

Syringocystadenoma papilliferum arising from a nevus sebaceus mimicking squamous cell carcinoma in a Filipino female: A case report

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ABSTRACT

INTRODUCTION Syringocystadenoma papilliferum (SCAP) is a relatively rare benign adnexal skin tumor which can manifest in a variety of clinical forms. Nearly one-third of cases are known to develop within a pre-existing nevus sebaceus (NS). The peculiar feature of this case was the appearance of a large exophytic tumor within a congenital verrucous plaque, which raised the suspicion of a malignant transformation. This is a case of a young Filipino adult with an unusual presentation of syringocystadenoma papilliferum in a nevus sebaceus mimicking squamous cell carcinoma.

CASE REPORT A 27-year-old Filipino female presented a persistently enlarging exophytic pedunculated cribriform tumor within a congenital verrucous plaque on the left temporal area. The tumor started to appear when she was 20 years old. One month prior to consult, it rapidly increased in size and bled on gentle manipulation. She has neither comorbidities nor any family history of a similar condition. Her physical examination was normal, with no palpable lymphadenopathies. The biopsy showed syringocystadenoma papilliferum on a nevus sebaceus. She underwent carbon dioxide (CO₂) laser excision under local anesthesia. The procedure was uneventful and the patient is on regular follow-up and close monitoring for any possible malignant change or recurrence.

CONCLUSION A case of syringocystadenoma papilliferum on a nevus sebaceus mimicking squamous cell carcinoma in a Filipino female treated with carbon dioxide laser excision was presented. The unusual presentation of SCAP can mimic malignancy and histopathologic evaluation is warranted to rule out malignant transformation for proper management.

KEYWORDS Syringocystadenoma papilliferum, secondary tumors, nevus sebaceus

INTRODUCTION

Syringocystadenoma papilliferum is a relatively rare benign adnexal skin tumor, which can manifest in a variety of clinical forms. Nearly one-third of cases develop in a pre-existing nevus sebaceus.¹⁻³ Using the Health Research and Development Information Network (HERDIN), there are no reported cases of syringocystadenoma papilliferum arising within a nevus in the Philippines. Tumors associated with nevus sebaceus are primarily benign. However, malignant neoplasms such as basal cell carcinoma, squamous cell carcinoma, and sebaceous carcinoma have also been reported to arise within a nevus sebaceus.^{1,2} To rule out malignant transformation, histopathologic examination is usually warranted.

This report describes a 27-year-old Filipino female who consulted due to a rapidly growing

exophytic tumor over a nevus sebaceus. Suspicion of malignant transformation was raised due to the abrupt increase in size (>4 cm over one month) of lesion, malodorous exudates, and associated spontaneous bleeding. The lesion was clinically diagnosed as squamous cell carcinoma but subsequent histopathologic results showed syringocystadenoma papilliferum.

CASE REPORT

A 27-year-old Filipino female was born with a yellow to dark brown, linear, and hairless plaque on the left temporal area. The plaque enlarged gradually and became more verrucous during her adolescent years. A malodorous exophytic pedunculated mass emerged in the center of the plaque when she was 20 years old. One month prior to the consult, the tumor rapidly increased in size, and bled on gentle manipulation.

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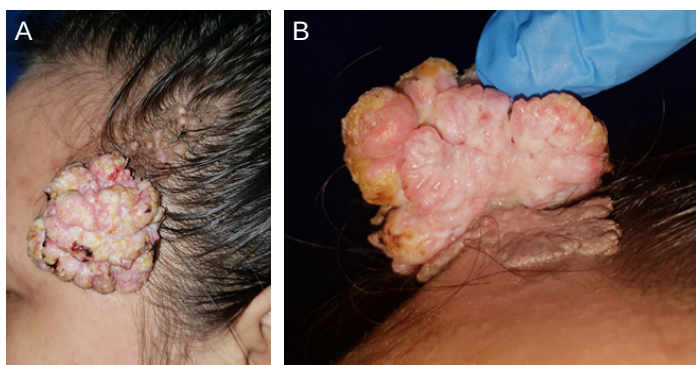


Figure 1. Exophytic pedunculated cribriform tumor measuring 4.5 x 4 x 2.5 cm attached to a yellow-brown verrucous plaque.

Skin examination showed a solitary flesh-colored exophytic, pedunculated cribriform tumor topped with yellow, hemorrhagic crusts with clefts filled with purulent, foul-smelling exudates measuring 4.5 x 4 x 2.5 cm (Figure 1). Its base showed a hairless, yellow to brown, well-demarcated verrucous linear plaque measuring 3.5 x 6 cm (Figure 1). The lesions were movable, not indurated, not friable and non-tender on palpation. There were no palpable lymphadenopathies.

