

Linear Syringocystadenoma Papilliferum with Unusual Presentation: A Rare Case Report

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Abstract

Syringocystadenoma papilliferum (SP) is a skin hamartoma also known as nevus syringocystadenomatous papilliferus. The histogenesis of this tumor is unclear. It is believed to derive from undifferentiated pluripotential cells with either apocrine or eccrine differentiation and still classified under tumors with apocrine differentiation because of evidence of decapitation secretions pointing toward an apocrine origin. It is relatively a rare neoplasm, and most of the patients presented with solitary lesions in the head and neck region at birth or early childhood. Multiple erythematous papules like lesions are rarely a presentation and often misdiagnosed clinically. We report a case of SP in a 10-year-old female, which was located in axilla and was clinically misdiagnosed as molluscum contagiosum.

Key words: Molluscum contagiosum, Skin hamartoma, Syringocystadenoma papilliferum

INTRODUCTION

Syringocystadenoma papilliferum (SP) is a rare hamartomatous skin adnexal tumor which is believed to originate from undifferentiated pluripotential cells which may show apocrine or eccrine differentiation but still considered under the category of tumors with apocrine differentiation.¹ It is predominantly a childhood tumor and is present since birth in almost 50% of cases another 15-30% of them developing before puberty with equal frequency in both sexes. Mostly, it is seen on the head and neck region with face and scalp being the most common locations, however, these tumors may be seen in other locations like vulva, external ear, lower leg, scrotum eyelids and breast.^{1,2}

A present case report is of linear SP in 10-year-old female, which was located in axilla. Though clinically it was very much similar to that of molluscum contagiosum but could

not be considered as a differential diagnosis because of the history of long duration.

CASE REPORT

A 10-year-old female child was brought with a complaint of multiple asymptomatic papules over the right axillary region since birth which gradually increased in size. The lesion developed a change in texture and grew rapidly over the last 1 year. There were no associated systemic symptoms.

On examination, there were multiple erythematous papules of size 1-10 mm in linear pattern of arrangement. Most of the papules were discrete, pink, and dome shaped few of the papules were eroded with yellowish creamy slough on it. Central umbilication and crusting were also noted on some of the papules. There was no regional lymphadenopathy. Hematological, biochemical and radiological investigations were normal. Lesions were similar to that of molluscum contagiosum but because of long duration history no particular presumptive diagnosis was made, and skin biopsy was performed for the final diagnosis (Figure 1 and Table 1).

Pathological Findings

We received two punch biopsies of skin each measuring 5 mm.

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Microscopic Findings

Section showed epidermis and dermis. Epidermis revealed hyperkeratosis, parakeratosis acanthosis, and papillomatosis mimicking like a verrucous lesion (Figure 2). Adjacent to it was a focus of cystic, papillary and ductal invagination extending into deep dermis. These invaginations were lined by double layers of cells made of outer layer of cuboidal cells with round nuclei and scanty cytoplasm and a luminal layer of tall columnar cells with oval nuclei and eosinophilic cytoplasm (Figure 3). The core of the papillae was filled with dense plasma cell infiltrate, few lymphocytes and dilated capillaries (Figure 4).

Table 1: Distribution of sites and sub types of SP

Case report	Type	Site	Age group
Vyas <i>et al.</i> ²	Plaque	Scalp	12 year female
Chauhan <i>et al.</i> ¹	Linear	Right lower abdomen	10 year female
Park <i>et al.</i> ³	Nodular	Right supra pubic	65 year male
Present case report	Linear	Axilla	10 year female

SP: Syringocystadenoma papilliferum



Figure 1: Linear, multiple erythematous papules with crusting and umbilication on axilla

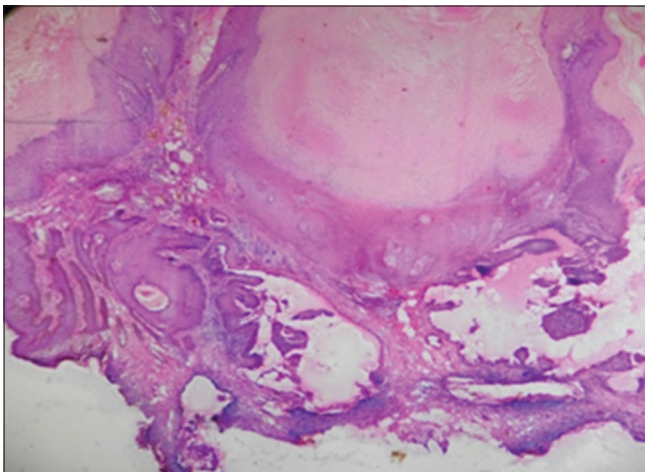


Figure 2: Epidermis with hyperkeratosis, parakeratosis and cystic invaginations with papillary projection into the lumen of cyst

Deep in dermis groups of tubular glands with large lumina were seen. The cells lining the lumina were flattened and showed evidence of active decapitation secretion.

