Case Report:
Verrucous Carcinoma Arising Within Syringocystadenoma Papilliferum

Natalie Lama Monticciolo, Jodie D. Schmidt, and Michael B. Morgan
Dermatology Department, Sun Coast Hospital/Nova Southeastern, Largo, Florida; Pathology Department, University of South Florida College of Medicine, and James Haley Veteran’s Hospital, Tampa, Florida

Abstract. Syringocystadenoma papilliferum (SP) commonly arises in conjunction with nevus sebaceous. Other less common lesions reported with SP include apocrine adenoma, condyloma acuminatum, hidradenoma papilliferum associated with hidrocystoma, poroma folliculare, and a single case of verrucous carcinoma. We report the second case of verrucous carcinoma in conjunction with SP. (received 15 March 2002, accepted 23 April 2002)

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Introduction

SP was originally described by Werther [1] in 1913 as an unusual tumor that was termed “naevus syringadenomatous papilliferus.” SP usually occurs on the head and neck, but cases have been reported that involve the chest [2], breast [3], axilla [4], genitalia [5], arm [6], thigh [7], and eyelid [8]. This tumor is more common in women and is typically present at birth or in early childhood. It appears as hairless plaques until puberty, when the lesion becomes more nodular or verrucous. The plaques commonly contain small cystic and umbilicated papules [6]. Their color range from brown to red to yellow. The plaques often become indurated and crusted, owing to stromal fibrosis, dried blood, or exudates from associated apocrine glandular secretions [6].

Approximately one-third of recorded cases of SP arise in association with a pre-existing nevus sebaceous of Jadassohn. Less common lesions that are associated with SP include apocrine adenoma [9], condyloma acuminatum [5], hidrocystoma and hidradenoma papilliferum [10], poroma folliculare [11], apocrine acrosyringeal keratosis [12] and giant comedo [13]. Malignant degeneration within SP is rare and includes ductal sweat gland carcinoma [14], malignant SP [15,16], and a single reported case of verrucous carcinoma [17]. We report the second case of verrucous carcinoma in conjunction with SP.

Case history

A 62-yr-old white woman presented with complaint of a dull aching sensation in the sacral area. The patient stated that a birthmark had been present in the area for “as long as she could remember.” She noted that the birthmark lesion had begun to grow within the past 6 mo.

Physical examination revealed a fluctuant subcutaneous mass (3 x 3 x 3 cm) located in the superior portion of the gluteal cleft and surrounding sacrum. The mass was associated with a mamillated plaque (6 x 3 cm). There were no other cutaneous lesions; no lymph nodes were palpated. The entire plaque and underlying mass were excised under local anesthesia. The patient’s post-operative course was uneventful. At 1-yr follow-up, there is no evidence of local recurrence or metastatic dissemination.
**Fig. 1.** (Above, left) Low-power photomicrograph of the endophytic mass depicting verrucoid epidermal hyperplasia with adjacent glandular lumina (H&E stain, 25X magnification).

**Fig. 2.** High-power photomicrograph depicting the lateral walls of the lesion, with apocrine epithelium and plasma cell infiltrate (H&E stain, 100X magnification).

**Fig. 3.** High-power photomicrograph depicting the deep aspect of the lesion and the pushing border of verrucous carcinoma (H&E stain, 100X magnification).
**Pathology**

The gross specimen (7 x 6 x 5 cm) consisted of a slightly hyperpigmented and mamillated cutaneous plaque. The plaque was contiguous to a well-circumscribed, roughly spherical, cystic mass (4 x 4 x 4 cm). The cut surface of the mass showed a thickened and indurated wall containing a greasy foul-smelling substance.

Histologic sections showed irregular epidermal hyperplasia contiguous with an endophytic cystic mass that occupied all levels of the dermis and superficial subcutaneous fat (Fig. 1). The lateral walls of the lesion comprised bi-layered basilar cuboidal and superficial columnar epithelium with apical snouts (Fig. 2). The surrounding stroma contained increased numbers of chronic inflammatory cells including numerous plasma cells.

The basilar portion of the cystic mass consisted of irregular islands of partially keratinized squamous epithelium. The outermost layer of epithelium showed a broad border that contained scattered typical and rare atypical mitoses and dyskeratotic cells (Fig. 3). Immunohistochemical staining of the squamous cells showed an increased proliferation index and increased antibody labeling of MIB-1, compared to the overlying epidermis. The lateral portions of the cystic structure were labeled by an antibody to gross cystic disease fluid protein-15 (GCDPF-15).

**Discussion**

Herein, we describe an unusual case of verrucous carcinoma associated with SP. As noted in the introduction, SP has been reported in conjunction with a variety of benign and malignant cutaneous entities.

Basal cell carcinoma was found to occur in 9% of cases with SP and concomitant nevus sebaceous [6]. Ductal sweat gland carcinoma has been reported in 9 cases associated with nevus sebaceous; all of these cases had an indolent biological behavior. In only one case was the ductal sweat gland carcinoma connected to SP arising in nevus sebaceous [14].

Syringoadenocarcinoma papilliferum is a rare malignant counterpart of SP; only 4 cases have been described [15,16]. It is considered a type of sweat gland carcinoma. Histologically, the lesion consists of solid and cystic areas lined by a multilayer of disorderly and atypical cuboidal and columnar cells.

One case of verrucous carcinoma associated with SP was previously reported [17]. It was a slowly expanding, exophytic mass that had been present for 15 yr on the thigh of an elderly man. On histologic examination, the lesion was a verrucous tumor formed by strands of large downward epithelial proliferations with mild atypia and a moderate lymphocytic infiltrate at the tumor-stroma interface. Situated within the verrucous mass were numerous ductlike invaginations lined by a bi-layer of cuboidal and columnar cells. An inflammatory infiltrate of plasma cells and lymphocytes was also present within the connective tissue [17].

Verrucous carcinoma is classified as a low grade, extremely differentiated, squamous cell carcinoma with a propensity for localized growth and recurrence [18]. It can occur at a variety of sites and afflict all ages and both genders, but it classically presents as an ulcerated polypoid mass on the digits or genitals of elderly men. The tumor's metastatic potential is low, although metastases have been reported with dedifferentiation following radiation therapy.

The histogenesis of SP is controversial. Pinkus [4] postulated that the adenomatous portion is formed secondary to a hamartomatous proliferation of the involved skin and that the papillomatous areas may be derived from apocrine or eccrine glands. Lever [19] proposed that this lesion is a hamartoma derived from undifferentiated pluripotential cells. Hackney and Helwig [7] suggested that syringocystadenoma papilliferum is either an adenoma of eccrine ductal origin or is derived from the ducts of a gland intermediate between eccrine and apocrine. Mammino and Vidmar [20] concur with the theory that SP develops from pluripotential appendageal cells and hybrid type apoecocrine glands.

The histogenesis of verrucous carcinoma has been variably attributed to human papillomavirus infection, chronic irritation, and/or inflammation [17,18]. Repeated testing for human papillomavirus by a variety of methods has yielded conflicting results [21,22]. Irritation related to the anatomic location and/or inflammation, as documented in the present
case, may predispose to oncogenic transformation. In view of the rarity of the association, it is also possible that the occurrence of verrucous carcinoma in conjunction with SP is serendipitous and etiologically unrelated.

References