Syringocystadenoma papilliferum of the lower limb with ulceration

Ano-Edward GH[1], Aramide KO[2], Amole IO[3], Adesina SA[3], Lasisi ME[1]and Jooda RK[1]

[1]Department of Histopathology, College of Health Sciences, Bowen University, Iwo, Nigeria
[2]Department of Histopathology, Babcock University, Ilsan Remo, Ogun State, Nigeria
[3]Department of Family Medicine, College of Health Sciences, Bowen University, Iwo, Nigeria

Abstract

Syringocystadenoma papilliferum is an exceedingly rare skin adnexal neoplasm of apocrine gland origin, commonly located primarily on the scalp and appearing as a hairless nodular plaque lesion. We report the case of a 35-year old man with history of an ulcer on the right leg which started three years earlier as a small excoriation after trauma to the leg over a lesion present from birth. Histological examination confirmed the diagnosis of syringocystadenoma papilliferum, which is a very rare lesion on the leg. We report this case due of its rarity and unusual clinical presenation.

Keywords: Syringocystadenoma papilliferum, Leg

Introduction

Syringocystadenoma papilliferum is a rare benign hamartomatous adnexal tumor, which originates from the apocrine or the eccrine sweat glands. It is relatively a rare neoplasm, predominantly a childhood tumor. About 50% of cases are present at birth and in 15-30%, the tumor develops before puberty[1]. Syringocystadenoma papilliferum occurs with equal frequency in both sexes [2].

In approximately one third of cases, it is associated with organoid nevus. [3] Seventy-five percent of the cases are reported in the head and neck region.[4]

Case Report

O.O is a 35- year old healthy non-smoking male that presented at the general out patients clinic of the hospital with a 3 years history of chronic leg ulcer on the right leg. The ulcer measures 6x4 cm on the lateral surface of the middle part of the right lower extremity. The edges are raised and hyperpigmented with a non – indurated base covered by granulation tissue and adhered to underlying structures. There is a hairless, painless nodule of 0.5cm diameter at the center of the ulcer, which had been present from birth. The ulcer started as a small skin excoriation after a mild trauma to the leg. The skin around the ulcer was itchy, painful, discharge serous and bleeds occasionally. There

Correspondence to: Ano-Edward GH, Department of Histopathology, College of Health Sciences, Bowen University, Iwo, Nigeria

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is no regional lymphadenopathy nor any other skin lesion elsewhere. Subsequently, the ulcer

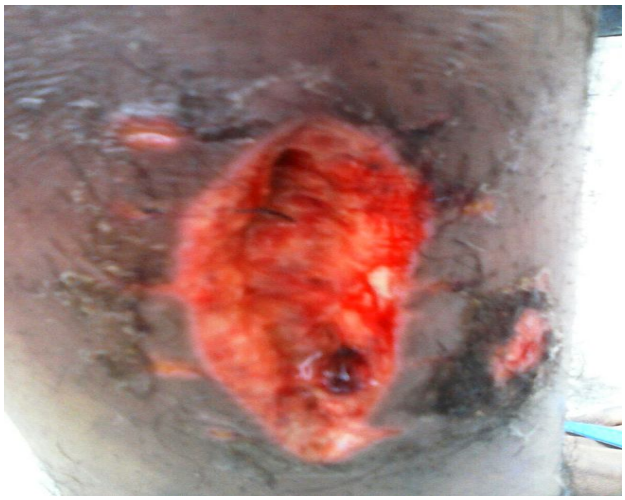

Fig. 1: Ulcer before excision biopsy

was completely excised under anaesthesia with a normal margin of 1cm and a depth extending to the subcutaneous layer. Presently, he has

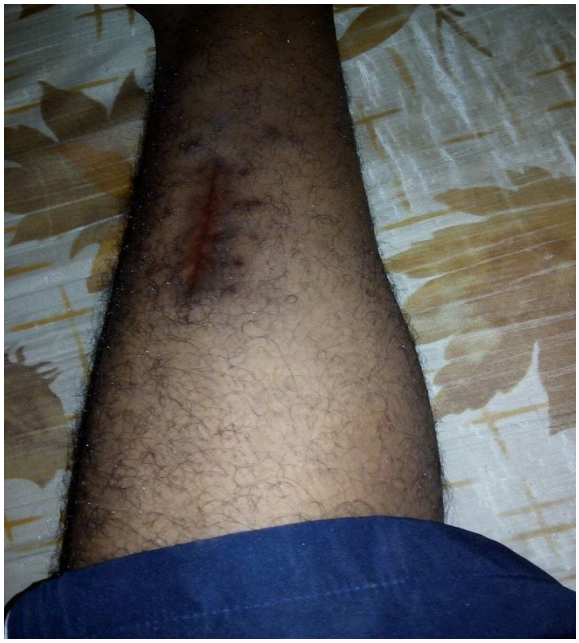

Fig. 2: Post excision of ulcer

been followed up for more than 4 months and the wound healed well without any history of recurrence as at the time of writing this article.