Skin punch biopsy on the verrucous plaque, and shave biopsy of the exophytic tumor was done. Histologic examination of the verrucous plaque showed serum crusts, hyperkeratosis, papillomatosis, irregular acanthosis, and dense superficial perivascular mixed cell infiltrates consisting of lymphocytes, histiocytes, and few eosinophils. There were immature hair follicles with enlarged mature sebaceous glands in the dermis (Figure 2). Microsections of the exophytic tumor showed hyperkeratosis, varying degrees of papillomatosis, and irregular acanthosis. There were cystic invaginations from the epidermis extending to the dermis with numerous papillary projections in which the

luminal rows of cells displayed evidence of active decapitation secretion. There were dilated and congested dermal blood vessels, dense superficial and deep perivascular mixed cell infiltrates consisting of lymphocytes, histiocytes, eosinophils, and plasma cells (Figure 2). There was absence of atypia, mitotic figures, or nuclear pleomorphism. The case was signed out as syringocystadenoma papilliferum in nevus sebaceus.

The nodular-type of tumor appeared on the temporal area and scalp, which are areas where cosmesis is a concern. Since histopathology results revealed a benign lesion, carbon dioxide laser excision was done at an output of 8-12 watts in ablative continuous mode under local anesthesia with curettage and healing by secondary intention (Figure 3). The postoperative wound healed with granulation tissue formation and good reepithelialization. The patient tolerated the procedure well and healing was uneventful. The patient is on regular follow-up and close monitoring for any possible malignant change or recurrence.

DISCUSSION

Nevus sebaceus is a congenital hamartoma with epidermal, sebaceous, and apocrine differentiation.^{4,5} It occurs in 0.3% of newborns or early childhood.⁴ It is predominantly distributed on the head and neck, presenting as hairless verrucous plaque.¹ Nevus sebaceus usually undergo three clinical and developmental stages. The alopecic or infantile stage (stage I) appears as a hairless yellowish plaque with primordial hair follicles and hypoplastic sebaceous glands on histopathology. The plaque becomes more prominent and firm during the verrucous-papillomatous plaque stage (stage II). Benign and malignant neoplasms usually develop at the tumoral stage (stage III).^{4,6} The lesions usually become more verrucous at puberty, and hormonal influence may play a role in its pathogenicity.^{1,4}

The susceptibility of a nevus sebaceus for secondary tumors has been associated with somatic mosaicism. Mutations in HRAS and KRAS in nevus sebaceus lesional keratinocytes led to the

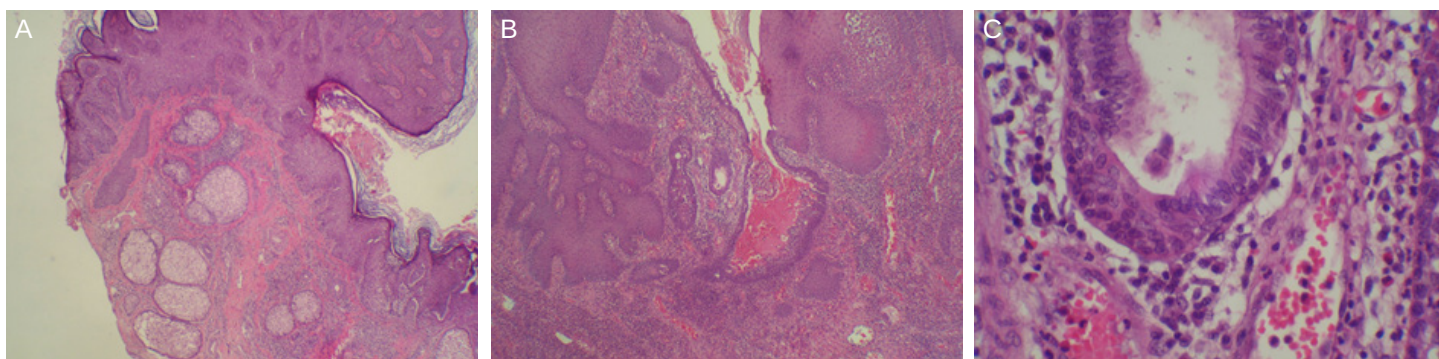


Figure 2. A. Skin punch biopsy of the verrucous plaque showed an exophytic structure showing hyperkeratosis, papillomatosis, and irregular acanthosis with immature hair follicles and enlarged sebaceous glands in the dermis (H&E stain; 10x). B. Shave excision biopsy of the exophytic tumor showed hyperkeratosis, varying degrees of papillomatosis, irregular acanthosis with cystic invaginations from the epidermis extending to the dermis (H&E stain; 10x). C. Numerous papillary projections in which the luminal rows of cells displayed evidence of active decapitation secretion (H&E stain; 40x).

