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Pathology in Focus

Turban tumour with involvement of the parotid gland

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Abstract
Familial autosomal dominant cylindromatosis (FADC, turban tumour syndrome, Brooke-Spiegler-syndrome and many more, MacKusick catalogue numbers 123850, 313100) is a rare hereditary disease usually presenting in the second or third decade. With female preponderance dermal cylindromas predominantly arise in hairy areas of the body with approximately 90 per cent on the head and neck. Transformation to malignancy seems to be scarce. Although cylindromas of the skin resemble basal cell adenomas of the salivary gland, there is usually no salivary gland involvement. On the other hand, patients with basal cell adenomas of a salivary gland usually do not show dermal lesions. We report one of the rare cases of FADC combined with multiple basal cell adenomas of the parotid glands and present a review of the literature.

Key words: Parotid neoplasms; Adenoma; Skin neoplasms

Introduction

Cylindromatosis (MacKusick MIM Numbers 123850, 313100)(MacKusick, 1992) may be inherited as an autosomal dominant trait with a different degree of penetrance and possible female preponderance. Recently, the linkage of cylindromatosis to chromosome 16q12-q13 has been pointed out (Biggs *et al.*, 1995). Schuermann and Weber (1937), Vernon *et al.* (1988), Burrows *et al.* (1992) and many others describe a disease with multiple cylindromas, trichoepitheliomas, and eccrine spiradenomas, but without

TABLE I
SURVEY OF CASES WITH CO-EXISTENT BASAL CELL ADENOMAS OF SALIVARY GLANDS AND DERMAL ADNEXAL TUMOURS

Case	Sex/age at histological diagnosis	Salivary gland tumour	Dermal tumours (exclusively in the head and neck region)	Familial occurrence reported	Authors
1	70/m	MBCA of both parotid glands	Cylindromas and trichoepitheliomas	No	Headington *et al.* (1977)
2	43/m	MBCA of both parotid glands	Cylindromas; trichoepitheliomas; eccrine spiradenomas	Yes	Reingold *et al.* (1977)
3	68/m	MBCA of both submandibular glands	Cylindromas	No	Bataskis and Brannon (1981b)
4	74/m	MBCA of one parotid gland	Cylindromas	No	Batsakis and Brannon (1981b)
5	?/m	MBCA of both parotid glands	Cylindromas	Yes	Alawi *et al.* (1982)
6	54/f	MBCA of both parotid glands	Cylindromas	Yes	Herbst and Utz (1984)
7	63/m	MBCA of one parotid gland	Cylindromas, *malignant* transformation, lung metastases	No	Pingitore and Campani (1984)
8	55/m	MBCA of both parotid glands	Cylindromas, eccrine spiradenomas	Yes	Ferrandiz *et al.* (1985)
9	66/m	MBCA of both parotid glands; *malignant* transformation	Cylindromas, trichoepitheliomas, eccrine spiradenomas	Yes	Hyman *et al.* (1988)
10	78/m	MBCA of both parotid glands	Cylindromas, trichoepithelioma, *malignant* dermal cylindromas	No	Rockerbie *et al.* (1989)
11	58/m	Monomorphic adenomas of dermal cylindroma type	Cylindromas, trichoepitheliomas, eccrine spiradenomas	Yes	Zarbo *et al.* (1985)
12	56/m	MBCA of both parotid glands	Cylindromas	Yes	own case

MBCA = membranous basal cell adenomas

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affectation of the salivary glands. In 90 per cent of the FADC patients tumours are found on the head and neck (Crain and Helwig, 1961). In these patients the disease is commonly called turban tumour syndrome. In general, lesions may arise from any hairy region of the body and the disease usually presents in the second or third decade. Transformation to malignancy (i.e. metastasis) seems to be extremely rare, but is on record (Urbanski *et al.*, 1985; Hammond *et al.*, 1990). The present report describes the coincidence of multiple dermal cylindromas of the skin and membranous basal cell adenomas of the salivary glands. This coincidence is rare and has been noted before only in 11 case reports (for an overview see Table I).

