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Microcystic adnexal carcinoma

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Abstract
Microcystic adnexal carcinoma (MAC) is a rare adnexal tumour which has only recently been recognized as a separate clinicopathological entity. It typically affects the face of the middle-aged and often requires extensive surgical excision, due to its locally invasive nature. Its clinical significance is that, despite being locally invasive, MAC is typified by a lack of metastatic spread. We present a case and review of the literature.

Key words: Carcinoma; Skin appendage diseases; Sweat gland neoplasms

Case report

A 55-year-old woman initially presented to her beautician with a lump in the nasal septum. This was painless and had only recently been noticed. In the past she had been successfully treated for breast carcinoma. There was no previous history of head and neck irradiation. Physical examination revealed a large cartilaginous swelling at the antero-inferior aspect of the nasal septum, extending into the sub-labial region. Clinically there were no signs of regional lymphadenopathy or distant metastasis. The lesion was excised from the anterior end of the septum, via a right-sided lateral rhinotomy and Killians' incision. Complete excision was not deemed feasible, as the lesion macroscopically had no defined margin. The patient made an uneventful post-operative recovery. A histological diagnosis of MAC was made. In view of the positive resection margins, the patient received a course of radical post-operative radiotherapy (tumour dose of 55 Gray in 20 fractions over four weeks). Six months post-operatively there is no evidence of local recurrence.

Pathology

Macroscopically, an irregular-surfaced nodule, measuring 22 × 12 × 9 mm was obtained. Microscopically, the sections contained skin with underlying cartilage, connective tissue and voluntary muscle. Within the dermis of the skin, there was a tumour composed of small keratocysts and narrow trabeculae of tumour cells (Figure 1), which were moderately pleomorphic with prominent nucleoli (Figure 2). Many of the tumour cells, especially in the more superficial parts of the tumour, showed ductular differentiation. The trabecular portion of the tumour infiltrated deeply into the muscle, with evidence of perineural spread (Figure 3). Trabeculae of tumour cells involved the margins of resection. This appearance was consistent with a microcystic adnexal carcinoma. Morphologically the tumour was unrelated to the patient's previous breast carcinoma.

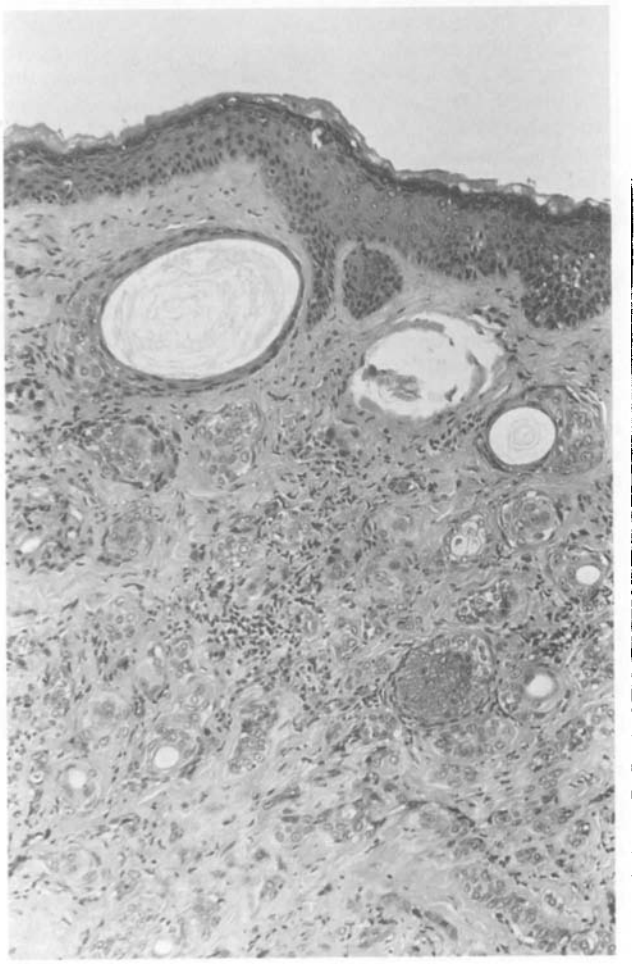

FIG. 1
Small keratocysts and narrow trabeculae of tumour cells within the dermis. (H & E; × 150).

