Case Report

Syringocystadenoma papilliferum on the thigh: An unusual location

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ABSTRACT

A 28-year-old man presented with a partially eroded growth on the back of his right thigh for the last 15 years. Tuberculosis verrucosa cutis and pyogenic granuloma were suspected on clinical examination. Histopathological examination revealed syringocystadenoma papilliferum (SCAP). SCAP is rare on the thigh and a review of the English literature revealed eight cases. Non-descript clinical presentation in an unusual location and the rare linear and segmental variants lead to misdiagnosis. Onset at puberty should alert one to the possibility of the unusual location of SCAP.

Key words: Leg, syringocystadenoma papilliferum, thigh

INTRODUCTION

Syringocystadenoma papilliferum (SCAP) is a benign adnexal neoplasm occurring during childhood or adolescence. It usually presents as a papular lesion or a smooth hairless plaque on the scalp and forehead. Nodular or verrucous transformation is noted at puberty. The microscopic appearance is characteristic and shows ducts connecting to the surface, containing papillary processes and lined by two epithelial cell layers. Seventy-five percent of the cases are reported in the head and neck region.[1] The lower extremity is an uncommon site. We report a case of SCAP presenting on the thigh, clinically misdiagnosed due to the unusual location.

CASE REPORT

A 28-year-old man came with a partially eroded growth on the back of his right thigh of 15 years duration. He gave history of a localized eruption that started as a small papule on the back of the thigh and gradually grew in size. There was no history of any discharge or pain. It could be felt by the patient while sitting, for which he sought advice. Some local medications were applied but the lesion never subsided. Over the past 5 years, the lesion became more noticeable and became ulcerated, with pain and blood-tinged discharge that stained his clothes. At this point, he sought our advice. On examination, a soft verrucous nodule was seen on the middle of the posterior aspect of the right thigh. The lesion was a circumscribed nodule with a keratotic base surrounding it like a collar and eroded in the lower half, showing a red surface. The rest of the skin examination was normal and there was no regional lymph adenopathy. A working diagnosis of traumatized wart, verrucous tuberculosis and pyogenic granuloma was considered. Hemogram was normal. Mantoux test was 16 mm X 14 mm and an X-ray of the chest revealed no abnormality. The nodule was excised and sent for histopathology [Figure 1].

The histopathological section showed hyperplastic epidermis with cystic invaginations into the dermis. The epidermis showed prominent hyperkeratosis. The invaginations were lined by squamous epithelium near the surface, with transition to a double layer of cuboidal and columnar epithelium below. Multiple

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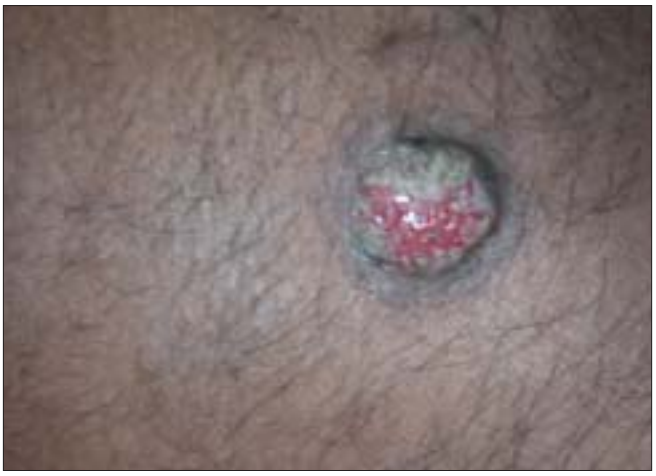

Figure 1: An eroded verrucous nodule on the right thigh

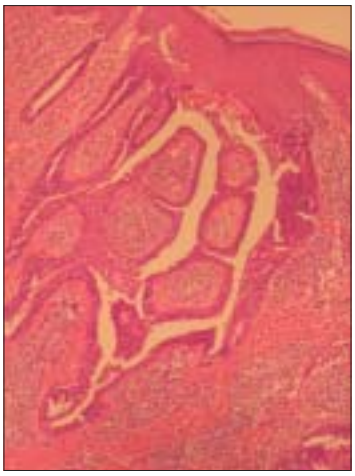

Figure 2: Cystic invagination of the epidermis with multiple papillary projections in the lumen (H and E, x100)

