

## Case Presentation

### Agminated syringocystadenoma papilliferum: a new clinical presentation of a rare benign adnexal neoplasm

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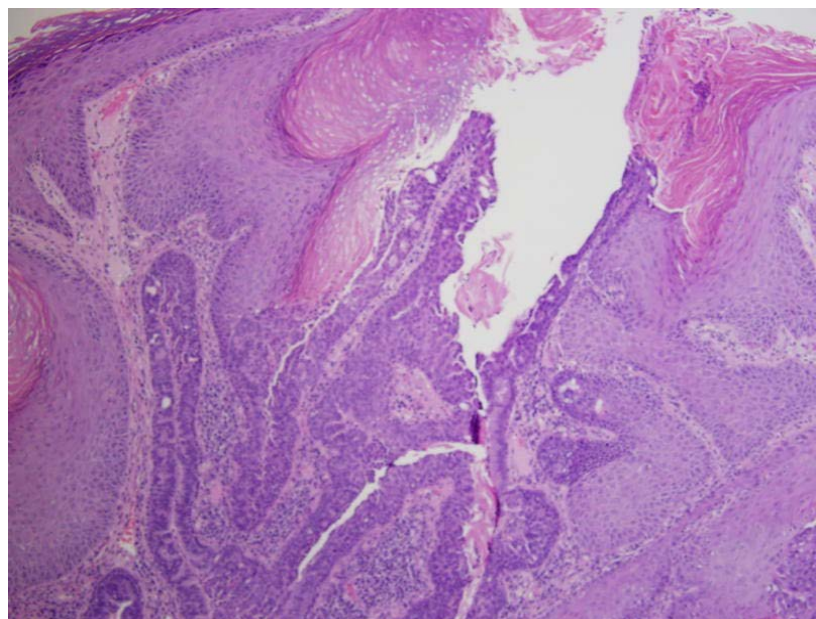
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## Abstract

Syringocystadenoma papilliferum is a rare adnexal tumor that often occurs as a solitary tumor in the head and neck region, although occurrences on other anatomical locations have been described. Linear configurations have been described, but an agminated form is a more rare and underreported variant of this tumor. We describe a case of a healthy 10-year old female with agminated syringocystadenoma papilliferum occurring on her left supraclavicular region, with the clinical appearance of grouped molluscum contagiosum papules. Case synopsis

A healthy 10-year-old girl was referred for the treatment of a "collection of molluscum contagiosum" of the left supraclavicular region of several years duration. The lesions were asymptomatic and refractory to cryotherapy. The patient was a healthy girl with no significant systemic findings. Cutaneous exam revealed a clustered group of pink, dome shaped, umbilicated papules over a 1.5 x 1 cm area within the left supraclavicular fossa (Figure 1a).

An excisional biopsy was performed. Routine H&E stained sections revealed cystic epidermal invaginations with papillary projections. The superficial portions of the cyst were lined by stratified keratinizing epithelium, whereas the deeper papillated portion exhibited a double layer of basal-like cells and luminal eosinophilic columnar cells with focal decapitation secretion. The papillary structures contained fibrovascular cores and lymphoplasmacytic infiltrates. A component of hamartomatous follicular growth was not identified (Figure 1b-d.). A diagnosis was made of agminated syringocystadenoma papilliferum.



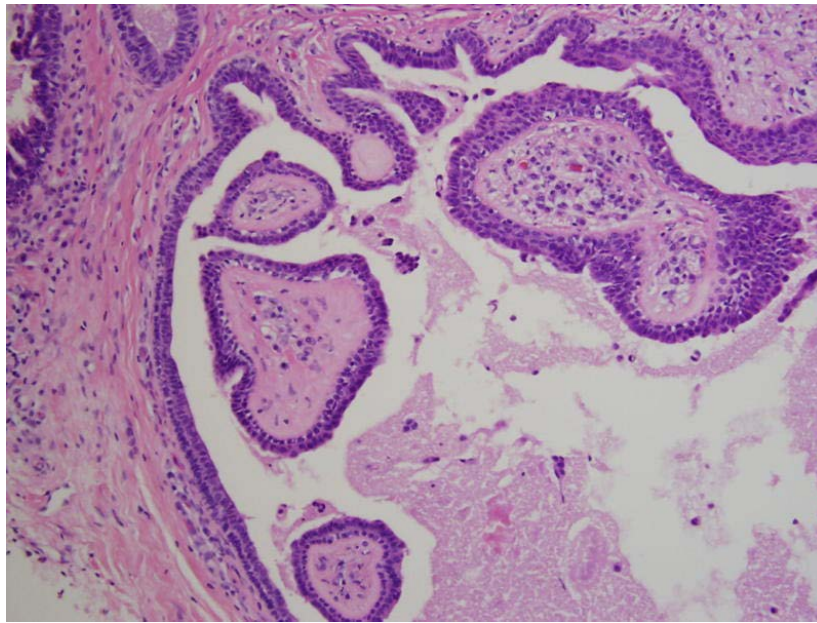


Figure 1a) Clinical photo of agminated pink umbilicated papules; b) Cystic invaginations within the dermis 10x; c) Deep cystic portions exhibited prominent papillary projections 20x; d) The papillae were lined by a double layer of basal-like cells and luminal eosinophilic columnar cells with focal decapitation secretion 40x.

## Discussion

Syringocystadenoma papilliferum is a rare benign adnexal tumor with variable clinical presentation. The vast majority of tumors arise within the head and neck region. However, other locations such as the arm, axilla, chest, breast, eyelid, scrotum, and leg have been reported [1-5]. About half of these tumors are present at birth with the remaining cases arising within early to late childhood. Lesions tend to enlarge during puberty and may become papillomatous and crusted. Approximately one-third of lesions arise in association with organoid nevi, such as sebaceous nevus, and 10% of lesions are associated with basal cell carcinomas.[1] Malignant variants have been reported [6].

The histogenesis of these tumors is highly disputed. The cells lining the papillary fronds exhibit decapitation secretion, suggesting apocrine differentiation. However, 90% of tumors arise in regions devoid of apocrine glands. Both apocrine and eccrine features have been documented by light microscopy, electron microscopy, and immunohistochemistry. Proposed theories of histogenesis include hamartomatous origin and hybrid apoeccrine differentiation, a well known occurrence in lesions of nevus sebaceous, a tumor commonly associated with syringocystadenoma papilliferum [7].

Clinically, *de novo* lesions tend to have a nonspecific and misleading presentation. Two different clinical distribution patterns have been reported, solitary plaques and linear papules [1, 8]. Ours appears to be the first case in which the entity presented clinically as agminated papules. Solitary plaques may be clinically misdiagnosed as viral warts or pyogenic granulomas. Linear papules may be misdiagnosed as herpes zoster or epidermal nevi. In our case, the agminated nature of the lesions suggested a clinical diagnosis of molluscum contagiosum. Within the literature, *de novo* linear tumors are more frequent in women and have been reported on the extremities, trunk, and neck [8, 9]. The phenomenon of an agminated syringocystadenoma papilliferum appears to be unique. Agminated is an adjective meaning aggregated or clustered and has been described in other cutaneous neoplasms including acquired nevi, Spitz nevi, lentigines, blue nevi, trichodiscomas, dermatofibromas, capillary hemangiomas, and angiofibromas. The exact mechanism by which a clonal benign proliferation becomes agminated is unclear. In this case the lesion could be excised in its entirety.

## References

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