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Extraocular Sebaceous Carcinoma

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A 79-year-old man with no history of previous irradiation presented with a large ulcerated tumour of the cheek. The histological features favoured a true sebaceous carcinoma. Neither squamous nor basal differentiation were seen. Sudan stainings were positive. Tumour cells expressed suprabasal keratins and were negative for carcinoembryonic antigen, vimentin and S 100 protein. Extraocular sebaceous carcinomas occurring without previous irradiation are rare tumours which behave aggressively. Treatment regimens are discussed.

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Extraocular sebaceous carcinomas are rare neoplasms with an aggressive behaviour. We report a case.

CASE REPORT

A 79-year-old man with no history of previous irradiation presented with an ulcerated tumour of the left cheek, 4 cm in diameter (Fig. 1). He had had the lesion for 5 months. The regional lymph nodes were not enlarged. A wide surgical excision was performed. Histologically, the tumour was well circumscribed, non-encapsulated and deeply involved the subcutaneous tissue. It consisted of variably sized lobules of pleiomorphic cells with atypical hyperchromatic nuclei and foamy vacuolated eosinophilic cytoplasm (Fig. 2). There were numerous mitoses. Focal necrosis occurred amid some lobules, leading to cyst formation. No squamous or basal cell differentiation was seen.

In frozen-tissue sections, Sudan red staining revealed numerous cytoplasmic lipid droplets. Tumoural cells expressed suprabasal keratins as defined with KL1 monoclonal antibody (Fig. 3) and were negative for carcinoembryonic antigen, vimentin and S 100 protein. There was recurrence 7 months later. Metastatic lesions similar histologically to the primary tumour were detected in three regional lymph nodes. Abdominal CT-scan, chest X-ray and bone scintigraphy proved normal. A new wide surgical excision and lymph node dissection were performed without adjuvant chemotherapy. No recurrence was noted after an 8-month follow-up.

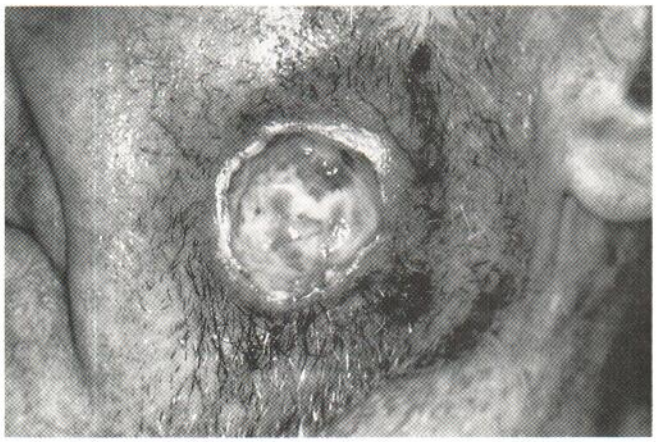

Fig. 1. Clinical appearance of the sebaceous carcinoma.

DISCUSSION

Pure sebaceous carcinomas are unusual (1). Tumours of the sebaceous gland are usually separated into three main categories: sebaceous adenoma, basal cell carcinoma with sebaceous differentiation, and true sebaceous carcinoma (2). Confusion in the nomenclature has arisen from the many terms used to define epidermal neoplasms having a sebaceous component (sebaceous epithelioma, metatypical mixed sebaceous epithelioma, spinosebaceous carcinoma). Troy & Ackerman proposed to call sebaceoma a benign neoplasm of adnexal epithelium with sebaceous differentiation (3).

The criteria used to characterize a sebaceous differentiation are poorly defined. The presence of cytoplasmic vacuoles with positive staining for lipids (Sudan IV technique) remains the most reliable histologic feature for defining sebaceous tumours (4).

