A woman in her 20’s presented with a right knee papule that had been present since childhood but grew larger and firmer over the past two years. (Figure 1). She denied any symptoms. Examination showed a pink, translucent, firm papule with telangiectasias on the right knee without palpable regional lymphadenopathy. A 5-mm punch biopsy was performed. (Figure 2). Histopathology revealed a cyst-like lesion below the epidermis comprised of clear fluid material. Both solid and cystic components were present alongside pleomorphic tumor cell aggregates characterized by mitotic figures, hyperchromasia, and necrosis. The combination of clinical presentation and the presence of ductal structures on histopathologic analysis led to a diagnosis of EPC. This case reinforces the critical importance of histopathologic analysis of a nonspecific lesion that has evolved over time and may resemble other forms of cutaneous malignancy.

EPC is a rare cutaneous adnexal tumor derived from eccrine sweat glands. Typically, EPC presents as a nonspecific erythematous papule or nodule most commonly on the head, neck, or lower extremities of females in the sixth or seventh decade of life. This tumor may arise from an existing benign eccrine poroma or occur de novo. Herein, we report a case of a growing erythematous nodule with overlying telangiectasia on the lower extremity of a woman in her twenties. Biopsy revealed a cyst-like lesion beneath the epidermis comprised of clear fluid material. Both solid and cystic components were present alongside pleomorphic tumor cell aggregates characterized by mitotic figures, hyperchromasia, and necrosis. The combination of clinical presentation and the presence of ductal structures on histopathologic analysis led to a diagnosis of EPC. This case reinforces the critical importance of histopathologic analysis of a nonspecific lesion that has evolved over time and may resemble other forms of cutaneous malignancy.

Ultrasound did not reveal abnormal lymph nodes. Three months later, local clinical lymphadenopathy was appreciated. Ultrasound-guided fine needle aspiration was performed, which was negative for carcinoma and favored a reactive process. The patient will continue serial monitoring long-term.

EPC is a rare cutaneous adnexal tumor derived from eccrine sweat glands. Some studies report that it arises from an existing eccrine poroma, while others suggest that it occurs de novo. EPCs most commonly occur in the sixth or seventh decade, though cases range from 6 months to 97 years. Lesions are nonspecific but can present as an erythematous papule with ulceration or crusting. On dermoscopy, EPCs contain linear irregular vessels surrounded by structureless white-pink halos.
Figure 1. Pink, translucent, firm papule on patient’s right inferior knee.

Figure 2. Original magnification x 200, hematoxylin and eosin staining. Histopathologic findings include a cyst-like lesion with solid components containing aggregates of pleomorphic tumor cells with ductal structures, mitotic figures, hyperchromatic nuclei, and cellular necrosis.

representing the mesh-like proliferation of involved tumor cells.\(^1\) Histopathology shows clonal proliferation of atypical cells with interspersed ductal differentiation. Lymph node metastases at diagnosis occur in approximately 30% of cases, discovered by clinical examination or by sentinel node biopsy.\(^2\) Currently, no guidelines exist regarding routine lymph node evaluation for EPC due to its rare nature.

The presence of a solitary firm, growing pink papule led to consideration of a broad differential diagnosis, including dermatofibroma, amelanotic melanoma, and nodular basal cell carcinoma (BCC). Dermatofibromas are benign dermal proliferations of fibroblasts commonly on the lower extremities. The most common variant, common fibrous histiocytoma, appears microscopically as an ill-defined dermal lesion of fibroblasts, macrophages, and blood vessels, with possible extension to
subcutaneous fat, often with interlacing fascicles of spindle cells within a loose collagenous stroma. Amelanotic melanoma, a rare melanoma subtype, most commonly affects the limbs of females over 50 years, and appears as an erythematous macule, skin-colored dermal plaque, or exophytic nodule lacking pigmentation. BCC, the most common skin cancer, often affects sun-exposed areas in individuals after the fifth decade of life. Nodular BCCs are shiny, smooth, pearly papules or nodules, with rolled borders and arborizing telangiectasias. Histopathology reveals discrete nests of malignant basaloid cells in the dermis, peripheral palisading, and mucoid stroma with plump spindle cells.

Management of EPC has historically included a combination of chemotherapy, radiation, and surgery. More recently, surgery has been the primary treatment, with adjuvant chemotherapy for metastases or recurrence. According to results from the National Cancer Database, WLE is the most common treatment approach, though other studies have suggested Mohs surgery as an efficacious and safe approach.

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