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Linear synngocystadenoma papilliferum of the limb: a rare localization of an uncommon tumour☆

Dear Editor,

Syringocystadenoma Papilliferum (SCAP) is a benign adnexal neoplasm that most frequently arises from an organoid nevus on the head and neck. It usually occurs during childhood or adolescence, varying in morphological character from smooth and flat to verrucous form. Most reported cases in the literature are single lesions presenting as a solitary raised warty plaque, and less commonly multiple papules. Here we report a case of multiple SCAP with a warty surface presenting on the limb distributed along a Blaschko line and without pre-existing lesions in an adult.

A 45-year-old female presented with several pink nodules on the left upper limb for seven years. The lesions were pruriginous and prone to bleed after scratching. Physical examination revealed multiple, verrucous papules, measuring 1 to 2.5 cm in the left upper extremity following a line of Blaschko. Central umbilication was seen in several lesions (Fig. 1A, B). She was misdiagnosed at another hospital with verruca vulgaris, laser was used to remove some of the lesions but soon recurred. One of the lesions was surgically excised and histopathology was performed. Features of SCAP were identified, with the tumor located in the superficial layer of the dermis without connection to the overlying epidermis, composed of cystadenoma-like structures and folded papillary structures. The cystic spaces and papillary structures were lined with single columnar epithelium and surrounded by a layer of small cuboidal myoepithelial cells, forming a special double-layer structure (Fig. 1C, D). DNA tested for Human Papillomavirus (HPV) was negative. After excision, there was no recurrence or new lesions at the 3-month and 6-month follow-ups. As the patient did not want to excise the other papules, we arranged for a subsequent visit after 6 months.

SCAP was first described by Stokes in 1917. The pathogenesis of SCAP remains unclear, HPV DNA and mutations in the RAS/mitogen-activated protein kinase signaling pathway have been detected.[1,2] In our case we failed to identify HPV infection though the lesions show verrucous growths. SCAP frequently arises in puberty within organoid nevi in the head and neck region. As far as we have observed, there have been 17 previous cases of linear SCAP reported in the literature in English, and only two of these cases developed in adults, aged 21 and 34 respectively. The reported 17 cases include 10 females and 7 males, 6 cases occurred in the head and neck, 5 cases were in the trunk, 5 cases were in the extremities and one case was in the inguinal fold. In the extremities, 3 were found on the leg and 2 on the upper limb.[3–5] The unique features of our case are Blaschkolinear distribution, the localization on the upper limb, late-onset in an adult, and the tumor without connection to the overlying epidermis. So far, neither organoid naevus nor epidermal naevus has been demonstrated in the linear form of SCAP. Therefore, multiple linear SCAP may represent a distinct clinical form, the relationship of linear SCAP with organoid naevus or other adnexal tumors needs further investigation.

☆ Study conducted at the Chengdu Second People's Hospital, Chengdu, Sichuan, China.