Case report

In 1987, a 56-year-old Caucasian male presented with multiple slow-growing tumours on the scalp and a gradually enlarging mass in the left parotid gland. He had a history of multiple tumoral lesions that had been excised from the scalp at the age of 25 and stated that his father and his older brother also had had several nodules on their scalps. They had died (of causes unknown to the patient) at the ages of 73 and 66, respectively. Apart from his father and his brother no-one else in his family suffered from dermal lesions. On admission, the patient's scalp had more than 20 pink-coloured tumours (diameters, between 0.5 and 2.5 cm) with a consistency ranging from smooth to firm (Figure 1). The left cheek was swollen by a firm, painless, well-demarcated movable mass (5 cm in diameter) located in the parotid gland. No signs of facial palsy were evident. His health was otherwise inconspicuous, all laboratory studies were normal. Findings of fine needle aspiration cytology of the parotid gland tumour were equivocal. Consecutive lateral parotidectomy along with excision of the 20 most prominent cutaneous nodules was performed. Histological investigation of the skin lesions revealed numerous, irregular shaped islands of basophilic staining tumour cells surrounded by a prominent PAS-positive and diastase-resistant acidophilic hyaline material which was also present as hyaline droplets intermixed with the tumour cells. Most of the cell islands were composed of two cell types: dark-nucleated small pallisading cells in the periphery and (more centrally located) larger cells with light nuclei. Duct-like structures (some with amorphous luminal material) were present in some of these islands. The lesions were interpreted as dermal eccrine cylindromas (Figure 2). The multicentric parotid gland tumour was composed of several nodular epithelial masses of variable size, most of which were surrounded by prominent PAS-diastase-positive hyalinized sheaths. In some areas this hyaline material severed the epithelial cells forming large acellular areas, exclusively comprised of PAS-diastase-positive hyaline material, and were found to be basal cell adenomas of the dermal subtype (Figure 3).

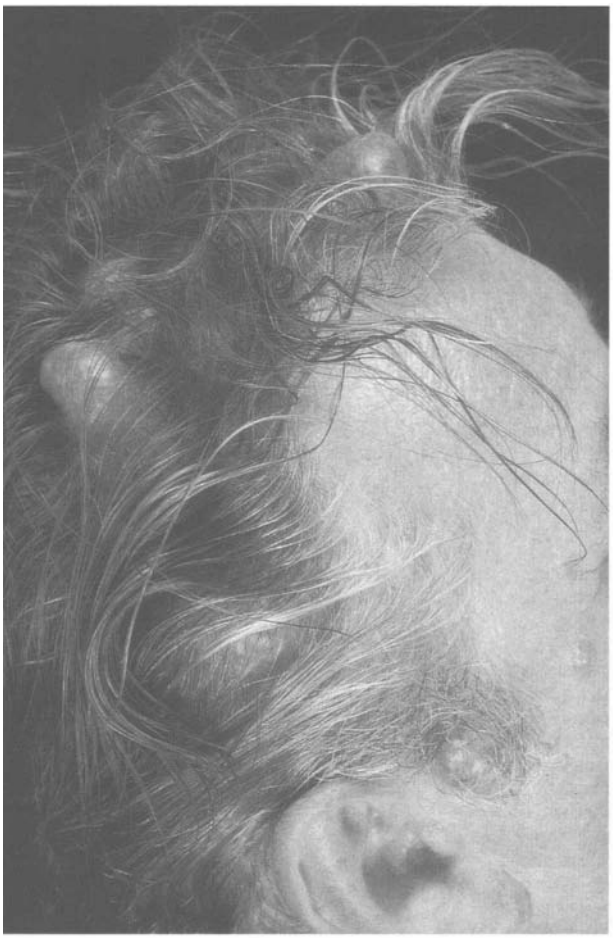

FIG. 1a

Multiple pink-coloured tumours emerging from the scalp with a diameter between 0.5 and 2.5 cm. The consistency ranged from smooth to firm. Fifty-six-years-old Caucasian man.

