

Painful Nodule on the Scalp: A Case of Primary Cutaneous Adenoid Cystic Carcinoma

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Sir,

Primary cutaneous adenoid cystic carcinoma (PCACC) is a particularly rare variant of sweat gland carcinoma which was first described by Boggio (1) in 1975. More than 40 cases have since been reported (2). The natural history of PCACC is that of an indolent but progressive tumour with a high risk of local recurrence after excision. Most of the reported tumours had been present for ≥ 5 years before diagnosis (3). Approximately 40% arise in the scalp (4). Herein we report a case of PCACC presenting with a painful mass on the scalp.

CASE REPORT

A 55-year-old healthy woman was seen in our dermatology clinic complaining of a slowly enlarging plaque on her scalp that had been present for approximately 1 year. It had recently become painful.

Examination revealed a firm, mobile, blue-violet-coloured, smooth surfaced, tender, well-defined, 1.5–2 cm nodule without hair loss on the right side of the occipital region. The nodule was excised with the tentative diagnosis of dermatofibroma or basal cell carcinoma. The patient's medical and family history were non-contributory. She did not complain of fever, chills, dyspnoea, weight loss or bone pain.

Histology of the excised tumor revealed solid epithelial sheets in the reticular dermis and areas of cribriform islands consisting of basaloid cells with rather uniform round spaces containing myxoid material giving a punched-out appearance. Tumour cells had a uniform character and mitosis was rare (Fig. 1). These findings were consistent with a diagnosis of adenoid cystic carcinoma. Perineural invasion was noted in some of the sections. There was no palpable lymphadenopathy or organomegaly. The ultrasonography of the head and neck showed no mass in

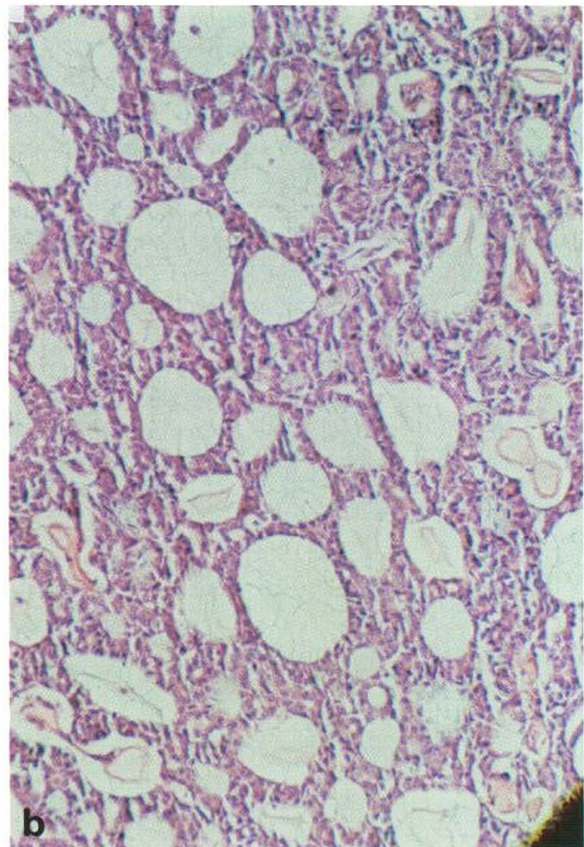


Fig. 1. (a) Cribriform islands forming a nodular structure in deep dermis. H&E $\times 35$. (b) High power view of cribriform islands with uniform round spaces containing myxoid material. H&E $\times 165$.

parotid glands or lymph nodes. The chest X-ray was normal.

A wide local re-excision was performed. At the time of writing the patient is 10 months postoperatively and there is no evidence of recurrence or metastasis.

DISCUSSION

The scalp is a unique anatomical site that can be involved with a variety of malignant and premalignant conditions. Although most lesions seen by the dermatologist will be benign, the evaluation of head masses involves an orderly approach based on a careful history and accurate physical examination. Certain lesional characteristics, such as pain, a hard consistency, immobility or a large size might need further evaluation using appropriate laboratory and radiological investigations (5).

PCACC appears to affect men and women equally, and the scalp seems to be the site of predilection, as seen in our case (6). Early diagnosis is important for preventing lymph node or lung metastases (3, 7). It is reasonable to rule out a direct cutaneous spread or a cutaneous metastasis of an adenoid cystic carcinoma of the salivary glands before a diagnosis of PCACC is made.

In our case, pain was the alarming symptom that led us to excise the whole mass in a 55-year-old woman who presented with a slowly enlarging nodule on the scalp. Perineural invasion, which is present in at least half of the patients, is responsible for pain in PCACC, as it was in this case. Local recurrence is noted in 57% of patients (4) and the presence of perineural invasion seems to increase the likelihood of recurrence. The reported interval between surgery and recurrence has ranged from 4 months to 20 years, necessitating long-term follow-up (6, 8). Because of its tendency to recur, wide local excision has been recommended as the treatment of choice for PCACC (6, 9).

Although PCACC is a rare tumor, we think that its frequency might have been underestimated especially because of its uncertain clinical presentation. There have been several reports of tumours clinically

diagnosed as benign appendageal tumor, cylindroma or basal cell carcinoma which after histopathological examination turned out to be PCACC (3, 7, 10, 11). Unlike its uncertain clinical presentation, the histopathological picture of PCACC is distinctive, with its characteristic cribriform pattern which originates from the tumour cells capable of producing basal lamina (6).

One should consider the diagnosis of PCACC when confronted with a painful nodule on the head of an elderly person.

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