activation of the RAF-MEK-ERK and phosphoinositide 3-kinase signaling pathways with subsequent cellular proliferation and increased susceptibility to tumor formation. Secondary neoplasms may develop in approximately 25% of nevus sebaceus cases, with mostly benign tumors.⁵ Nevus sebaceus is associated most commonly with benign neoplasms such as trichoblastoma and syringocystadenoma papilliferum. However, malignant neoplasms such as basal cell carcinoma, squamous cell carcinoma, and sebaceous carcinoma have also been reported to arise within a nevus sebaceus.^{1,2,4,6} There are only two published reports in local literature with tumors arising in nevus sebaceus. Piansay-Soriano et al. reported a case of a 9-year-old girl with a basal cell carcinoma arising from a nevus sebaceus on the scalp and an infundibuloma arising from a separate nevus sebaceus on the face.⁷ Lagunzad et al. reported sebaceoma and squamous cell carcinoma cases arising on top of a congenital nevus sebaceus and epidermal nevus, respectively.⁸

Syringocystadenoma papilliferum usually appears at birth and before puberty in 50% and 15%-30% of cases, respectively.^{1,3} It occurs commonly on the scalp or the face and typically measures from 1 cm to 3 cm. Clinical presentations include plaque-type, linear-type, and nodular-type. The plaque-type is often associated with nevus sebaceus, characterized as a hairless patch on the scalp. The linear-type comprises of multiple reddish-pink papules or nodules on the face and neck region. The nodular-type are dome-shaped pedunculated nodules measuring between 5-10 mm with predilection on the trunk, shoulder, and axilla.³ Unusual presentations of a large syringocystadenoma papilliferum (>4 cm), with bleeding, and malodorous exudates have been reported in the literature. Agrawal et al. reported a 35-year-old male presenting with multiple papules and nodules on the right lower abdomen with an accelerated increase in size and foul-smelling discharge diagnosed as syringocystadenoma papilliferum.⁹ Hence, a biopsy is warranted for proper diagnosis and management.

Histopathologic examination of syringocystadenoma papilliferum often shows characteristic epidermal invaginations, and the presence of papillary processes lined with two epithelial cell layers. Decapitation secretion is usually found at the luminal surface. Another common feature is the presence of inflammatory infiltrates, mostly of plasma cells.⁹

Malignant transformations arising from a nevus sebaceus have been reported, and is related to aggressive behaviors, i.e. sudden accelerated growth, the large size of a developed nodule, bleeding, and the presence of metastatic lymph nodes.^{1,2} In our case, the rapid growth of the tumor warranted further investigation.

The treatment of choice for syringocystadenoma papilliferum is excision, followed by a detailed histological examination.³ The definitive treatment for nevus sebaceus is full-thickness epidermal and dermal excision. This is because the nevus extends deep as the subcutaneous tissue, including adnexal structures. Other treatment

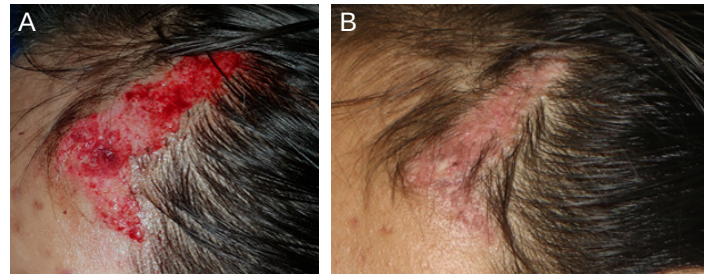


Figure 3. A. Photograph of the left temporal area immediately after CO2 laser treatment. B. There is a reduction in size and proper wound healing one month after (30 days after CO2 laser treatment).

options are Mohs micrographic surgery or carbon dioxide laser excision in unfavorable excision and grafting areas. Cribrier et al.^{2,6} reviewed 596 cases of nevus sebaceus and reported that most of the associated tumors were benign.² The authors believe that, in children, close observation and clinical follow-up are better than prophylactic excision. Patients must have a comprehensive understanding of the condition, and should be compliant with the recommended follow-up to observe new growth or changes suggesting malignancy.^{2,6}

Most studies suggest that full-thickness surgical excision is the treatment of choice. In cases where the ensuing defect is too large for primary closure, rotation flap and tissue expansion procedures are more suitable. However, these extensive surgeries will require a more prolonged recovery period, a slower cicatrization, and risk of hypertrophic scars and keloids.^{6,10} The carbon dioxide (CO₂) laser has been recognized as an excellent alternative to surgery. CO₂ laser has benefits of reduced healing time, less requirement for anesthesia, less bleeding and inflammation, and easy access to anatomically difficult areas.¹⁰ Side effects are minimal such as pain, erythema, and edema. The CO₂ laser could be a treatment option in areas unfavorable to excision and grafting, but it is critical to consider how much lesion is being removed. There is a greater chance of recurrence if fragments remain, so further follow-up is necessary.⁶

CONCLUSION

A case of syringocystadenoma papilliferum in nevus sebaceus mimicking squamous cell carcinoma in a Filipino female treated with CO₂ laser excision was presented. It is essential to be mindful of the various presentations of syringocystadenoma papilliferum because atypical forms can mimic malignancy. Histopathologic evaluation is warranted to rule out malignant transformation for proper management. CO₂ laser ablation can be a treatment alternative to produce a cosmetically acceptable result but close observation and further follow-ups are necessary for monitoring of relapse.

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