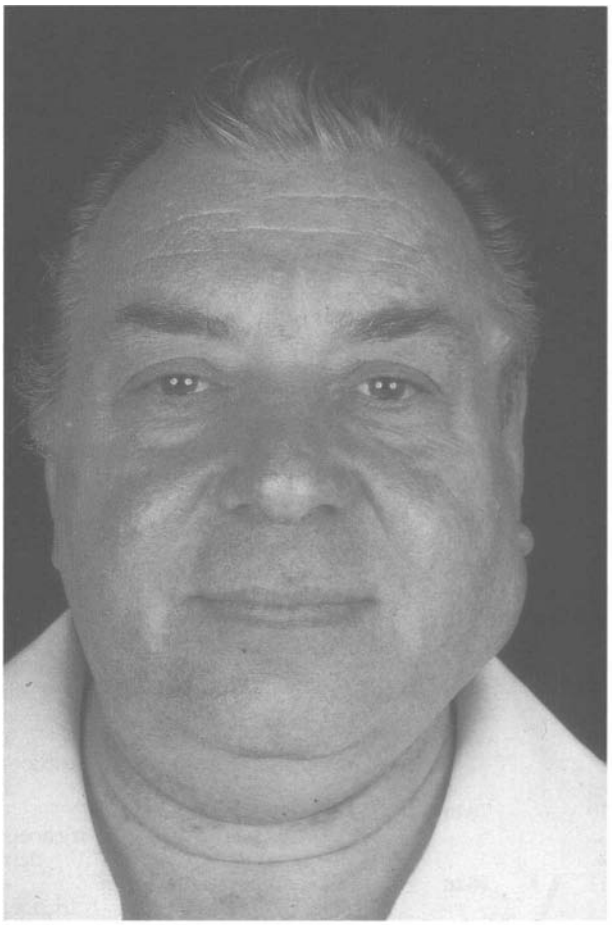

FIG. 1b

The left parotid gland region is protruded by a painless, well-demarcated movable mass (5 cm in diameter). Facial nerve function is not restrained.

