

BRIEF ARTICLES

Syringocystadenoma Papilliferum of the Scalp Arising De-Novo in an Elderly Female

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INTRODUCTION

Syringocystadenoma papilliferum (SCAP) is a rare benign hamartomatous tumor of adnexal origin stemming from either apocrine or eccrine glands. This tumor appears early in life with 50% of cases developing at birth and 15-30% of cases arising in puberty.¹ Presentation during adulthood is uncommon and more likely to represent malignant syringocystadenocarcinoma papilliferum (SCACP).²⁻³ SCAP arises in three forms: plaque, linear, and solitary nodular type. The plaque type appears as an alopecic patch on the scalp, often enlarging during puberty into a warty, nodular, or crusted plaque. The linear type presents as multiple red papules or nodules 1-10 mm in size. The nodular type presents as domed pedunculated nodules 5-10 mm in size, typically found on the trunk.⁴ We present a case of large plaque SCAP without malignant transformation arising in a 73-year-old adult woman and a concise description of our histopathologic findings.

CASE PRESENTATION

The patient was a 73-year-old woman who presented with a non-healing, alopecic,

oozing, bleeding lesion on the crown of her scalp for the past four years (Figure 1). Initially, the lesion was biopsied and diagnosed as erosive pustular dermatosis with a culture positive for *Pseudomonas* bacteria. Oral levofloxacin and clobetasol ointment were prescribed. One year later the lesion appeared as an eroded atrophied plaque with adherent yellow-green crust measuring 6 x 3.5 cm. Again, she was diagnosed with a bacterial infection of the skin. Three months later with no improvement after antibiotic treatment tacrolimus 0.3% ointment was started. Approximately one year later the patient saw a different dermatologist. A shave biopsy of the area was taken and diagnosed as actinic keratosis. The patient switched dermatologists again after six months. The lesion was now a 3 x 3 cm exophytic plaque with a 5.5 x 4 cm surrounding scar. An 8 mm punch biopsy was taken with a final diagnosis of syringocystadenoma papilliferum.

Histologic examination of the biopsy demonstrates loss of the surface epithelium (Figure 2a,b) with replacement by a downward growth of duct-like structures lined by numerous papillary projections. The base of the lesion is well defined with no

July 2020 Volume 4 Issue 4

evidence of an infiltrative architecture. Some papillary structures are lined by squamous type epithelium (Figure 3a,b) while in other areas the papillary structures are lined a double-layer of cuboidal and columnar type epithelium. Within the stroma of the papillary structures, there is a mixed inflammatory infiltrate composed of neutrophils, lymphocytes and plasma cells. There is no significant cytologic atypia or mitotic activity.

Figure 1. Syringocystadenoma papilliferum lesion of the scalp plaque type in a 73-year-old woman.



Figure 2. (A) The lesion elevates off the surface of the epidermis. The normal epidermis has been replaced by an irregular downward proliferation of papillary structures. The base of the lesion is well defined with no evidence of an infiltrative growth pattern (20x, H&E).

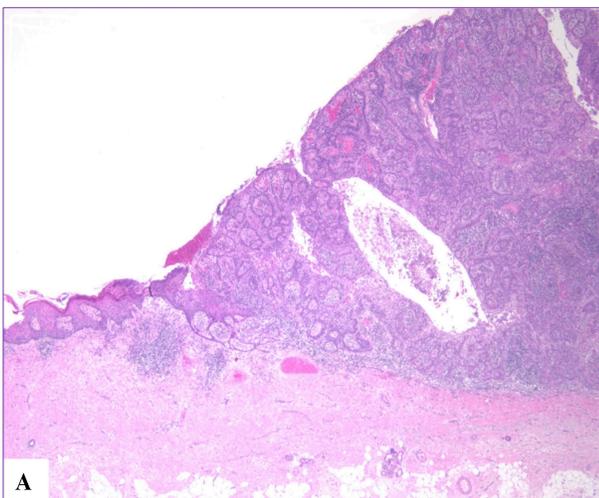


Figure 2. (B) Papillary structures that are lined by squamous epithelium in some areas and cuboidal type epithelium in others (100x, H&E)

