Verruciform genital-associated xanthoma with acantholysis: Report of a rare case

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INTRODUCTION

Verruciform xanthoma occurs most commonly in the oral cavity. Another predilection site is the genital region, where it has been termed *verruciform genital-associated xanthoma* (Vegas xanthoma).[1] We report an unusual case on the scrotum that demonstrated extensive acantholysis.

CASE REPORT

A 43-year-old male patient presented with an exophytic and pedunculated 0.8-cm papule on the scrotum that had been present for approximately 6 months and that was clinically believed to represent a skin tag. After surgical removal, the histologic specimen demonstrated an exophytic and pedunculated architecture (Fig 1, *A*). The epidermis was papillomatous and acanthotic and showed extensive acantholysis, which comprised the entire thickness of the spinous layer in some areas and its superficial portion in others (Fig 1, *B* and *C*). Corps ronds or grains were not observed. The dermal papillae contained an infiltrate of epithelioid cells with a foamy cytoplasm (xanthoma cells) (Fig 1, *B* and *C*). These cells were immunohistochemically positive for the histiocytic marker CD163 (Fig 1, *D*) and negative for the melanocytic marker SOX-10 (not shown).

DISCUSSION

The combination of histopathologic features observed in our patient appears to be rare, and we are aware of only 1 other case in the literature with a similar constellation of findings. This case occurred on the penile skin of a 53-year-old patient and histologically showed acanthosis and papillomatosis of the epidermis with acantholysis and numerous foamy cells within narrow dermal papillae. These findings were interpreted as a verruciform xanthoma with acantholysis.[2] Even though this case demonstrated more superficial acantholysis than our case and a broad-based papillary rather than a taglike architecture, the similarity of its key features suggests that it represents the same entity as in our case.

Because the combination of a papillary architecture with xanthomatosis and acantholysis has not been firmly established as a histopathologic entity, it is difficult to assign an unequivocal diagnosis in our case. Depending on which of the histologic features are considered as the primary process, at least 2 diagnostic appellations appear possible. In the previous case report, the process was interpreted as a verruciform xanthoma with superimposed acantholysis,[2] but alternatively it could also be designated as an acantholytic acanthoma with incidental xanthoma cells. Such incidental xanthoma cells have been observed in association with a variety of primary processes.[3] These 2 alternative diagnoses will be considered in the following.

Verruciform xanthomas occur most often in the oral cavity, and their histopathologic features have been well defined in several case series.[4-6] The defining histopathologic features common to all verruciform xanthomas include acanthosis, papillomatosis, or both, with numerous foamy histiocytes within dermal papillae. Based on the predominant histologic growth pattern, 3 subgroups have been distinguished. The verrucous type shows features similar to a wart, the papillary type has an exophytic

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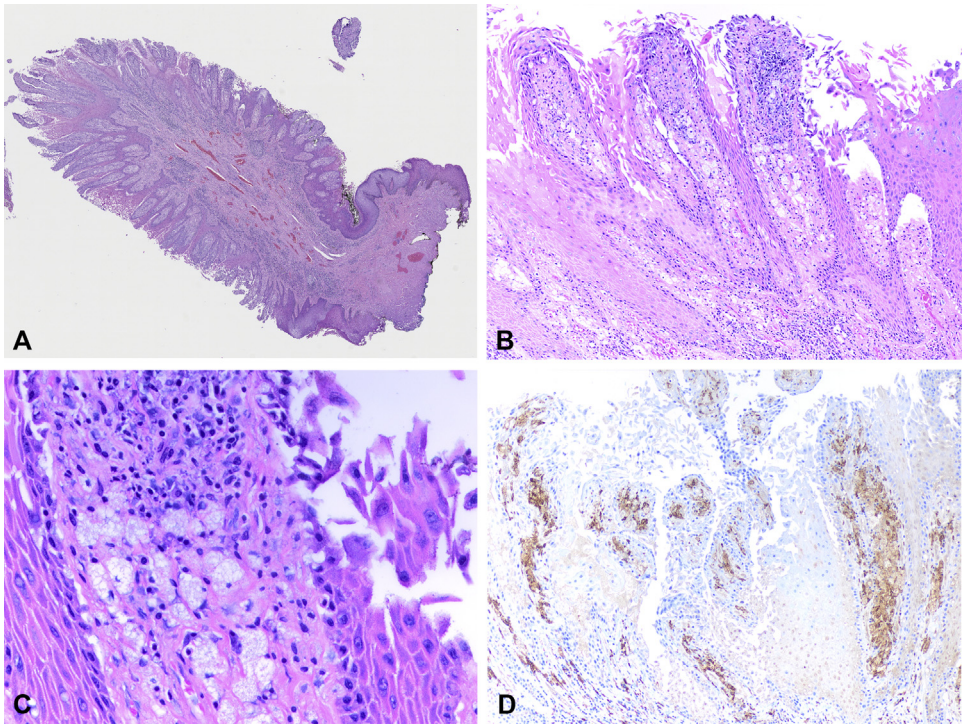

Fig 1. Verruciform xanthoma with acantholysis. **A**, An exophytic and pedunculated architecture with acanthosis and papillomatosis. **B** and **C**, The dermal papillae are covered by acantholytic epidermis and contain foamy histiocytes. **D**, The foamy histiocytes stained with CD163 on immunohistochemistry. (Hematoxylin-eosin stain; original magnifications: **A**, ×20; **B**, ×100; **C**, ×400; **D**, ×100.)

architecture with fingerlike projections, and the flat type demonstrates minimally elevated acanthosis of the epidermis.[4-6] In addition to the oral cavity, verruciform xanthomas are also encountered in the skin, where they have a similar verrucous, papillary, or flat architecture and are predominantly located in the genital area.[1] Exophytic and pedunculated cases have also been described.[1] In contrast, acantholytic acanthomas occur on the skin in a widely distributed localization, most commonly on the trunk, and in the largest series reported to date only 2 of 31 cases were located in the groin or genital area.[7] The architecture of acantholytic acanthomas is predominantly acanthotic/verrucous and only rarely papillary.[8,9] Thus, our patient's case, which was located in the genital area and had a papillary architecture, matches the features of an acantholytic acanthoma less well than those of a verruciform xanthoma, which we therefore consider the more likely diagnosis.

It could also be argued that the findings observed in our case represent a collision of a verruciform xanthoma with an acantholytic process (eg, Hailey-Hailey disease, papular acantholytic dyskeratosis, pemphigus vulgaris). In our patient, this appeared unlikely because no clinical stigmata of these diseases were present.

The combination of acantholysis and foamy histiocytes appears to be a rare phenomenon. In addition to the current case and the reported case mentioned earlier, the only other case we are aware of was an intraoral warty dyskeratoma with foamy histiocytes. This case was clearly different from the current case because it showed the typical endophytic architecture with a craterlike depression of a warty dyskeratoma.[10]

With only 2 cases on record, the papillary, acantholytic, and xanthomatous proliferation presented here can obviously not be classified as an entity at this point, and its final nosology awaits reporting of additional similar cases.

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