Keratoacanthoma on an Epidermal Nevus in a Filipino Male

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ABSTRACT

Keratoacanthoma is a cutaneous tumor that most often presents as a solitary lesion in middle-aged to elderly, light-skinned individuals on sun-exposed areas. It is characterized by rapid growth and may be followed by spontaneous involution. This is a case of a seventy-year-old Filipino male who had a one month history of a solitary nodule that suddenly appeared on an epidermal nevus. Histopathologic diagnosis revealed a keratoacanthoma.

INTRODUCTION

Epidermal nevus is a benign epithelial tumor that presents sporadically at birth, or may have an onset in infancy or childhood and may grow or extend in size until puberty. The incidence has been reported to range from 1 to 3 per 1000 live births with equal sex predilection.[1,2] It may present as localized or widespread tumors which are characteristically skin-colored, brown or dark brown verrucous papules forming warty plaques usually in linear or whorled distribution. Sites of predilection are the trunk and extremities.

Rare cases have been reported on neoplasms such as basal cell carcinomas and squamous cell carcinomas arising on an epidermal nevus, most commonly arising in the middle aged and the elderly.[1, 2, 3, 4] There are only few cases of reported keratoacanthoma and keratoacanthoma-like lesions arising in an epidermal nevus.

Case Report:

A 70-year-old Filipino male consulted because of sudden appearance of a solitary bud-shaped nodule over a linear verrucous plaque, which extended from the right posterior auricular area to the right lateral side of the neck. The plaque is asymptomatic and has been present since birth.

The condition started four weeks prior to consult, when he noted a pink papule with an adherent central scaly center associated with intermittent mild pain. This gradually enlarged to form a nodule. Patient is a known hypertensive and diabetic. He is a smoker with fifty pack years and an alcoholic beverage drinker. The patient has a history of constant sun exposure for almost forty years, as his hobby includes playing outdoor tennis. His family history was negative for malignant skin tumors.

Physical examination showed a bud-shaped slightly erythematous movable smooth firm nodule measuring 0.8 cm x 0.5 cm topped with white scale on a hyperpigmented verrucous plaque arranged in a linear configuration over the right retroauricular area extending over the lateral side of the neck (Fig. 1). Regional lymphadenopathy was absent. Excision biopsy of the nodule and part of the verrucous plaque was done. Histologic findings of the nodule revealed a keratoacanthoma while that of the verrucous plaque showed an epidermal nevus. He followed up six weeks later, and there was no recurrence nor new growth noted.

Discussion

Keratoacanthoma is a common epithelial tumor of the skin most commonly in elderly males.[9] Predisposing factors include UV light exposure and smokers, seem to be more affected than non-smokers.[2,5] Most lesions develop on hair bearing skin, and may arise on a site of previous trauma ex. surgery. It has three clinical stages namely 1) Proliferative or growth phase, which is characterized by a rapidly growing erythematous papule with a smooth surface reaching a size of 1-2 cm. This phase lasts about 2-10 weeks. 2) Mature or stationary phase of similar duration with a central keratotic core on the nodule. 3) Involution or resolving phase that takes around 8-50 weeks characterized by flattening of the nodule leaving a scar. [6]

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The clinical and histopathologic features of the warty papules and plaques in a linear distribution on the right retroauricular area extending to the lateral neck, which has been present since birth is consistent with an epidermal nevus. The sudden appearance of a solitary pinkish papule, which rapidly increased in size over a period of four weeks, forming a firm erythematous nodule with a white scaly center, were likewise consistent with the clinical features of keratoacanthoma. The diagnosis of keratoacanthoma was confirmed by histopathologic findings of keratin-filled invagination of the epidermis with surrounding areas of glassy eosinophilic appearance, keratinocytes with nuclear atypia and mitotic figures, dyskeratosis, irregular acanthosis and dense superficial perivascular lymphohistiocytic cell infiltrates admixed with numerous eosinophils.

Epidermal nevus is a benign proliferation of keratinocytes that usually present at birth or childhood. Lesions may be localized, following linear patterns known as "the lines of Blashko." When extensive in distribution it is referred to as systematized epidermal nevus and may be associated with extra-cutaneous manifestations such as CNS and skeletal abnormalities.

Previous reports have indicated that the linear epidermal nevus may serve as a site of development for skin tumors.[1] Verrucous and adnexal carcinomas, bowen's disease and basal cell carcinoma although rare have all been reported to arise in linear epidermal nevus. Squamous cell carcinoma arising in an epidermal nevus is extremely rare. According to Massod et.al only less than ten cases of such have been reported in literature.[4] Such malignant transformations are most common in middle aged and elderly.[1,2,4] Keratoacanthoma arising in an epidermal nevus is an infrequent phenomenon. Review of literature, showed only two reported cases of such, both in 1982 on a 26 - year-old white woman and a 32-year-old white man. Presentations were on an epidermal nevus on the right arm for the former and the left arm for the latter.[3] Other reported cases of keratoacanthoma development were on a nevus sebaceous. This also has a low incidence.[7]

