Giant Naevoid Syringocystadenoma Papilliferum

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Sir,

Syringocystadenoma papilliferum (SCAP) is a rare tumour first described in 1893 (1). To date, 400 cases have been reported worldwide. SCAP usually appears at birth or in early childhood, most frequently on the face or scalp. In approximately 25% of cases it is observed on other locations such as the trunk or the extremities. In 40% of cases, SCAP evolves contiguous to a pre-existing naevus sebaceous, but cases without an antecedent naevus sebaceous have also been described (2). In 10% of cases SCAP was associated with naevus sebaceous and a basal cell carcinoma. SCAP typically consists of one papule to several papules in a linear arrangement or alternatively as a solitary plaque. Macroscopically, the mature lesion consists of clusters of generally pinkish brown nodules 2–10 mm in diameter with an occasional central opening. During puberty, SCAP may increase in size and develop a crusted and papillomatous texture (3). Here, we report the case of a giant naevoid SCAP that did not develop malignancy despite a long disease course and an absence of medical treatment.

CASE REPORT

A 43-year-old man was referred to our department in January 2004, with a giant cutaneous tumour located on the left side of the ventral thorax. The patient had several linear verrucous papules on the left side of the breast, present since birth. The patient initially presented at the age of 13 with a 14-cm organoid naevus on the left side of the thorax that displayed histological features consistent with SCAP (Fig. 1A). Between 1973 and 1977, he underwent three subtotal surgical excisions of the SCAP. Following these procedures he did not seek the advice of a physician again until his presentation at our clinic. The patient reported that during this time the tumour had slowly, but consistently increased in size. In January 2004, the patient finally sought treatment because of local pruritus and bleeding from the tumour. On examination, several scars were present on the left side of the thorax consistent with the previous surgical revisions. In the same area, a giant, 4×10 cm brownish, verrucous tumour was found (Fig. 1B). The tumour surface was serous and crusted and had a putrid fetor. Two sharply demarcated 1×1 cm erythematous papules with a smooth surface were located in close proximity to the tumour. There was no palpable regional lymphadenopathy and all routine laboratory tests were unremarkable. The tumour and the papules were excised under local anaesthesia and the resulting tissue defect was corrected by an autologous full-thickness graft following the granulation phase.

Histopathologically, the tumour was characterized by a number of cystic invaginations extending downwards from a papillomatous epidermis with a distinct verrucous hyperplasia. Numerous papillary projections extended into the lumen of these invaginations. Papillary projections were lined by a two-layer epithelium comprising an inner cylindrical and an outer cuboidal layer. The basement membrane remained intact. An inflammatory infiltrate consisting of lymphocytes, histiocytes and plasma cells was present in the stroma of papillary projections. The histological findings were consistent with a verrucous form of SCAP (Fig. 2A–C).

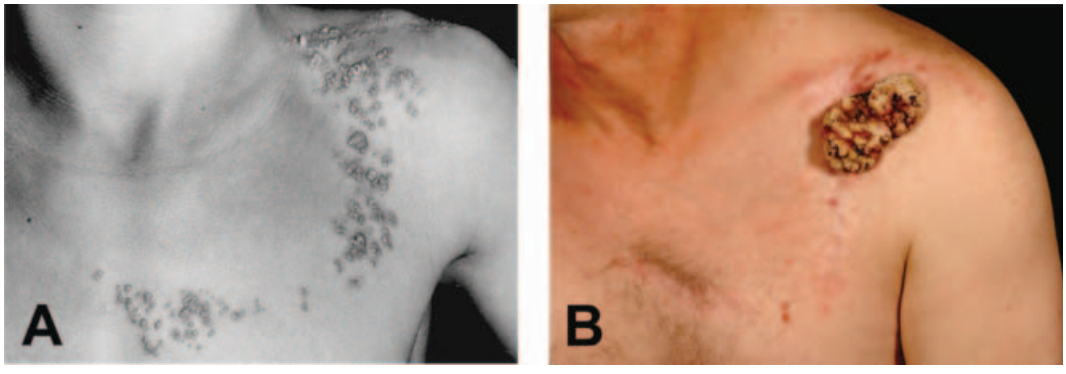

Fig. 1. (A) Typical features of the papulous form of syringocystadenoma papilliferum: linear papules associated with a naevoid malformation located at the left side of the thorax (1977). (B) 26 years later: 4×10 cm brownish, verrucous tumour on the thorax.

