An 80-year-old male visited our outpatient clinic with the presence of a progressively enlarging eczematous plaque over the right neck close to the post-auricular area for 2 years. The skin lesion was unresponsive to cryotherapy and topical steroids. At this presentation, the physical examination revealed a ten dollar coin-sized, polygonal, salmon-red firm plaque with erosions and crusts, accompanied with a pinkish small nodule which was located in the center of the plaque (Fig. 1). Under the suspicion of chronic eczema or deep fungal infection, we performed a skin biopsy. The pathology revealed a malignant neoplasm of skin composed of pleomorphic basaloid and squamoid cells with frequent mitoses and apoptosis. The tumor was epidermal-based with dermal extension and anastomosing tumor cords showing multiple connections to the epidermis (Fig. 2A, 2B). Ductal differentiation is seen and can be accentuated under both epithelial membrane antigen (EMA) and carcinoembryonic antigen (CEA) stains (Fig. 2C, 2E). Taken together, an eccrine porocarcinoma (EP) was diagnosed. The patient received a wide local excision thereafter. The pathology reported eccrine porocarcinoma with Bowenoid features. Focal en mass necrosis (Fig. 2D), focally infiltrative border, ductal differentiation, and intracytoplasmic lumina. Squamous differentiation was also noted. There were frequent mitoses (>30 per 10 HPFs) and perineural invasion, without evidence of lymphovascular invasion. The mainly pushing advancing behavior and focal dyskeratosis were characteristic of Bowenoid pattern. The post-operative course was smooth and there was neither recurrence nor evidence of metastasis in eleven months of follow-up to date.

EP was first described by Mishima an Morioka in 1969 as a malignant counterpart of eccrine poroma, with both neoplasms putatively arising from the intraepidermal portion of the eccrine sweat duct or acrosyringium. Since then about 200 cases of EP have been reported in the literature, but there have been few large series. The tumor either arises spontaneously or develops in a long-standing eccrine poroma. Although often red and papular, the lesion can be flesh-colored or appear as a plaque, polypoid or verrucous lesion or as an ulcer, which sometimes bleed with minor trauma. Unlike benign eccrine poroma, which is often found on the palms and soles, most eccrine porocarcinomas arise on the lower extremity with the trunk and head also representing common sites, and the least common being on the palm and neck.

The histologic diagnosis of EP was predicated on the basis of an irregular tumor at least partly formed of characteristic poromatosous basaloid epithelial cells displaying ductal differentiation, and significant cytologic atypia. Most of them contained mature well-formed eccrine ducts having an eosino-
philic luminal cuticle, with the remaining tumors containing small ill-formed ducts and/or intracytoplasmic lumina. Histologically, EP is characterized by epidermotropism and pagetoid diffusion in the epidermis. It should be differentiated from squamous cell carcinoma (which usually shows more nuclear pleomorphism, intercellular bridge formation, keratinization, absence of ducts and is negative for CEA and other markers of glandular differentiation), sebaceous carcinoma (which is composed of clear cells with bubbly cytoplasm due to lipid vacuoles that can be highlighted with a fat stain and is negative for CEA) and proliferating trichilemmal tumour (which shows lobules of squamous cells and trichilemmal keratinization).

EP may have variable histological changes. The “bowenoid pattern” is identified as an invasive tumor having a broad pushing advancing edge with focal dyskeratosis and often-bizarre cytonuclear atypia reminiscent of Bowen’s disease. Shaw and his coworkers later reported that 8 of the 27 cases of EP in their series showed bowenoid histologic changes. Robson and his colleagues also investigated that bowenoid features were seen in 14% of their 69 cases of EP. In all cases identified in the literature, bowenoid features arose within the cluster of cuticular cells. It may be speculated that nuclear atypia in the cuticular cells may reflect the biologic behavior and show a greater tendency towards malignant transformation than poroid cells, and that there may be a pathologic continuum according to the degree of nuclear dysplasia.

Whether it was an EP with bowenoid features, EP coexisted with Bowen’s disease (BD) or EP arising from BD, remains to be debated. Both of them, although rarely, have been reported. The designation of “Bowen’s disease with invasive adnexal carcinoma (BD-CA)” was originally used by Kao in 1981. Among these, few cases of EP arising from BD, in which the two distinct neoplastic areas exhibited continuity both clinically and histologically, have been reported in the past. There were also few reports of EP coexisted with BD, of which both tumors were separated completely with sufficient distance to allow each lesion to be clearly identified and different in immunohistochemical stains. Whether the coexistence occurs only by change or indicating a progression from BD to EP, the correlation between these two remains to be debated.

Although EMA is a useful marker of some benign tumors from eccrine gland structure, Mariko found that EP with Bowenoid changes cannot be differentiated from BD by immunohistochemical staining with EMA. Moreover, the cytologic similarities between SCC in situ and EP with bowenoid features may be superficially striking. Nevertheless, the presence of ductal differentiation readily establishes the correct diagnosis. In addition, the EP with bowenoid features has a verrucose architecture rarely seen in BD. Furthermore, dyskeratotic cells are less prevalent in EP with bowenoid features in comparison with BD, which, when
invasive, typically shows keratinization.

In summary, we reported a rare case of EP with bowenoid features arising from the neck, which is the less common site for EP. The case reminds us that EP, although a rare neoplasm, should always be considered in the differential diagnosis of any erythematous plaque poorly responsive to treatments. Sometimes, accurate distinction between Bowen’s disease and EP with bowenoid features may be quite difficult, particularly in a small biopsy or in the absence of ducts lined by atypical cells. In that event, the specimen should be carefully reviewed, since the latter needs more aggressive treatment.

REFERENCES