of SCACP have been reported, of which 13 were located on the head and neck, three on the extremities, one each on the back and the chest, and three were located in the perineal areas.[1–17] Syringocystadenocarcinoma papilliferum is prevalent in older ages, ranging from 46 to 86 years, with a median age of 65 years. There is no gender predilection. Clinically it presents as a nodule, inflammatory plaque, or tumor. On physical examination, the lesions are usually flesh-colored to hyperpigmented exophytic nodules, which can persist maximally up to 30 years. The maximum dimension of the lesion varies from 1.5 cm to 13 cm. Of the reported 21 cases, seven were diagnosed as SCACP in situ, and 14 were diagnosed as invasive SCACP. Three of the 14 invasive SCACP cases also had coexistent in situ carcinoma elements.[15,16] Regional lymph node metastasis was observed in only two cases.[4,5]

Syringocystadenocarcinoma papilliferum shows all the cardinal histomorphologic features of syringocystadenoma papilliferum, which includes irregular papillomatous invaginations from the epidermis, papillae lined by cuboidal cells with decapitation secretion (evidence of apocrine differentiation), plasma cell rich fibrovascular cores, and interstitial mucin. Anisonucleosis of variable degree, partial to full thickness loss of nuclear polarity, and stratification are present in SCACP. Central dirty necrosis within the glandular or cyst lumen with nuclear debris and neutrophils, reminiscent of a colonic adenocarcinoma may be seen focally in SCACP. Pagetoid spread of tumor cells into overlying epidermis, squamous differentiation, and adenoid cystic-like areas may be observed.[7,10,14–16]

There is no specific immunohistochemical marker for SCACP. The areas with basaloid and squamous differentiation demonstrate CK5/6 and CK7 immunoreactivity. CK5/6, p63, and podoplanin (D2-40) expression seems to be valuable to distinguish cutaneous adnexal neoplasms from metastatic adenocarcinoma. Furthermore, the combined expression of p63 and D2-40 has been reported to be useful in distinguishing the primary skin tumor from metastatic carcinomas of the skin.[18–21] Some authors have suggested that the pattern of CK7 staining may be helpful in discriminating primary adnexal neoplasms from metastatic adenocarcinoma because primary lesions tend to have focal rather than diffuse staining pattern for CK7. Expression of CK7 in luminal surface further supports an adnexal origin.[18] CEA immunostain was positive in the luminal columnar cells, accentuated at the apical cell membrane. The immunohistochemical studies for CEA and CK7 may be useful, particularly in cases where diagnosis of SCACP is considered on histology, where the non-invasive component is predominantly of syringocystadenoma papilliferum and the invasive component shows only squamous differentiation.

Recognition of SCACP and its accurate distinction from various metastatic malignancies is very important as this differentiation has significant therapeutic and prognostic implication. While the majority of reported cases of SCACP do well with surgical excision only,[1] with rare