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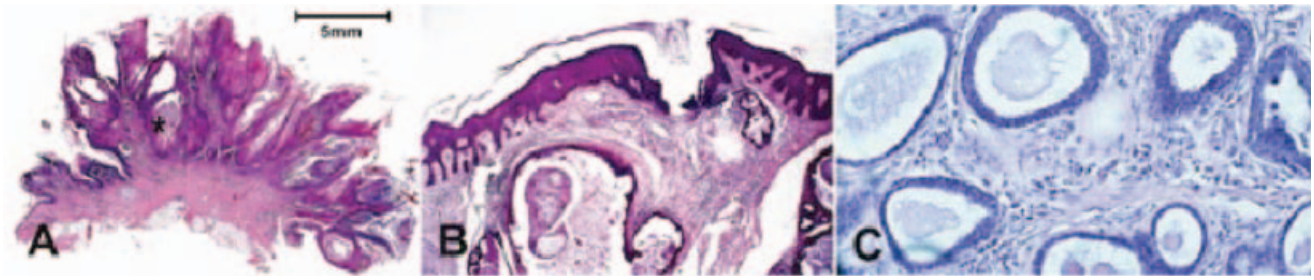

Fig. 2. (A) Overview: upper part of syringocystadenoma papilliferum (SCAP). Numerous cystic invaginations with a verrucous surface (H&E stain; original size). (B) Histopathology of the SCAP. Cystic invaginations extend downwards from a papillomatous epidermis. Cyst-like ducts with keratin (H&E stain; $\times 50$). (C) Papillary processes lined by a two-layered epithelium with decapitations on the luminal surface of the cells. The inflammatory infiltrate consists of lymphocytes, histiocytes and plasma cells. Apocrine glands are obvious beside the typical ducts of SCAP (H&E stain; $\times 100$).

DISCUSSION

SCAP is a rare cutaneous tumour generally thought to arise from apocrine sweat glands. Despite evidence of an apocrine origin, the histogenesis of SCAP remains unclear (3). Our case fulfils the typical criteria including the onset of the disease in early childhood and an association with an organoid naevus (4). Linear and plaque varieties of SCAP are more commonly found in the scalp, face and neck and only 20% of lesions are located on the trunk (5). On the trunk, nodular growth appears to predominate. The lesions generally become more elevated and verrucous during puberty. All published cases of nodular SCAP have to date measured <4 cm in diameter (2). A characteristic feature of the present case is the giant size of the tumour that, to our knowledge, has never been previously reported.

Due to the dimensions of the tumour, its long history and recent onset of bleeding from areas of the tumour itself, malignant transformation of SCAP into syringocystadenocarcinoma papilliferum (SCACP) had to be ruled out (6). To date only six cases of SCAP transforming to SCACP have been described (7). Published cases of malignant transformation of SCAP have described intervals of between 20 and 50 years from the time the lesions were first noted (7). Verrucous squamous cell carcinoma also needed to be excluded as it shares similar morphology to SCAP at this stage of development.

On histological examination, duct-like structures and cystic spaces with protruding papillary projections containing a plasma cell-rich stroma were found and were consistent with SCAP. The high degree of differentiation of the tumour cells and the intact basal membrane in the tumour allowed us to rule out both squamous cell carcinoma and SCACP. In cases of SCAP it is important for treating physicians to consider both the possibility of malignant transformation and the difficulty in distinguishing SCAP from other clinically similar lesions. Therefore we recommend early excision followed by histological analysis.

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