Bilateral eyelid myxomas in Carney's complex

Hans E Grossniklaus, Ian W McLean, John J Gillespie

Abstract

A 21-year-old man with a history of an excised soft tissue mass of the groin and spotty cutaneous pigmentation underwent excision of nodules of the right lower and left upper eyelids. The patient subsequently had a cutaneous mass of the left ear removed. All excised lesions were classified as myxomas. A diagnosis of multiple myxoma, spotty pigmentation, and endocrine overactivity (Carney's) complex was made.

In 1985 Carney and associates described a complex consisting of various combinations of myxomas, spotty cutaneous pigmentation, and endocrine overactivity.[1] The importance of recognition of this complex is that a significant proportion of patients with the complex have a cardiac myxoma, a potentially lethal lesion.[1,2]

Case report

A 21-year-old man with lesions of the right lower and left upper eyelids which had been present for two years was examined (Figs 1A, B). There were 0·5×0·5×0·5 cm soft nodular lesions of the right lower and left upper eyelids. Small, inconspicuous areas of spotty pigmentation were present on the patient's face. The rest of the ophthalmic examination showed nothing abnormal.

The eyelid nodules were excised. Histological examination of the right lower eyelid lesion showed a fibromyxoid stroma covered by epidermis. The left upper eyelid lesion was composed of a loosely myxoid stroma containing spindle and stellate cells covered by epidermis (Figs 2A, B). The patient's past history was significant for a recently excised soft tissue lesion of the left groin. Review of the histology of the groin lesion showed findings similar to those of the eyelid lesions.

Approximately one month after examination a 0·8×0·6×0·3 cm cutaneous nodule of the left ear was excised. Histological examination showed findings virtually identical to those of the eyelid and groin lesions. The groin, eyelid, and ear lesions were classified as myxomas. Owing to the spotty pigmentation and the histopathological appearance of the excised lesions the diagnosis of Carney's complex was made. The results of the rest of the physical examination were normal. An echocardiogram and serum cortisol level were normal. Subsequently it was found that one of the patient's parents and two of his siblings had multiple myxomas and spotty cutaneous pigmentation.

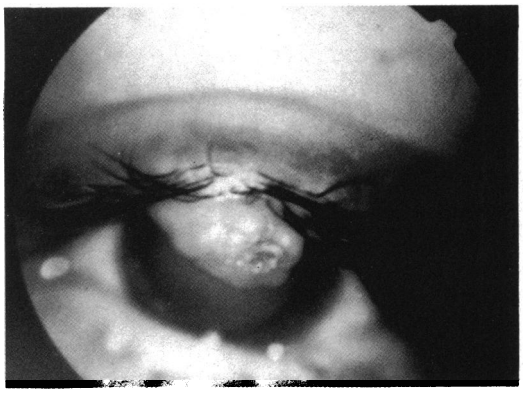

Figure 1B Left upper eyelid nodule is similar in appearance to the right lower eyelid nodule.

Discussion

The presence of multiple myxomas, spotty pigmentation, and a positive family history suggest a diagnosis of Carney's complex. Carney and coworkers found reports of 36 patients in the literature with various combinations of myxomas, spotty cutaneous pigmentation, and endocrine overactivity, and they combined these patients with four new patients in their original definition of this complex.[1] There are various histological counterparts of the spotty cutaneous pigmentation including lentigo, ephelis, junctional naevus, compound naevus, and atypical

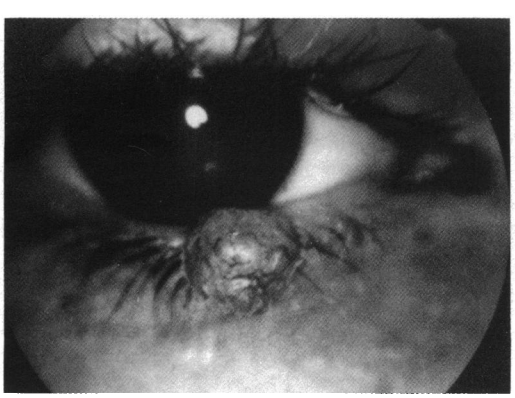

Figure 1A Nodular, white to yellow right lower eyelid lesion.

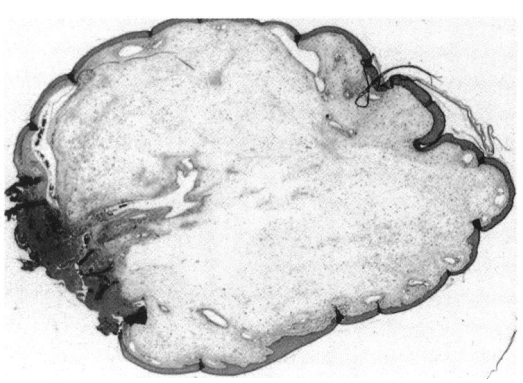

Figure 2A The left upper eyelid nodule is composed of a myxoid stroma covered by overlying epidermis. (Haematoxylin-eosin, ×153.)

L F Montgomery Eye Pathology Laboratory, Emory Eye Center, Atlanta, Ga, USA
H E Grossniklaus

Department of Ophthalmic Pathology, Armed Forces Institute of Pathology, Washington DC
I W McLean

Department of Pathology, St John's Hospital, Springfield Ill
J J Gillespie

Correspondence to:
Dr Hans E Grossniklaus, L F Montgomery Eye Pathology Laboratory, Emory Eye Center, 1327 Clifton Road, NE, Atlanta, Ga 30322, USA.