Some authors believe that keratoacanthoma is a subtype of squamous cell carcinoma, where as others say that it is a precursor lesion to SCC. [8] Some however, believe that it is an abortive malignancy that rarely progresses to invasive squamous cell carcinoma.[9] Both keratoacanthomas and SCC may grow rapidly, but regression has been suggested as the gold standard for the diagnosis of keratoacanthoma.[8] Due to the uncertainty of differentiating one from the other, the recommendation is that it be treated rather than observed. The use of topical, intralesional and systemic treatments like 5-fluorouracil, methotrexate, triamcinolone and interferon-**α** have proven successful. But in most solitary keratoacanthomas, complete surgical excision is the treatment of choice. [2,10]

Conclusion

We report a seventy-year-old male, who presented with a neoplasm clinically and histopathologically consistent with keratoacanthoma on an epidermal nevus. We must exercise vigilance with any new growth that develops rapidly within an epidermal nevus and always regard it as suspicious for malignancy. There are no clinical or histologic criteria to classify a potential keratoacanthoma as a benign tumor that might spontaneously regress, or as a neoplasm with metastatic potential.[11] Some authors believe that in terms of treatment, it is best to regard a keratoacanthoma as a variant of squamous cell carcinoma hence, treatment should be instituted once diagnosed. Wide excision is the treatment of choice for solitary keratoacanthoma. The rate of recurrence is 4 -8% within a few weeks after surgery and maybe the result of incomplete excision[11]. Patients should be advised to perform regular skin examination. Sun protective measures should be practiced at all times. This includes the use of sunscreen, protective clothing, wide-brimmed hats and sun avoidance during the peak hours. Predisposing factors like potential carcinogens such as smoking should be avoided.

It is highly recommended that follow-up be done with complete skin examination during each visit at regular intervals between 3-12 months. Sites of previous lesions and treatment should be assessed for recurrences and appearance of any new lesion.

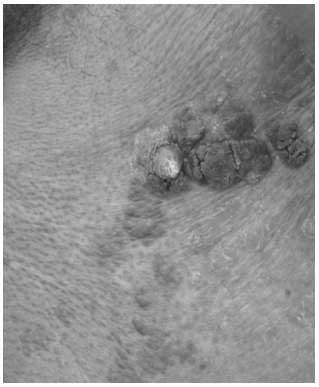
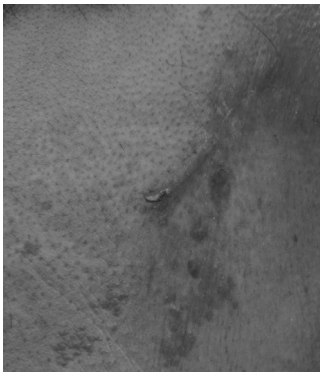


Figure 1. solitary bud shaped pinkish nodule topped with keratotic dry scales on the center of an epidermal nevus



Post-excision of the keratoacanthoma on follow-up six weeks later.

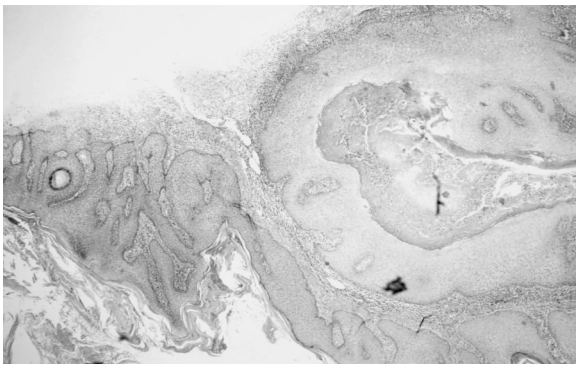


Figure 2. Right side: keratin-filled invagination of the epidermis Left side: hyperkeratosis, focal parakeratosis, papillomatosis, irregular acanthosis with horn cysts (Hematoxylin-eosin stain, magnificaction x 25)

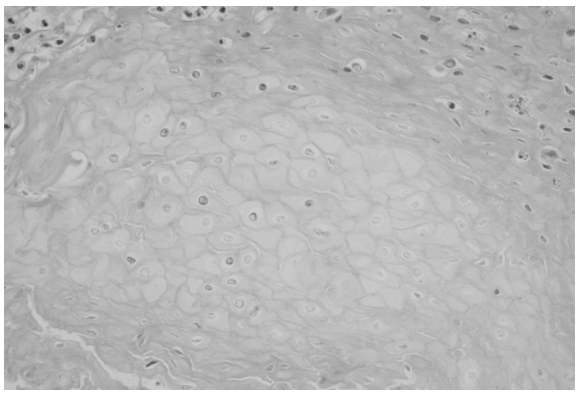


Figure 3. Presence of dyskeratotic cells (above) Surrounding areas with eosinophilic glassy appearance (below) (Hematoxylin and eosin; magnification 40x)

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