CORRESPONDENCE

A large fungating verruciform xanthoma of the scrotum in association with arteriovenous malformation mimicking giant condyloma

Dear Editor,

A 97-year-old man with underlying hypertension and benign prostate hyperplasia, had a large tumor on the right side of his scrotum for decades, during which time the tumor gradually enlarged. Easy bleeding upon irritation was recently noted, which prompted him to seek medical help. Physical examination revealed a 5 cm × 3 cm × 3.4 cm, pinkish fungating tumor, with a warty surface and focal hemorrhagic crust. Engorged blood vessels were noted near the stalk (Figure 1). Sonography showed hypervascularity. Projectile bleeding from the stalk base was noted during tumor excision.

Histopathological examination revealed a large, exophytic fungating tumor consisting of numerous individual fibroepithelial polyp-like structures (Figure 2A). The epidermis showed acanthosis, relatively uniform elongation of thin rete ridges, and spongiosis with infiltration of neutrophils, but without granular layer or hyperkeratosis (Figure 2B). There was no koilocytosis or keratinocytic atypia. The connective tissue core showed marked edema, many dilated blood vessels, and a variable amount of inflammatory infiltrate consisting of abundant plasma cells, some small lymphocytes, and numerous histiocytes with abundant, pale foamy cytoplasm, which are CD68-positive (Figure 2C). The stalk of the lesion contained many dilated or congested large blood vessels, some of which had an uneven thickness in their walls (Figure 2D). Based on these findings, a diagnosis of fungating verruciform xanthoma (VX) in association with arteriovenous malformation was made. No recurrence was observed at the 6-month follow-up.

VX is an uncommon type of mucocutaneous lesion, mostly affecting the oral cavity. Cutaneous involvement is rare, and usually presents as a pink to orange nodule or plaque, with a warty surface primarily affecting the anogenital area. The rate of scrotal involvement is higher in Japanese than in Western populations. The main histopathological features are epidermal hyperplasia without keratinocytic atypia or viral inclusion body and numerous foamy or xanthomatous cells in the accentuated papillary dermis. The exophytic lesion frequently shows hyper- or parakeratosis and neutrophilic exocytosis. The foamy cells are positive for CD68 and weakly positive for factor XIIIa, but not S–100 protein, thus supporting that these foamy cells are histiocytes of macrophage/monocyte or dermal dendrocyte lineage. The inflammatory infiltrate also contains plasma cells and lymphocytes. The reticular dermis typically shows vascular ectasia, but does not have xanthomatous cells, which is different from lipidemic xanthoma.

The etiology of VX remains elusive. Since it has been reported to be associated with many inflammatory and neoplastic conditions, it is considered a reactive phenomenon rather than a neoplastic process. Several possible pathogenic mechanisms are proposed, including chronic irritation resulting in keratinocyte damage and xanthomatous cell response, congenital defects, abnormal immune status, and anatomic factors. The present case is very similar to the case described by Kishimoto et al, in which the authors hypothesized that the formation of VX might be related to the vascular ectasia deep in the lesion in the reticular dermis. Their case was a 75-year-old Japanese man presenting with a yellowish, verrucous, pedunculated nodule on the scrotum. The lesion was accompanied by a pulsating subcutaneous induration covered by lightly purplish skin. The deep dermis showed severe vascular ectasia, including capillaries and venous blood vessels and variable numbers of thin-walled and thick-walled blood vessels in close association with one another, i.e., arteriovenous malformation. They hypothesized that deep dermal vascular ectasias are initially induced by an unknown etiology, causing the activation of both endothelial cells and pericytes in the papillary dermis. Then, these activated cells release various cytokines and growth factors, which induce hyperplasia of epidermal keratinocytes and accumulation of macrophages. Finally, accumulated macrophages scavenge lipids and are converted to xanthomatous cells. The underlying arteriovenous malformation affects the nutrients and metabolism of overlying blood vessels, and may lead to the deep dermal vascular ectasia.

Figure 1 A large fungating pedunculated tumor with verrucous surface is present at the scrotum. An engorged violaceous vessel is connected to the base of the stalk.
Another unusual feature of the present case is that the lesion is fairly large. There are only a few reported cutaneous lesions with sizes larger than the present one, and almost all of the lesions were plaques instead of polypoid, as in the present case. The large size of the current lesion may be because it had been allowed to grow over several decades. A large verrucous tumor over a scrotal area usually leads to the first impression of giant condyloma, among other differential diagnoses such as verrucous carcinoma, squamous cell carcinoma, seborrheic keratosis, giant molluscum contagiosum, condyloma latum, and xanthoma. Therefore, it is important for clinicians and pathologists to be aware of this disease and to avoid unnecessary overtreatment.

In summary, we present a rare case of giant VX on the scrotum, mimicking giant condyloma. This is the second reported case of VX in association with arteriovenous malformation in the literature.

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Figure 2 (A) A large fungating tumor consisting of numerous fibroepithelial polyp-like structures; (B) the epidermis shows characteristic hyperplasia with neutrophilic infiltration; (C) there are abundant foamy cells in the papillary dermis bounded by elongated thin rete ridges. (D) arteriovenous malformation present in the stalk of the lesion shows uneven thickness in the vessel wall. Inset: These foamy cells are CD68-positive, which are histiocytes of macrophage/monocyte or dermal dendrocyte lineage.