True sebaceous carcinoma appears as a slow growing, yellowish nodule. The commonest site of occurrence is the ocular adnexa. Extraocular cutaneous locations are rare and concern especially the head and neck (5). Only 2 cases have been described on the external genitalia (4). The average age at diagnosis is 65. The male/female ratio is 2:1. Sebaceous carcinoma carries a risk of spread to regional lymph nodes, bones and viscera (6). Histologically it shows large asymmetrical, poorly circumscribed lobules composed of atypical basaloid and sebaceous cells with severe nuclear atypia, abundant eosinophilic cytoplasm and foci of squamous differentiation. The tumour invades the adjacent stroma and the deep dermal tissues. Mitosis may be frequent (1, 4, 6). In our case, tumour cells expressed high molecular weight keratins (Fig. 3) but

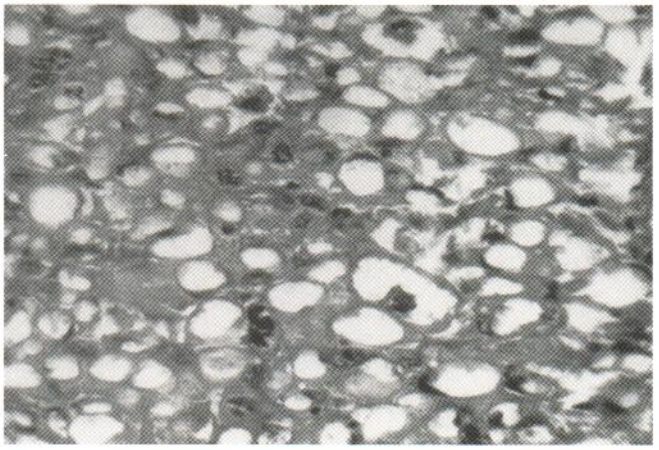

Fig. 2. Well-differentiated sebaceous carcinoma with large and small cytoplasmic vacuoles (H & E, ×250).

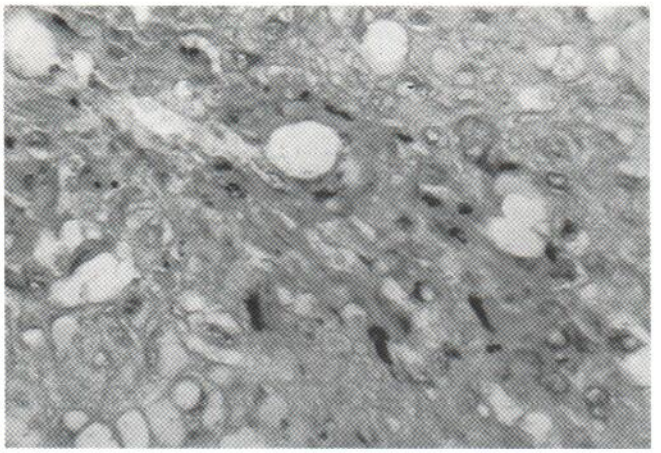

Fig. 3. KL1 monoclonal antibody staining of human tumour cells (peroxidase-anti-peroxidase technique, ×250).

were negative for carcinoembryonic antigen, vimentin and S 100 protein. In one case, stains for keratins have been reported to be negative (5).

Beside basal cell carcinoma with sebaceous differentiation and basal cell carcinoma associated with the naevus sebaceus of Jadassohn (which share the presence of cystic cavities) all neoplasms containing clear cells which have to be distinguished from sebaceous carcinomas. They include poorly differentiated squamous cell carcinoma, metastatic renal cell carcinoma, clear cell sweat gland carcinoma, clear cell melanoma.

The risk of developing sebaceous carcinoma are unknown. However, the occurrence of several sebaceous carcinomas in areas of chronic radiodermatitis suggests a close association (7). In our case, no previous irradiation had been recorded. Wide surgical excision with microscopically tumour-free margins and elective lymph node dissection are the treatment of choice for extraocular sebaceous carcinomas (4). The small number of reported cases does not allow us to further define optimum treatment regimens. The benefit of prophylactic lymph node dissection and the sensitivity to cytotoxic drugs or irradiation remain to be defined (4).

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