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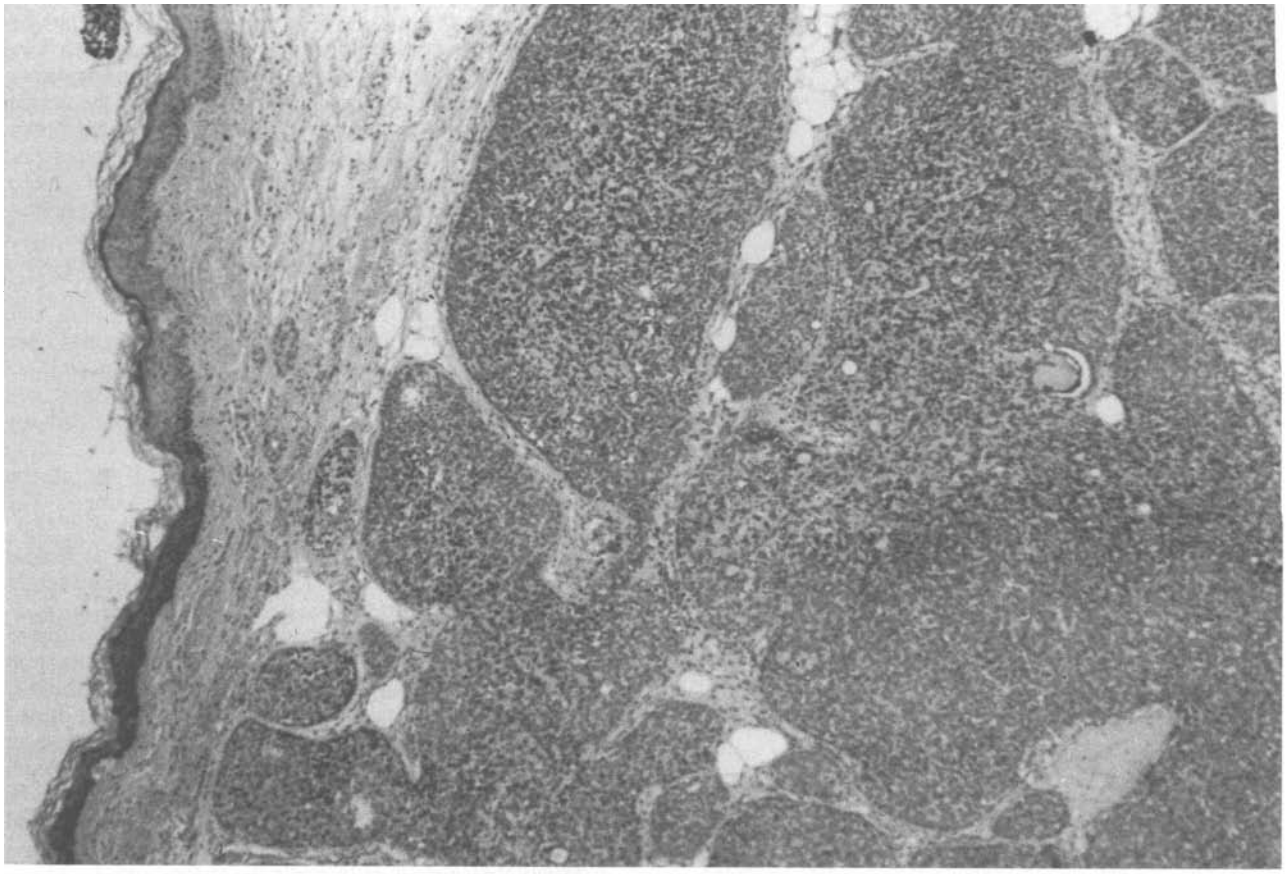

FIG. 2
Dermal eccrine cylindroma of the scalp. (H & E; × 12.5).