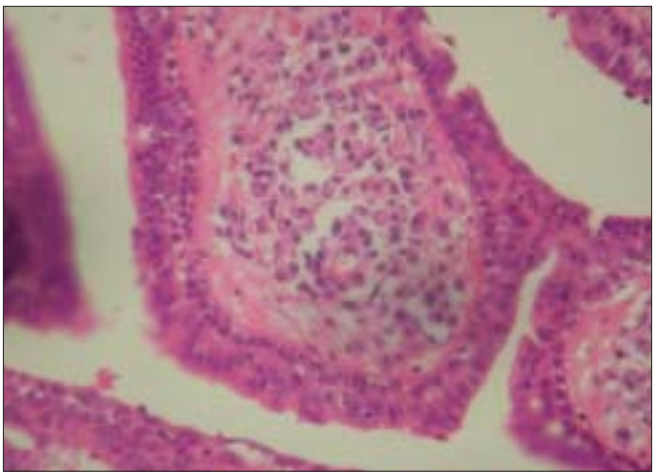

Figure 3: Papillae lined by bilayered epithelium with an abundance of plasma cells in the stroma (H and E, x400)

papillary projections were noted in the lumen of the cystic invagination. These were lined by bilayered epithelium composed of a luminal row of high columnar cells and an outer row of small cuboidal cells. A fair number of the columnar cells showed decapitation secretion. A dense inflammatory infiltrate composed of plasma cells and a few lymphocytes was noted within the papillae and in the underlying dermis [Figures 2 and 3]. There was no evidence of nevus sebaceous. A final diagnosis of SCAP was entertained.

DISCUSSION

SCAP occurs most commonly in the head and neck region, *de novo* or associated with nevus sebaceous. Uncommon sites of occurrence include chest, arms, breast, eyelids, axilla, scrotum, lower limb and inguinal and perineal regions.[2,3] Most of these are sporadic cases diagnosed on histopathology, clinical presentation being non-specific and misleading. Although an apocrine origin has been postulated, SCAP is rare in the axilla. Ninety percent of the cases are observed in anatomic sites normally devoid of apocrine elements.

Rare clinical patterns include linear and segmental variants.[4] This warrants inclusion of this tumor in the list of adnexal tumors forming linear arrangements. Segmental tumors tend to be misdiagnosed as herpes zoster, epidermal nevus and other lesions with a zonal distribution.

SCAP is rare on the thigh. A comprehensive review of the literature revealed eight cases,[1,5-7] which include multiple linear lesions[5] and one tumor arising in a giant comedo.[6] Yamamoto and Mamada[7] recently described a thigh lesion without connection to the overlying epidermis. The nodule was located in the middermis and the overlying epidermis was intact, without acanthosis. However, the authenticity of the report remains questionable as serial sections were not performed. Interestingly, the first case of SCAP was described on the thigh by Stokes in 1917. Other than the thigh, in the lower extremity, sporadic cases have been described on the lower leg[8] and toes.

The lesion increases in size at puberty, becoming papillomatous and crusted. One-third of all the cases arise in an organoid nevus, such as naevus sebaceous.[9] Coexisting basal cell carcinoma is noted in 10% of

the cases. Association with condyloma acuminatum has been described.[10] Syringocystadenocarcinoma papilliferum is the malignant counterpart, characterized by solid areas and cytologically malignant cells.[11]

The histogenesis of this lesion is disputed. The cells lining the lumina show evidence of decapitation secretion point to an apocrine origin. However, results of enzyme histochemistry, immunohistochemistry and electron microscopy have been conflicting. Hamartomatous origin from mature apocrine, eccrine or undifferentiated pleuripotential cells is suggested.[12] Helwig and Hackney[9] have suggested that the lesion represents an adenoma of eccrine duct origin or that it is derived from the ducts of a gland that is intermediate between eccrine and apocrine. The concept of apoeccrine gland has been proposed.[13] This exhibits microscopic, immunohistochemical and ultrastructural features of both eccrine and apocrine glands.