On the basis of clinical features and histopathological finding diagnosis of linear SP was made.

DISCUSSION

In the present case report, the lesions were located in axilla in a linear pattern of arrangement, this particular location and the pattern of arrangement is highly uncommon and has not been described in available literature.

Three clinical subtypes of SP have been described:^{1,2,4}

- a. Plaque type: They present mostly as a hairless area in the scalp and enlarge during puberty to become nodular, verrucous or crusted. It is often seen in

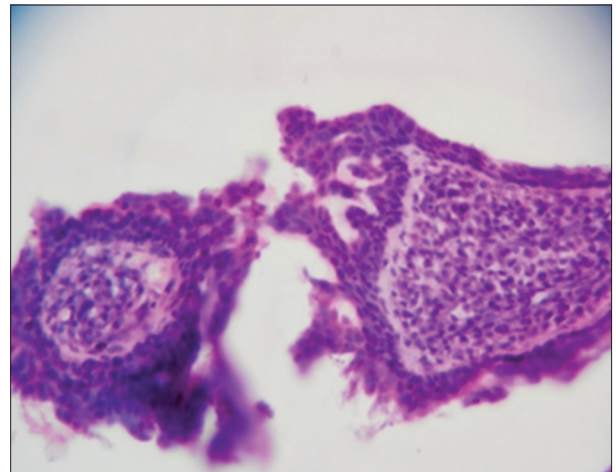


Figure 3: Papillary projections lined by two rows of epithelial cells. Luminal row is high columnar outer row consist of small cuboidal cells

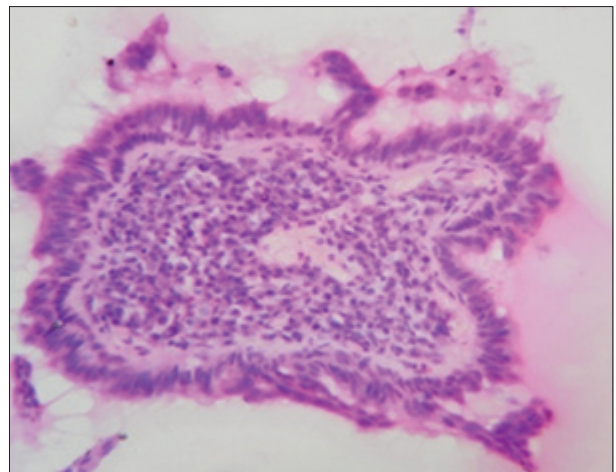


Figure 4: Plasma cells infiltrate in the core of papilla, decapitation secretion at the luminal surface

association with sebaceous nevus of Jadassohn or may appear *de novo* without preexisting lesions.

- b. Linear type: Nearly, all cases involve neck and rarely chest arm abdomen and thigh. The lesions are present since birth and consist of multiple linear reddish pink firm papulovesicles with dome-shaped surface of size 1-5 mm, as age progresses it may increase in size and number sometimes show central umbilication, ulceration, crusting, and yellowish foul smelling creamy discharge. At puberty, it may become verrucous and papillomatous.
- c. Solitary nodular type: This type show predilection for the trunk, shoulders, axillae, and the genital area and occasionally extensive verrucous or papillary plaques can be seen. It consists of solitary pedunculated nodules up to 5-10 mm.

A long list of lesions which are reported to be associated with SP includes viral warts, nevus sebaceous, linear nevus verrucous, nevus comedonicus, apocrine poroma, apocrine hidradenoma, tubulopapillary hidradenoma, hidradenoma papilliferum, papillary eccrine adenoma, verrucous carcinoma, apocrine acrosyringial keratosis, poroma folliculare, linear nevus verrucosa, atypical fibro xanthoma, clear cell syringoma, basal cell epithelioma, sebaceous epithelioma, trichoepithelioma and verruca vulgaris.⁵⁻⁹ It can be seen in association with malignant tumors such as verrucous carcinoma, basal cell carcinoma, sebaceous carcinoma and ductal carcinoma. The most common association is with a nevus sebaceous.^{10,11}

Differential diagnosis is syringoma for solitary lesions,⁴ other adnexal tumors which show linear arrangements include nevus comedonicus, trichodiscoma, trichoepithelioma, basaloid follicular hamartoma, cylindroma, eccrine nevus, syringoma, eccrine poroma, eccrine spiradenoma, and basal cell carcinoma.¹ Umbilicated nodules may mimic molluscum contagiosum, warty lesions often confused with verruca vulgaris. Clinical features of SP vary widely

but histopathology is invariably uniform and confirmatory in all clinical subtypes.^{1,4}

Results of enzyme histochemistry immunohistochemistry and electron microscopy obtained have been conflicting.¹¹ A complete surgical excision is a treatment of choice.⁴

CONCLUSION

The presented case illustrates a yet un-described location and pattern of the rare skin tumor linear SP, leading to a clinical misdiagnosis. Correct diagnosis rest on histopathology since clinical information does not narrow down among many differential diagnoses.

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