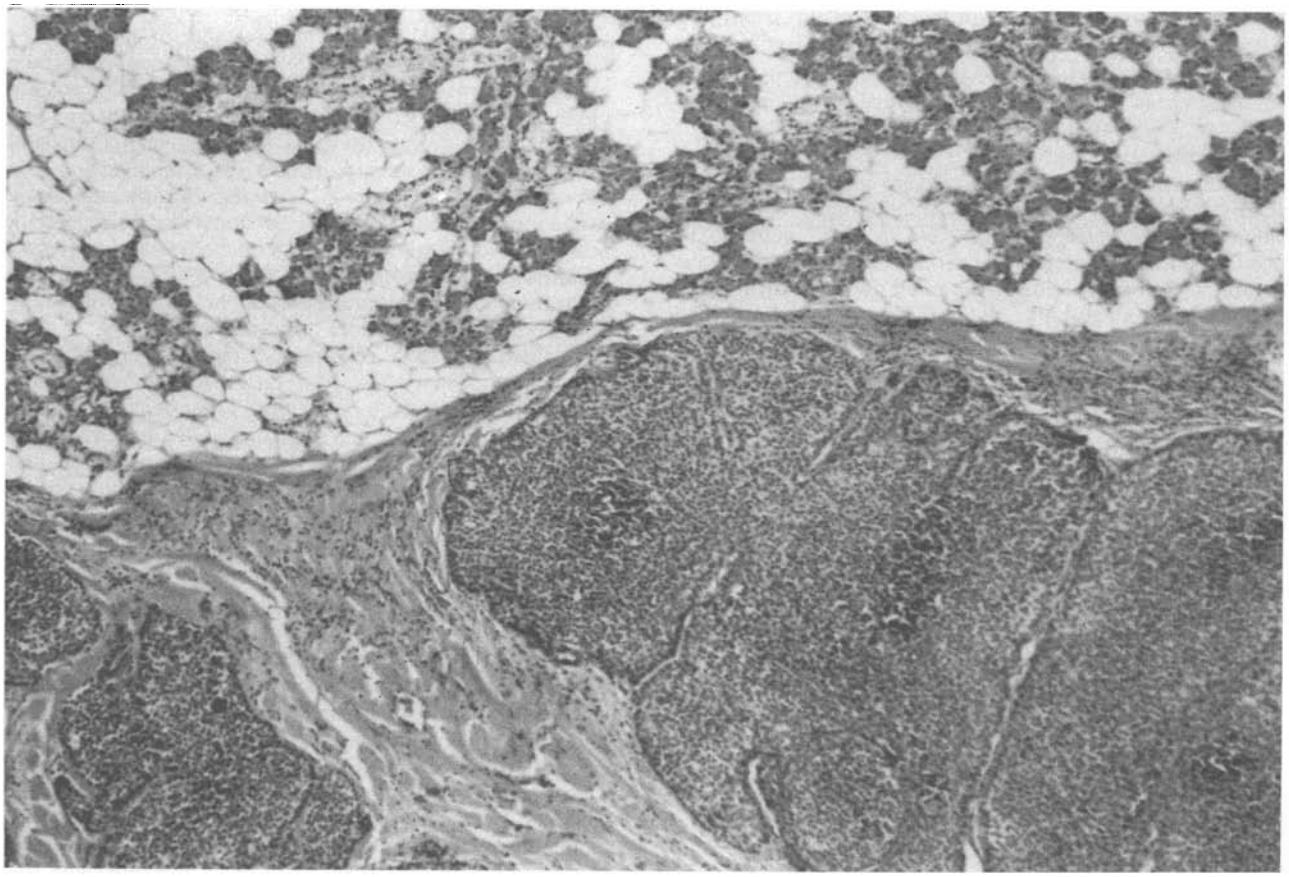

FIG. 3
Membranous basal cell adenoma of the parotid gland with remarkable resemblance to the dermal eccrine cylindroma of the scalp (H & E; × 12.5).

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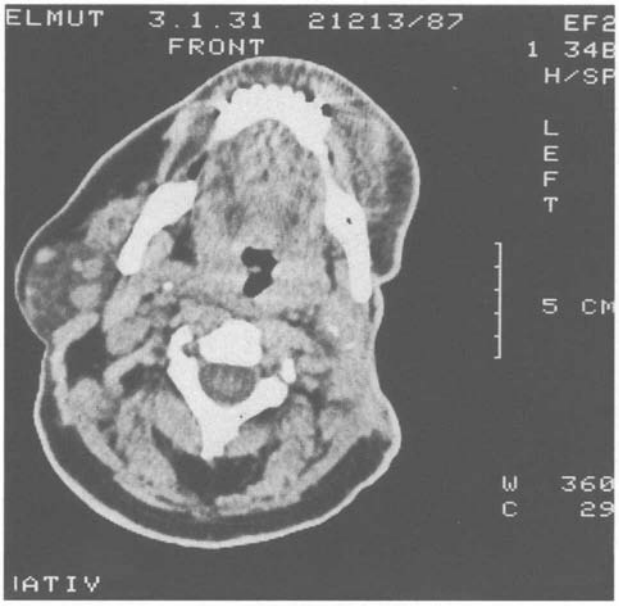

FIG. 4
Native transversal CT scan in the same patient; note the high density, well-defined subcutaneous mass in front of the right ear; Excisional biopsy revealed dermal eccrine cylindroma.