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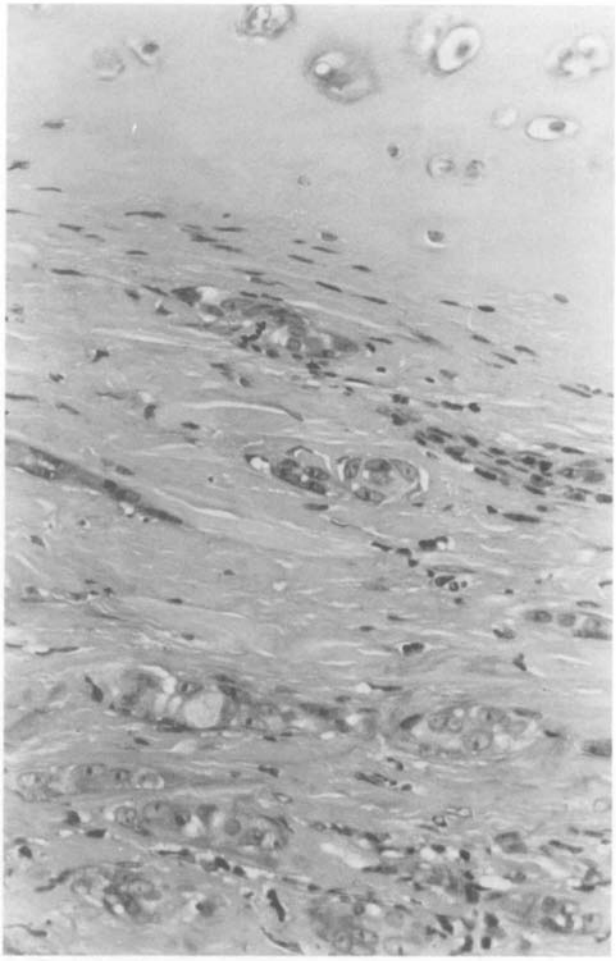

FIG. 2
Deeply infiltrative tumour cells with prominent nucleoli. (H & E; × 375).

Discussion

MAC was first described in 1982 (Goldstein *et al.*, 1982). It has several synonyms:- sclerosing sweat duct carcinoma, malignant syringoma, clear cell eccrine carcinoma and syringoid carcinoma. Despite its 'benign' clinical appearance, it has a tendency for extensive local invasion and local recurrence. In the past it has been confused with both benign and malignant adnexal tumours.

Typically it occurs in the mid-facial region of middle-aged patients. It usually presents as a slow-growing firm plaque or nodule and rarely ulcerates. Pain is occasionally a feature due to perineural invasion.

It is important to realize that despite its propensity to local invasion and local recurrence, to date there have been no reports of regional or systemic metastasis. It has been reported that previous irradiation may be a predisposing factor (Wallace and Bernstein, 1991).

The main histological findings are: 1) numerous superficial keratin-filled cysts; 2) dense fibrous stroma; 3) deep dermal invasion by narrow strands of tumour cells showing rudimentary tubular differentiation; 4) invasion of other local structures by tumour; 5) minimal cellular atypia and mitotic activity.

MAC is thought to derive from a pluripotential adnexal keratinocyte. Immunoperoxidase staining for carcinoembryonic antigen (CEA) supports a dual differentiation towards pilar and eccrine adnexal structures (Nickoloff *et al.*, 1986). Immunohistochemical and electron microscopic studies in the future will probably differentiate between the two.

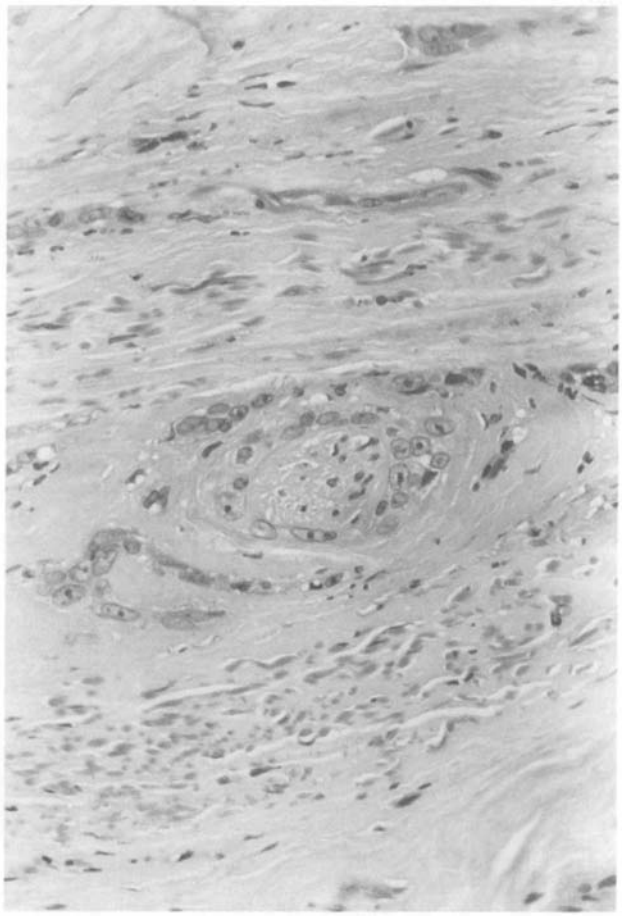

FIG. 3
Peri-neural infiltration by tumour (H & E; × 375).

Several of these cases in the literature have been treated by Mohs controlled surgical excision (Fleischmann *et al.*, 1984). This technique achieves clear resection margins with maximal preservation of normal tissue. The role of radiotherapy in this condition has not been extensively published. However, certain histological criteria should encourage its use post-operatively. These include dermal lymphatic invasion, perineural involvement, deep structure infiltration and positive resection margins (Harari *et al.*, 1990).

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