoma may spread to regional lymph nodes, and may cause metastatic death.[1,4] The histopathology of this tumour is characterised by an invasive adenocarcinoma with gland-like structures consisting of eosinophilic or opaque glassy cells forming irregularly shaped lumina of varying size.[7]

Case report

An 80-year-old Caucasian man was referred with a history of a rapidly growing painless lump on his right upper eyelid for 2 months. On examination there was an elevated blue-brown soft-elastic tumour, 6 × 4 mm at the medial upper lid margin (Fig. 1). There was notable lash loss from the site of the tumour. The palpebral conjunctiva of the right upper lid showed no abnormalities. Further ophthalmic examination was unremarkable. The regional lymph nodes were not enlarged. His past medical history did not include any malignancy. Full-thickness, wide block excision was performed. Systemic investigation revealed a biopsy-proven well-differentiated carcinoma of the prostate gland (Tlla GI Nx M0). A bone scan was negative for metastasis. The patient was treated for his prostate tumour with a course of hormone therapy. Over a 2 year follow-up there was no recurrent disease of the eyelid or in his regional lymph nodes. His prostate responded well to treatment.

Pathology. Histopathological examination revealed at the eyelid margin a partly cystoid papillary tumour, not in contact with the epidermis, hair shafts or tarsal Meibomian glands. The tumour was composed of cells with a strongly eosinophilic cytoplasm, and moderately pleiomorphic nuclei, containing nucleoli (Fig. 2). The cells showed at several places decapitation secretion, and contained iron-positive, PAS-positive diastase-resistant granules. There was moderate mitotic activity, and

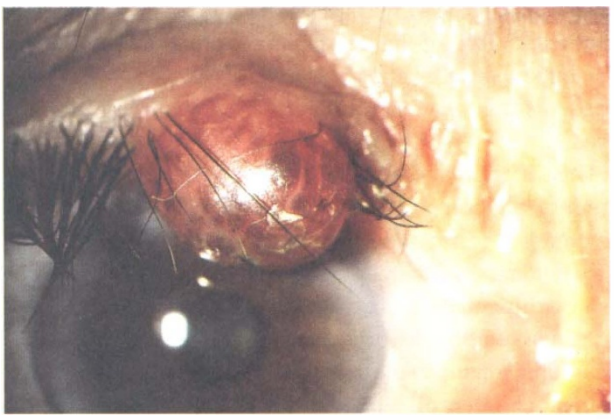

Fig. 1. *Clinical appearance of an apocrine sweat gland carcinoma of the right upper lid of an 80-year-old man. The nodular blue-brown soft elastic tumour measured 6 × 4 mm.*

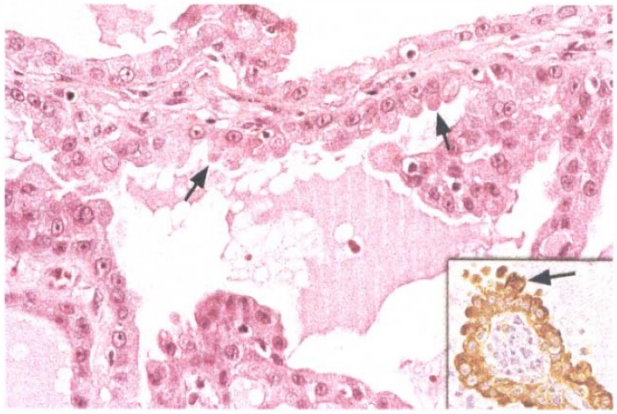

Fig. 2. *Papillary tumour composed of cells with a strongly eosinophilic cytoplasm, and moderately pleiomorphic nuclei, containing nucleoli. The cells show decapitation secretion (arrows) (H&E, ×400). Inset: The cells stained positive with antibody against epithelial membrane antigen (EMA) (EMA, ×400).*

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 Luxenberg[2] 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 cases[1–6] 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 mestastatic disease should be ruled out.

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References

1. Stout AP, Cooley SGE. Carcinoma of sweat glands. Cancer 1951;4:521–36.
2. Aurora AL, Luxenberg MN. Case report of adenocarcinoma of glands of Moll. Am J Ophthalmol 1970;70:984–90.
3. Futrell JW, Krueger GR, Chretien PB, Ketcham AS. Multiple primary sweat gland carcinomas. Cancer 1971;28:686–91.
4. Ni C, Wagoner M, Kieval S, Albert DM. Tumours of the Moll's glands. Br J Ophthalmol 1984;68:502–6.
5. Thomson SJ, Tanner NS. Carcinoma of the apocrine glands at the base of eyelashes: a case report and discussion of histological diagnostic criteria. Br J Plast Surg 1989;42:598–602.
6. Seregard S. Apocrine adenocarcinoma arising in Moll gland cystadenoma. Ophthalmology 1993;100:1716–9.
7. Rodgers IR, Jakobiec FA, Hidayat AA. Eyelid tumours of apocrine, eccrine, and pilar origins. In: Albert DM, Jakobiec FA, editors. Principles and practice of ophthalmology: clinical practice. Philadelphia: WB Saunders, 1994:1777–9.

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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