In 1997, the patient reconsulted our department with multiple painless whitish nodules emerging from the scalp (diameters 0.5 to 4 cm). The computed tomography (CT) scan of the remaining right parotid gland showed multiple small high-density lesions (Figure 4). The scalp tumours were excised and histological examination again showed multiple benign cylindromas. Fine needle aspiration cytology findings of the right parotid gland confirmed basal cell adenoma. Up to now, the patient suffers from no major ailments other than FADC.

Discussion

Basal cell adenomas of the salivary glands represent a relatively heterogenous group of adenomas with occasional tubular differentiation and a prominent PAS-positive basement membrane. Some basal cell adenomas exhibit secretory activities with small amounts of hyaline, PAS-positive intratubular material, that is sometimes also located between the cells of solid trabecular proliferations. Three histological features are essential: cords of basaloid cells, hyaline material and cystic and/or solid cell masses with ductline structures (Crain and Helwig, 1961). The arrangement of the tumour cells is categorized as the solid, trabecular, or membranous (dermal) analogue type. The term 'monomorphic adenoma' was originally proposed as a general category to encompass all varieties of salivary gland adenomas other than mixed tumours (pleomorphic adenomas). Ellis and Auclair point out that many published investigations of monomorphic adenomas have not adequately identified the various histological types within this heterogeneous group so that conclusions about the clinicopathological parameters of each type are not possible from these reports (Ellis and Auclair, 1996).

Headington *et al.* were the first to describe the notable histological similarity between dermal eccrine tumours (eccrine spiradenoma and cylindroma) and *membranous* basal cell adenoma (Headington *et al.*, 1977).

Whereas the recurrence rate for the other subtypes of basal cell adenomas is very low (Ellis and Wiscovitch, 1990), re-manifestation has been reported to be 25 per cent for the membranous subtype which is probably because of its tendency to be multifocal and unencapsulated (Batsakis *et al.*, 1991). Multinodularity and sometimes multifocal growth are also held responsible for frequent remanifestation after resection for the dermal counterparts, the dermal eccrine cylindromas (Batsakis *et al.*, 1981a). Malignant transformation of salivary gland basal cell adenomas seems to be rare, but has been reported (Hyman *et al.*, 1988). Batsakis *et al.* have cited a rate of malignant transformation of four per cent for the other types of basal cell adenomas, whereas it is considerably higher (28 per cent) for the membranous subtype (Batsakis *et al.*, 1991).

A combination of multiple dermal cylindromas and salivary gland membranous basal cell adenomas has been noted before in 11 reported cases (Table I). These cases share common traits with the membranous basal cell adenomas of salivary glands originally described by Kleinsasser *et al.* (1969).

Familial occurrence was pointed out in six of the reported 11 cases (see Table I) and in our patient. The male/female ratio of 11/1 differs from that in FADC without salivary gland affection, for which a female preponderance is known (MacKusick, 1992). This observation is consistent with that of Batsakis *et al.* (1991).

In our patient, during an observation period of more than 41 years, (since first excision of cutaneous tumours) our patient had recurrent dermal cylindromas without a tendency for malignant transformation. The extirpated salivary gland tumour of the left parotid gland showed no sign of malignancy, and until today, the membranous basal cell adenoma of the right parotid gland shows no signs of malignant transformation, such as loss of well-defined borders or abundant growth in ultrasound.

Mixed tumour, adenoid cystic carcinoma, and basal cell adenocarcinoma are different from the basal cell adenoma of the parotid gland. Batsakis points out however, that the lesion may easily be mistaken for an adenoid cystic carcinoma or basal cell carcinoma either in cutaneous or salivary sites, (Batsakis, 1989). Unlike adenoid cystic carcinomas, basal cell adenomas are composed of eosinophilic cells with smooth ovoid or round nuclei. The tumours lack cells with pale to clear cytoplasm and irregular angular-shaped nuclei. Morphologically, the cribriform pattern that is common in adenoid cystic carcinoma is rare in basal cell adenoma and is, if present, accompanied by more typical solid or trabecular growth patterns (Nagao *et al.*, 1982). Growth patterns such as infiltration and perineural invasion help distinguish adenoid cystic carcinoma from basal cell adenoma.