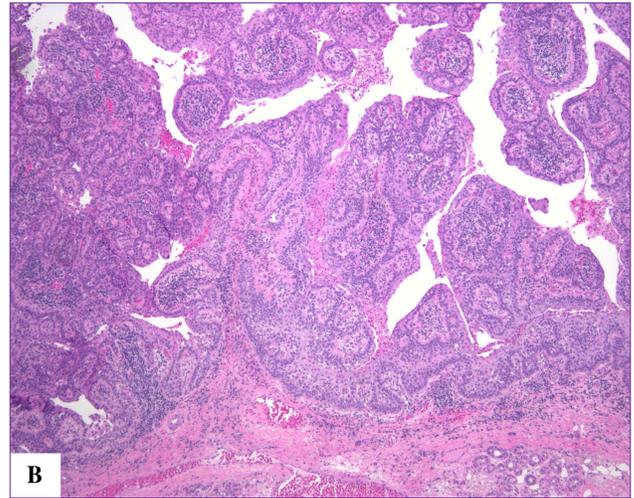
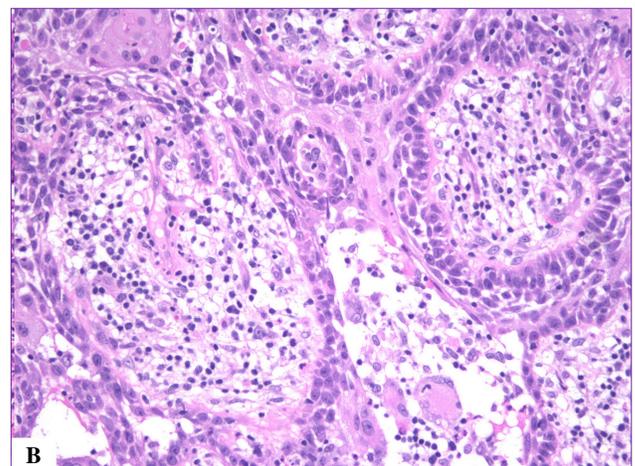
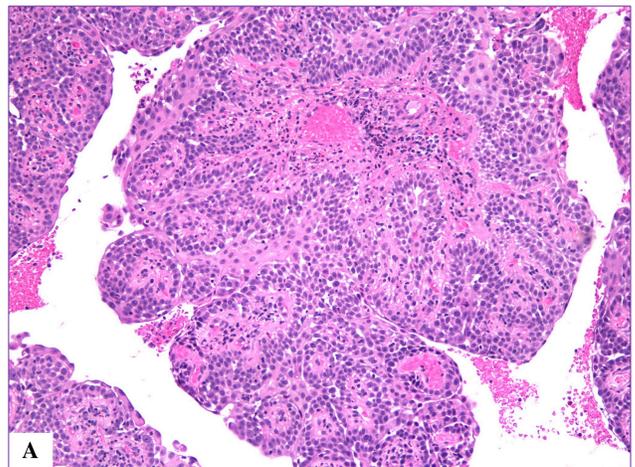


Figure 3. The papillary structures are lined by an inner layer of basaloid cells with some cytologic atypia. The outer layer that is typically lined by columnar cells has been replaced by squamous type epithelium (3A) 100x, H&E and (3B) 200x, H&E



DISCUSSION

The clinical picture of SCAP can be confusing due to variable clinical presentations. The lesion often appears as an exophytic, hairless, papillary structure combined with erosion or ulceration with various vessel morphologies. Dermatoscopy findings can be altered when SCAP arises from a preexisting lesion, nevus sebaceous being the most common. Biopsy with histopathology is necessary for confirmatory diagnosis.⁵

SCAP can be confused with a range of other cutaneous tumors including basal cell carcinoma (BCC), squamous cell carcinoma (SCC), cylindroma, and proliferating trichilemmal cysts. BCC and SCC have been reported to arise from SCAP; BCC has been reported in up to 10% of SCAP.^{6,7} Additionally, while both lesions can have a prolonged clinical timeline and ulcerate, they typically do not have a continuous relapsing and remitting course. They are also less likely to appear on a scalp as hair offers some protection from the sun. Histopathologically, SCC shows nests and cords of atypical epithelioid appearing cells extending from the epidermis into the underlying dermis. Depending on the degree of differentiation the individual cells may have abundant to minimal eosinophilic cytoplasm. Similarly to SCC, BCC also arises from the dermal-epidermal junction. Classic BCC will appear as nest and cords of basaloid appearing cells with areas of peripheral palisading and retraction artifact, with some nests at times associated with stromal mucin.⁸

Comparably, to SCAP, a cylindroma is a tumor of the eccrine and apocrine glands. These can present as single or multiple tumors arranged closely on the head or neck

giving rise to the colloquial name of “turban tumor” due to its appearance. While solitary tumors are not inherited in a familial fashion, multiple tumors are inherited in an autosomal dominant manner. These tumors are often smooth and firm with some hair loss as pedunculation occurs with tumor growth. Conversely to SCAP, cylindromas lack lymphoid cells and papillary structures and instead have a histologic appearance composed of dermal nests of basaloid cells arranged in a “jigsaw” pattern with a surrounding hyaline sheath and an abundance of dendritic cells. Regression does not occur, and plastic surgery is often necessary for removal.⁹ Lastly, proliferating trichilemmal cysts are benign freely mobile lesions often on the scalp of elderly females stemming from the keratinization of the epithelium encasing the hair follicle. Tenderness is common with trauma to the area. They can grow rapidly and ulcerate upon rare malignant degeneration. Histologically, epithelial cells line the cyst, without a granular layer, and increase in size as they grow closer to the cyst cavity containing keratin and cholesterol clefts.¹⁰ SCAP is a benign tumor that rarely undergoes malignant transformation. The conversion of SCAP to its malignant counterpart, syringocystadenocarcinoma papilliferum (SCACP), has been documented twice in the literature as of 2018.⁵ If rapid growth and ulceration occur malignancy should be considered.

Treatment options for SCAP are limited. Complete surgical excision with reconstruction, Mohs micrographic surgery, and CO2 laser excision used in areas unfavorable to surgery have all proven successful.^{1,7-8}

CONCLUSION

Benign SCAP can appear in adulthood and should not be excluded from the differential diagnosis based on age or lack of a pre-existing lesion. SCAP diagnosis requires a biopsy to differentiate from similar appearing lesions such as BCC, SCC, cylindromas, and proliferating trichilemmal cysts. Complete excision or reaching an adequate depth with a shave biopsy and appropriate histopathology is necessary to ensure the correct diagnosis as shallow biopsies can confuse the clinical picture and yield inaccurate pathology reports.

Conflict of Interest Disclosures: None

Funding: None

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