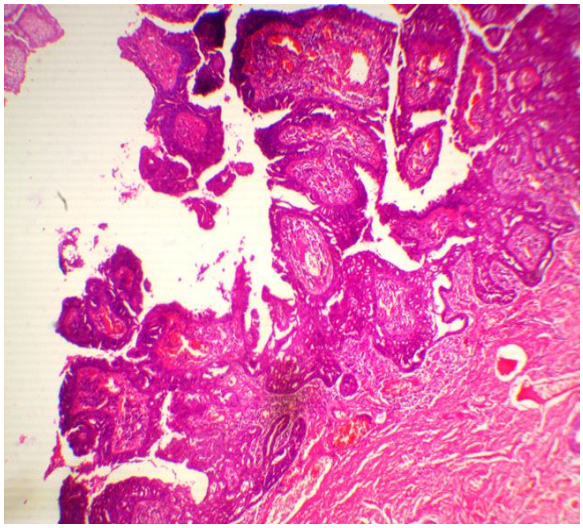

Fig. 3: 3x40 showing cyst with papillary projection lined by 2 layers of epithelium and containing decapitation secretions

We received a single piece of skin bearing elliptical tissue that measures 4x2.5x1 cm. there is a raised papillary lesion on the surface of the skin centrally. Cut surface is gray white with yellowish discoloration. Histologic examination

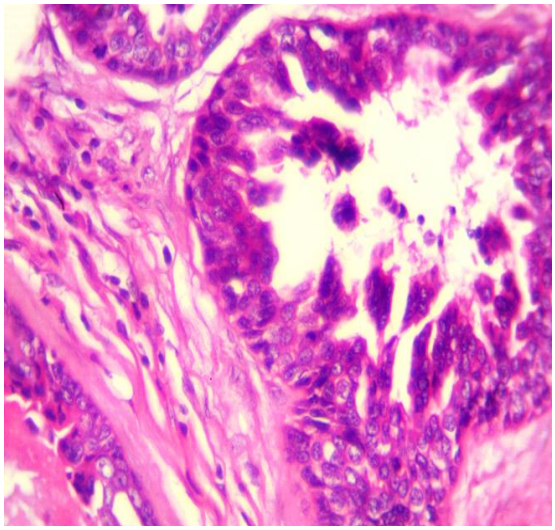

Fig. 4: 2 x400 showing duct with more than 2 layers of epithelium

reveals a skin lesion with mild acanthosis and papillomatosis in areas, there are cystic invaginations extending from the epidermis with villous papillary projections extending into the lumen. The lumen and papillary projections are lined by 2 rows of cells.

Decapitation secretions are seen in some of the cavities. Occasional cavities are lined by more than 2 layers of epithelium. No atypical cells are seen and the stroma is unremarkable.

Discussion

Syringocystadenoma papilliferum is a rare non-malignant adnexal sweat gland neoplasm characterized by asymptomatic, skin-colored to pink papules or plaques with a highly variable appearance, most commonly in the head and neck area. Syringocystadenoma papilliferum (SCAP) occurs most commonly on the scalp or the face; however, tumors have been reported in the vulva, external ear, scrotum and breast in about one fourth of the cases.[5,6,7,8] As of now, there has been just one case report of SCAP on lower leg from Japan. [9]

Three clinical types have been described.[10,11]

(1.) Plaque type: presenting as an alopecic patch on scalp and may enlarge during puberty to become nodular, verrucous or crusted.

(2.) Linear type: consists of multiple reddish pink firm papules or umbilicated nodules 1-10 mm in size commonly occurring over face and neck.

(3.) Solitary nodular type: they are domed pedunculated nodules 5-10mm in size with a predilection for the trunk shoulder and axillae. In the relatively rare cases of Syringocystadenoma papilliferum diagnosed in histopathological evaluation, they originate from the pluripotent cells [12,13,14]. It may spring up within a nevus sebaceous and present in different clinical forms: such as solitary nodule, plaque or in a linear pattern.

It has been reported that 10% cases of Syringocystadenoma papilliferum grew into basal cell carcinoma[15].

The histogenesis of this tumor is still largely controversial [16]. Eventhough, decapitation secretion in the luminal cells of Syringocystadenoma papilliferum suggests an apocrine origin. But it is paradoxical to observe the fact that 90% are found on location devoid of apocrine glands [17]. The only treatment for Syringocystadenoma papilliferum is excision biopsy, which also confirms the diagnosis[18].

Conclusion

Syringocystadenoma papilliferum is a rare tumor and location on the leg is also very rare. It is important to consider it in the differential diagnosis of any longstanding nodular lesion with history dating back to birth and having crusted surface that may ooze serosanguinous fluid because of its continuity with the epidermal surface.

Syrigocystadenoma papilliferum is not always associated with organoid nevus. Clinicians must have a high index of suspicion irrespective of its location and consider histopathological examination for confirmation.

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