Distinction of basal cell adenocarcinoma from basal cell adenoma is primarily based upon growth features indicative of more aggressive behaviour. These features include infiltration of parotid parenchyma and adjacent tissues such as fat, muscle, skin and bone and perineural and vascular invasion (Ackerman, 1996).

Conclusion

The case reported here involved turban tumour syndrome with multiple membranous basal cell adenomas of both parotid glands. Surgical treatment of both the dermal cylindromas and the parotideal membranous basal cell adenomas led to cosmetically acceptable results, but failed to prevent local and even distant re-manifestation (right parotid gland) of new tumours of the same type. Treatment of the tumours is often only required for cosmetic purposes, however, any clinical or histological suspicion of malignancy warrants an excision biopsy and clinical follow-up (Burrows *et al.*, 1992). Histologically, the tumours may be confounded with adenoid cystic carcinoma or basal cell carcinoma. Such confusion would cause

radical and inappropriate treatment (radical parotidectomy with facial nerve resection), severe functional and cosmetic damage would be the result (Batsakis, 1989). To prevent confusion, close communication between surgeon and pathologist is mandatory, and the patient's clinical appearance (turban tumours) has to be mentioned. In general, excision of the dermal and parotideal lesions depends on the extent of the lesions but does not prevent recurrence (Crain and Helwig, 1961; Burrows *et al.*, 1992).

It is not yet clear whether FADC with membranous basal cell adenoma of the salivary glands is a rare subset of FADC or (as indicated by differences in male to female ratio) a distinct syndrome; ongoing genetic analyses may help to elucidate this question.

References

Ackerman, A. B. (1996) Eccrine Cylindroma. In *Ackerman's Surgical Pathology/1*. (Rosai, J., ed.), Mosby, St Louis, pp 120–121.

Alawi, M. H., Hobby, J., Lesna, M. (1982) Familial dermal cylindroma with involvement of the parotid gland. *British Journal of Plastic Surgery* **35:** 167–170.

Batsakis, J. G. (1989) Dermal eccrine cylindroma. *Annals of Otology, Rhinology and Laryngology* **98:** 991–992.

Batsakis, J. G., Brannon, R., Sciubba, J. J. (1981a) Monomorphic adenomas of major salivary glands: a histologic study of 96 tumours. *Clinical Otolaryngology* **6:** 129–143.

Batsakis, J. G., Brannon, R. B. (1981b) Dermal analogue tumours of major salivary glands. *Journal of Laryngology and Otology* **95:** 155–164.

Batsakis, J. G., Luna, M. A., el-Naggar, A. K. (1991) Basaloid monomorphic adenomas. *Annals of Otology, Rhinology and Laryngology* **100:** 687–690.

Biggs, P. J., Wooster, R., Ford, D., Chapman, P., Mangion, J., Quirk, Y., Easton, D. F., Burn, J., Stratton, M. R. (1995) Familial cylindromatosis (turban tumour syndrome) gene localized to chromosome 16q12-q13: evidence for its role as a tumour suppressor gene. *Nature Genetics* **11:** 441–443.

Burrows, N. P., Jones, R., Smith, N. P. (1992) The clinicopathological features of familial cylindromas and trichoepitheliomas (Brooke-Spiegler syndrome): a report of two families. *Clinical and Experimental Dermatology* **17:** 332–336.

Crain, R. C., Helwig, E. B. (1961) Dermal cylindroma (dermal eccrine cylindroma). *American Journal of Clinical Pathology* **35:** 505–515.

Ellis, G. L., Auclair, P. L. (1996) Basal cell adenoma. In *Tumours of the Salivary Glands*. American Registry of Pathology, Washington, DC, pp 80–150.