Morphologically, SCAP is characterized by endophytic invaginations of the epithelium into the dermis. These are duct-like structures leading into the dermal cystic spaces. Papillary projections of variable shape and size protrude into the lumen of these spaces. These are lined by double-layered outer cuboidal and luminal high columnar epithelium. Dilated capillaries and a dense infiltrate of plasma cells are noted in the stroma of these papillary projections. Tumor cells show a positive staining reaction with carcinoembryonic antigen and gross cystic disease fluid protein-15.

The thigh being an unusual site, SCAP was not considered in our case. Viral wart was considered based on the appearance of the lesion and the long clinical history. Based on the strong Mantoux reaction and the high incidence of tuberculosis in our country, warty tuberculosis was considered as a second possibility. Pyogenic granuloma was considered based on the soft exuberant appearance with an eroded surface. However, the absence of a pedicle and relatively scanty bleeding did not favor this diagnosis. Non-descript clinical presentation in an unusual location leads to misdiagnosis.[2]

The only treatment for SCAP is excision biopsy, which also confirms the diagnosis. CO_2 laser excision of SCAP of the head and neck is a clinical treatment option in anatomic areas unfavorable to excision and grafting.[14] Syringocystadenocarcinoma papilliferum has been successfully treated with Moh's micrographic surgery.[15]

To conclude, onset at puberty should alert one to the possibility of unusual location of SCAP. Solitary lesions in unusual locations generate multiple differential diagnoses and must be sent for histopathological examination.

REFERENCES

1. Mammino JJ, Vidmar DA. Syringocystadenoma papilliferum. Int J Dermatol 1991;30:763-6.
2. Patterson JW, Straka BF, Wick MR. Linear syringocystadenoma papilliferum of the thigh. J Am Acad Dermatol 2001;45:139-41.
3. Rao VA, Kamath GG, Kumar A. An unusual case of syringocystadenoma papilliferum on the eyelid. Indian J Ophthalmol 1996;44:168-9.
4. Premlatha S, Rao NR, Yesudian P, Razack A, Zahra A. Segmental syringocystadenoma papilliferum in an unusual location. Int J Dermatol 1985;24:520-1.
5. de Bliek JP, Starink TM. Multiple linear syringocystadenoma papilliferum. J Eur Acad Dermatol Venereol 1999;12:74-6.
6. Nizuma K. Syringocystadenoma papilliferum developed from giant comedo: A case report. Tokai J Exp Clin Med 1986;11:47-50.
7. Yamamoto T, Mamada A. Syringocystadenoma papilliferum arising on the thigh without connection to the overlying epidermis. Am J Dermatopathol 2008;30:84-5.
8. Yoshii N, Kanekura T, Setoyama M, Kanzaki T. Syringocystadenoma papilliferum: Report of the first case on the lower leg. J Dermatol 2004;31:939-42.
9. Helwig EB, Hackney VC. Syringocystadenoma papilliferum. Arch Dermatol 1955;71:361-72.
10. Skelton HG III, Smith KG, Young D, Lupton GP. Condyloma acuminatum associated with syringocystadenoma papilliferum. Am J Dermatopathol 1994;16:628-30.
11. Bondi R, Urso C. Syringocystadenocarcinoma papilliferum. Histopathology 1996;28:475-7.
12. Lever W: Histopathology of the skin. 4th ed. Philadelphia: JB Lippincott Co; 1967. p. 546-9.
13. Sato K, Leidal R, Sato F. Morphology and development of an apoeccrine sweat gland in human axillae. Am J Physiol 1987;252:166-80.
14. Jordan JA, Brown OE, Biavati MJ, Manning SC. Congenital syringocystadenoma papilliferum of the ear and neck treated with the CO2 laser. Int J Pediatr Otorhinolaryngol 1996;38:81-7.
15. Chi CC, Tsai RY, Wang SH. Syringocystadenocarcinoma papilliferum: Successfully treated with Mohs micrographic surgery. Dermatol Surg 2004;30:468-71.