Accepted for publication 4 October 1990

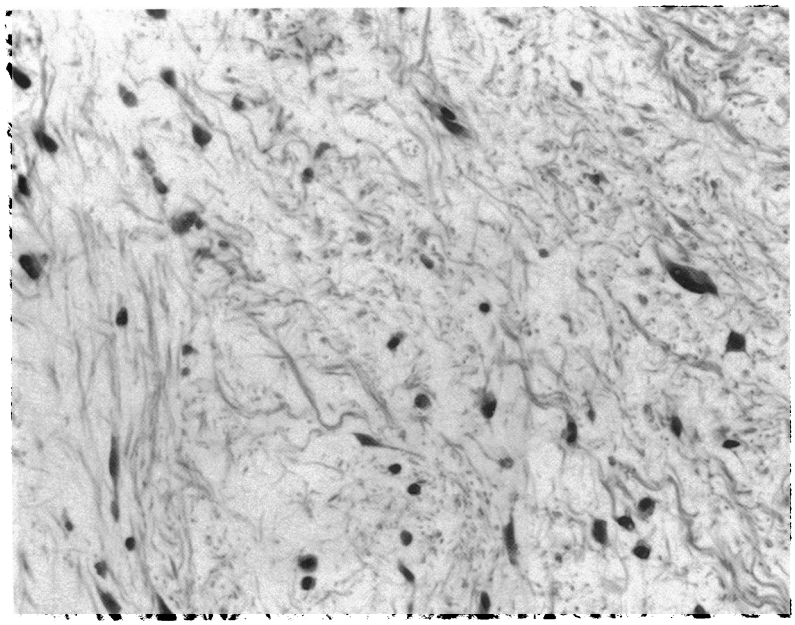

Figure 2B Higher magnification of the left upper eyelid lesion shows spindle and stellate shaped cells in a myxoid background. (Haematoxylin-eosin, ×300.)

blue naevus.[1] Our patient had spotty facial pigmentation with the clinical appearance of benign naevi; none of his pigmented lesions were biopsied.

Endocrine abnormalities in patients with this complex include adrenocortical disease, Sertoli cell tumours, and acromegaly.[1] The presence of two of the three criteria is needed to make the diagnosis. A subsequent report suggests an autosomal dominant inheritance pattern to this complex.[3] The syndrome may be due to a genetically determined malfunction of mesenchymal cells causing 'myxomatosis', akin to neurofibromatosis.[2] Several reports published after Carney and coworkers' original article describe other patients with the complex.[4-8]

Ophthalmic findings in patients with this complex include spotty pigmentation of the eyelids, pigmented lesions of the caruncle or conjunctival semilunar folds, and myxomas of the eyelids.[8] Cutaneous myxomas in this complex present most often in the skin of the head and neck, followed by the trunk, pelvic area, and limbs.[2] The cutaneous myxomas are most commonly of the eyelid, ear, and nipple. Eyelid myxomas are reported to occur in 10% of patients with the syndrome.[8] It is important to note that cutaneous myxomas are detected prior to the cardiac myxoma in the majority of patients.[2] Therefore, in a patient with multiple cutaneous myxomas, spotty pigmentation, and/or endocrine overactivity, cardiac ultrasonography should be performed.

We thank Dr J Aidan Carney for examining the patient and providing information on his family.

The opinions or assertions contained herein are the private views of the authors and should not be construed as being official views of the Department of the Army or the Department of Defence

1 Carney JA, Gordon H, Carpenter PC, Shenoy V, Go VLW. The complex of myxomas, spotty pigmentation, and endocrine overactivity. *Medicine* 1985; **64**: 270–83.
2 Carney JA, Headington JT, Su WP. Cutaneous myxomas. A major component of the complex of myxomas, spotty pigmentation, and endocrine overactivity. *Arch Dermatol* 1986; **122**: 790–8
3 Carney JA, Hruska LS, Beauchamp GD, Gordon H. Dominant inheritance of the complex of myxomas, spotty pigmentation, and endocrine overactivity. *Mayo Clinc Proc* 1986; **61**: 165–72.
4 Reed OM, Mellette JR, Fitzpatrick JE. Cutaneous lentiginosis with atrial myxomas. *J Am Acad Dermatol* 1986; **15**: 398–402.
5 Leedman PJ, Cohen AK, Matz LR. The complex of myxomas, spotty pigmentation and endocrine overactivity. *Clin Endocrinol* 1986; **25**: 527–34.
6 Rowe MH, Mullany CJ, Leitl G. Spotty pigmentation and atrial myxoma: a case report of Carney' s complex. *Med J Aust* 1988; **149**: 10–2.
7 Reinecke P, Frenzel H, Hort W, Willberg B. The combination of atrial myxoma and multiple skin myxomas: a characteristic symptom complex? *Dtsch Med Wochenschr* 1987; **112**: 1902–5.
8 Kennedy RH, Waller RR, Carney JA. Ocular pigmented spots and eyelid myxomas. *Am J Ophthalmol* 1987; **104**: 533–8.