Ellis, G. L., Wiscovitch, J. G. (1990) Basal cell adenocarcinomas of the major salivary glands. *Oral Surgery, Oral Medicine and Oral Pathology* **69:** 461–469.

Ferrandiz, C., Campo, E., Baumann, E. (1985) Dermal cylindromas (turban tumour) and eccrine spiradenomas in a patient with membranous basal cell adenoma of the parotid gland. *Journal of Cutaneous Pathology* **12:** 72–79.

Hammond, D. C., Grant, K., Simpson, W. D. (1990) Malignant degeneration of dermal cylindroma. *Annals of Plastic Surgery* **24:** 176–178.

Headington, J. T., Batsakis, J., Beals, T., Campbell, T., Simmons, J., Stone, W. D. (1977) Membranous basal cell adenoma of parotid gland, dermal cylindromas, and trichoepitheliomas. Comparative histochemistry and ultrastructure. *Cancer* **39:** 2460–2469.

Herbst, E. W., Utz, W. (1984) Multifocal dermal-type basal cell adenomas of parotid glands with co-existing dermal cylindromas. *Virchows Archive-A Pathological Anatomy* **403:** 95–102.

Hyman, B. A., Scheithauer, B., Weiland, L., Irons, G. B. (1988) Membranous basal cell adenoma of the parotid gland. Malignant transformation in a patient with multiple dermal cylindromas. *Archives of Pathology and Laboratory Medicine* **112:** 209–211.

Kleinsasser, O., Hübner, T., Klein, H. J. (1969) [Histogenesis of cylindromas]. *Archiv der klinischen und experimentellen Ohren-, Nasen-, und Kehlkofheilkunde-* **195:** 192–206.

MacKusick, V. A. (1992) *Mendelian Inheritance in Man: Catalogs of Autosomal Dominant, Autosomal Recessive, and X-linked Phenotypes*. The Johns Hopkins University Press, Baltimore, pp 289–289.

Nagao, K., Matsuzaki, O., Saiga, H. (1982) Histopathologic studies of basal cell adenoma of the parotid gland. *Cancer* **50:** 736–745.

Pingitore, R., Campani, D. (1984) Salivary gland involvement in a case of dermal eccrine cylindroma of the scalp (turban tumour). Report of a case with lung metastases. *Tumori* **70:** 385–388.

Reingold, I. M., Keasbey, L., Graham, J. H. (1977) Multicentric dermal-type cylindromas of the parotid glands in a patient with florid turban tumour. *Cancer* **40:** 1702–1710.

Rockerbie, N., Solomon, A., Woo, T., Beals, T., Ellis, C. N. (1989) Malignant dermal cylindroma in a patient with multiple dermal cylindromas, trichoepitheliomas, and bilateral dermal analogue tumours of the parotid gland. *American Journal of Dermatopathology* **11:** 353–359.

Schuermann, H., Weber, K. (1937) Beitrag zur Kenntnis der Spieglerschen Tumoren (Cylindrome) nebst einigen Bemerkungen zum Epithelioma adenoides cysticum. *Archiv für Dermatologie und Syphilis* **175:** 682–695.

Urbanski, S. J., From, L., Abarmowicz, A., Joaquin, A., Luk, S. C. (1985) Metamorphosis of dermal cylindroma: possible relation to malignant transformation. Case report of cutaneous cylindroma with direct intracranial invasion. *Journal of the American Academy of Dermatology* **12:** 188–195.

Vernon, H. J., Olsen, E., Vollmer, R. T. (1988) Autosomal dominant multiple cylindromas associated with solitary lung cylindroma. *Journal of the American Academy of Dermatology* **19:** 397–400.

Zarbo, R. J., Ricci, A., Kowalczyk, P. D., Cartun, R. W., Knibbs, D. R. (1985) Intranasal dermal analogue tumour (membranous basal cell adenoma). Ultrastructure and immunohistochemistry. *Archives of Otolaryngology – Head and Neck Surgery* **111:** 333–337.

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