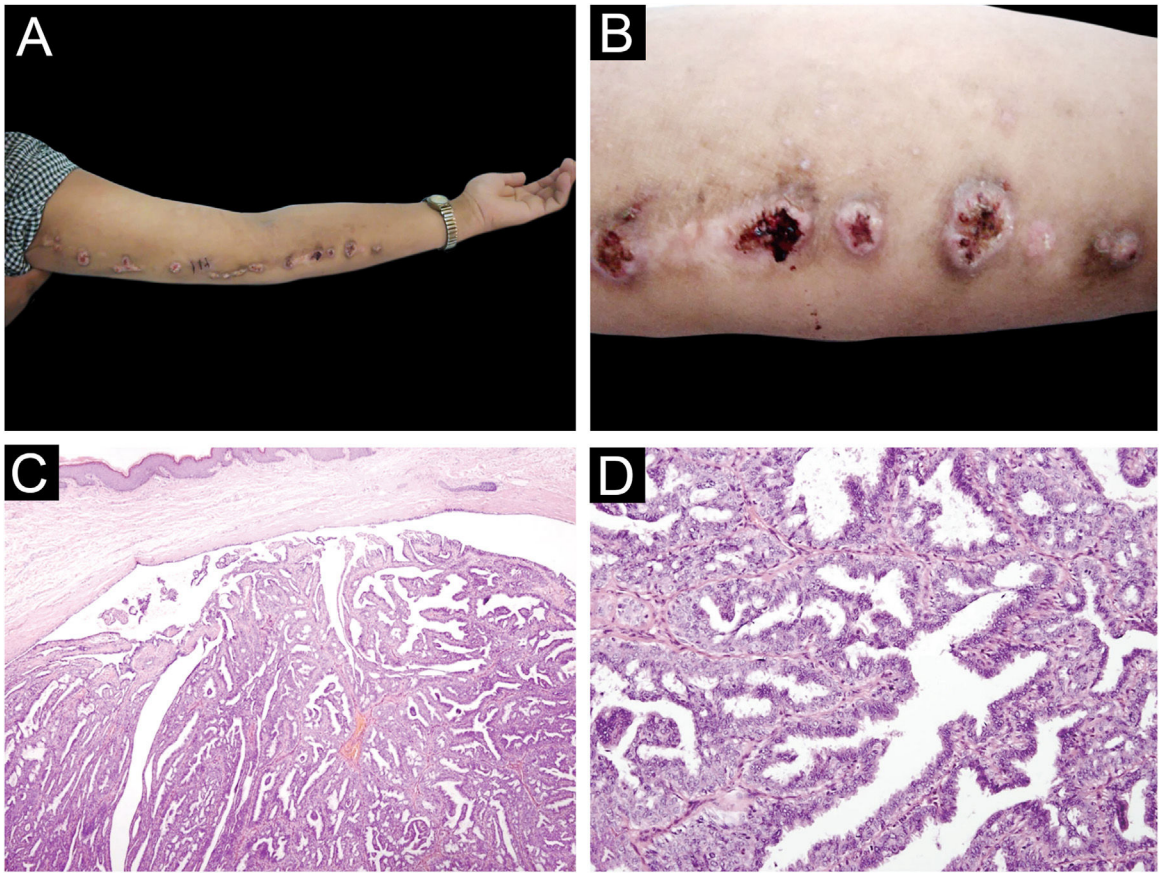

Figure 1 (A, B) Several normochromic papules, some with a warty surface, with a diameter of about 1 ∼ 2.5 cm on the left upper limb, arranged in a linear pattern. (C) The tumor is located in the superficial dermis without connection to the overlying epidermis, composed of cystadenoma-like structures and folded papillary structures. (Hematoxylin & eosin, ×40). (D) The cystic spaces and papillary structures are lined with single columnar epithelium and surrounded by a layer of small cuboidal myoepithelial cells, forming a special double-layer structure. (Hematoxylin & eosin, ×400)

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Author's contributions

Ming Yao: Pathological analysis, article writing and figure editing; literature search; approval of the final version of the manuscript; data collection, analysis and interpretation; critical review of the manuscript.

Lang Rao: Pathological analysis, article writing and figure editing; concept determination; approval of the final version of the manuscript; data collection, analysis and interpretation; critical review of the manuscript.

Conflicts of interest

None declared.

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Merkel cell carcinoma in a 38-year-old man: a case report☆

Dear Editor,

Merkel Cell Carcinoma (MCC) is a rare but aggressive cutaneous cancer, and it occurs mostly in older Caucasians, especially in immunocompromised patients. It is reported that up to 80% of MCC is associated with Merkel Cell Polyomavirus (MCPyV) infection, and 20% is related to ultraviolet.[1] The clinical presentation of MCC is nonspecific and varied, but most commonly presents with rapidly growing, solitary violaceous nodules with or without ulceration. Approximately 26%–36% of MCC patients have lymph node involvement and 6%–16% present with distant metastasis at their initial visit.[2] Surgery and radiotherapy are first-line treatments, while an emerging effective treatment modality is Immune Checkpoint Inhibitor (ICI).[3]

Herein, we report an extremely rare case of MCC in a young man with rapid deterioration to provide experience for the diagnosis and treatment.

Case report

A 38-year-old man with a five-year history of unspecified lesion on the left index finger presented to our clinic for an asymptomatic nodule at the same site. In the beginning, he presented with an eczematous lesion on the left index finger in 2015 and just accepted ointment treatment. Then an erythematous nodule appeared and was removed by surgery in 2016 without pathological examination. A growing red-purple painless nodule measuring 52×51×39 mm appeared at the same site four years later, which seemed to be covered with small, widened vessels (Fig. 1). Moreover, physical examination showed dark red nodules with scaly scabs on his right thenar and right ankle. And he reported no trauma to these lesions and no systemic symptoms such as fever and weight loss. He was a dentist, lived in urban areas without an epidemic and denied family medical history and long-term administration of drugs. Moreover, HIV infection was ruled out.

Wide excision of left index finger neoplasm and axillary lymph node dissection was performed in July 2020. The results of Hematoxylin-eosin staining and immunohistochemistry were in line with MCC histopathologic characteristics (Fig. 2). Besides, the pathological diagnosis for the other two lesions was Squamous Cell Carcinoma (SCC) and surgery was performed. The patient then accepted adjuvant therapy (Etoposide, Cisplatin, Pembrolizumab), routine blood tests, and renal and liver function were carefully monitored (Fig. 3). The disease maintained stable for four months. However, the liver was invaded by MCC in December 2020 (Fig. 4A), which suggested the adjuvant treatment was no longer effective. Given that he was still young, we offered a local radiotherapy (48Gy/16F) for hepatic metastases and tyrosine kinase inhibitor Apatinib was administered with the patient's consent. In February 2021, he presented with severe clinical worsening, and multiple enlarged cervical lymph nodes were observed (Fig. 4B). The patient eventually died 7 days after this admission.

The present report refers to an MCC patient under the age of 40 with poor therapeutic effect. Owing to its rarity, almost MCC treatment recommendations are derived from retrospective studies, and we take several potential factors that account for his poor immunotherapy response.

The first one to consider is age. Paulson[4] reported a higher fraction of metastases and a more aggressive course in younger MCC patients. MCC frequently was misdiagnosed due to non-specific clinical characterization and the patient mentioned above was treated as eczema for five years, which prevented him from timely treatment. Besides, there was a study indicating that neuroendocrine carcinomas associated with SCC had a higher incidence of local recurrence.[5] In our report, it is also noteworthy that the patient repeatedly appears lesions in the same skin site. But it is difficult to identify their connection because no pathological examination was carried out before this admission.

The Next Generation Sequencing results of peripheral blood showed no somatic variation within tumor genomes. What's more, this patient had a low tumor mutation burden (TMB) score (bTMB-L <1 Muts/Mb) as

☆ Study conducted at the Department of Radiation and Medical Oncology, Zhongnan Hospital of Wuhan University, Wuhan, China.