A Case of Clonal (Nested) Variant of Seborrheic Keratosis

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Summary: We report a case of clonal (nested) type of seborrheic keratosis. Although seborrheic keratosis is a commonly encountered lesion in dermatology OPD but clonal (nested) variant of seborrheic keratosis is quite rare and can be diagnosed only after proper histopathological examination.

Keywords: Seborrheic keratosis, clonal type, nested type.

I. Background

Seborrheic keratoses are common, benign, pigmented epidermal tumors.[1] Many terms such as senile wart, melanoacanthoma, basal cell papilloma, senile keratosis and seborrheic wart have been applied, but seborrheic keratosis is the most widely accepted term.

These usually develop after the age of 50 years although occasionally, seen in young adulthood without any sexual predilection.[1] The common site of involvement includes the trunk, particularly the interscapular area, sides of the neck, the face and the arms. The tumors are not, however, seen on the mucous membranes.[2] Lesions appear as coin-like, sharply demarcated, exophytic lesions and are “stuck on the skin” with a verrucous, rough, dull or punched-out surface. Flat lesions often have a smooth surface and are scarcely elevated above the surface of the skin.[2]

The etiology is not well-known, although heredity, sunlight and human papilloma virus (HPV) have been suggested as risk factors. Recent genetic studies have suggested that somatic mutations in Fibroblast Growth Factor Receptor 3 (FGFR3) gene are important in the development of these lesions.[3] Although seborrheic keratosis is a commonly encountered lesion by the dermatologists, the clonal variant is very rare and can be diagnosed only after histopathological examination.

II. Case Presentation

A 38 year old, muslim, female patient presented to our Dermatology OPD at School of Tropical Medicine, Kolkata with multiple, hyperpigmented, thickened, raised lesion distributed over face, neck, back, abdomen, upper limb along with some hypopigmented flat lesion over upper limb, back, abdomen. The lesions first appeared on back and neck 24 years back and gradually progressive to involve face, neck, trunk, upper limbs. Lesions were stable for last 10 years and were asymptomatic. There was no history of consanguineous marriage and no significant past, personal, family history.
• Multiple, hyperpigmented, dull looking, round to oval, hyperkeratotic plaques with verrucous surface over face and neck (Fig 1,2,3,4)

• Multiple hypopigmented macules over back and arm.(Fig5,6)

Routine investigations were done and were within normal limit. A clinical diagnosis of epidermodysplasia verruciformis was given and a punch biopsy was performed and the tissue was sent for histopathological examination.

Histopathological findings(Fig7,7a,8,8a): Showed uniform acanthosis of the epidermis with hyperkeratosis. Within thickened epidermal layer there are aggregates of basophilic epithelial cells clustered within the epidermis in a nested or whorled arrangement.
A thorough histopathological evaluation established the diagnosis of seborrheic keratosis, {Clonal(nested) type}.

III. Discussion

Seborrheic keratoses show a considerable variety of histologic appearances. Often, more than one type may be observed in the same lesion.

Acanthotic type
This type shows marked acanthosis of predominantly basaloid cells. Moderate papillomatosis and hyperkeratosis are present.[5] Horny invaginations that on cross-sections appear as “pseudo-horn cysts” are numerous. “True horn cysts” are also seen, which show sudden and complete keratinization with a very thin granular layer surrounding it.[4] About one-third of these lesions exhibit melanocytic proliferation and hyperpigmentation.[5] Formation of an in-situ carcinoma within this type, so-called “bowenoid transformation”, is seen occasionally in lesions of sun-exposed areas.[4]

Hyperkeratotic type
Pronounced papillomatosis is present in this variant. Acanthosis is mild but shows a verrucous appearance with elongated projections (“church spire” pattern).[5] There is prominent hyperorthokeratosis. While horn cysts and pseudocysts may be seen, they are less common than in the acanthotic form[5] Hyperpigmentation is unusual in this variant.

Clonal type
The hallmark of the clonal (nested) seborrheic keratosis subtype is the proliferation of sharply demarcated intraepithelial nests of basaloid cells. In some cases, the nests are composed of larger cells with conspicuous intercellular bridges, with nests separated by strand of cells with small dark nuclei.[5]

Reticulated type
The reticulated (or adenoid) type is characterized by numerous, thin, double rows of basaloid epidermal cells, which extend from the epidermis and show branching and interweaving in the dermis.[5] Hyperpigmentation is relatively common, although “horn cysts” and “pseudocysts” are not. There is clinical and histologic evidence of a relationship between solar lentigo and the reticulated subtype of seborrheic keratosis.[4]

Irritated type
Irritated seborrheic keratosis shows a lichenoid inflammatory infiltrate in the dermis and intraepithelial squamous eddies, which are composed of whorling aggregates of eosinophilic squamous cells.[4] In this type, the squamous cells outnumber the basaloid cells.[4] Most eddies appear to show at least one of the morphological features of intraepidermal hair follicle structures. Squamous eddies may be confused with horn pearls of squamous cell carcinoma but can be differentiated from them by their large number, small size and circumscribed configuration.[4]

Pigmented type
Any variant can show pigmentation but is often seen within the acanthotic and reticulated subtypes of seborrheic keratosis. Pigment is present mainly within basal keratinocytes, although in melanocanthoma of skin, a rare type of pigmented seborrheic keratosis, a marked increase in melanocytes containing melanin pigment is seen. The pigmented subtype may be clinically confused with other pigmented lesions, such as malignant melanoma, pigmented basal cell carcinoma or melanocytic nevus.[5]
Differential Diagnosis[3,4]

- Epidermodysplasia verruciformis
- Epidermal Nevus
- Actinic Keratosis
- Verruca vulgaris
- Acanthosis Nigricans
- Basal Cell Carcinoma
- Melanoma.

IV. Conclusion

Seborrheic keratoses are benign lesions. They generally are only of aesthetic concern to the patient. Some lesions may be bothersome because of pruritus and are usually treated for this reason. It may clinically mimic a malignant tumor, thus they are usually biopsied to exclude other tumors. The sign of Leser–Trélat (LT) is the sudden eruption of multiple seborrheic keratoses or increase in the number and size of existing seborrheic keratoses, associated with an underlying malignancy. Thus, patients should be carefully followed up and investigations repeated, especially if the sign progress and/or become more florid. Thorough routine and histopathological examination is necessary to differentiate clonal seborrheic keratosis from pagetoid squamous cell carcinoma in situ.

V. Learning Points

- Although seborrheic keratosis is a disease of elderly it can occur even at a very young age as in this case.
- Seborrheic keratosis may be associated with underlying malignancy, or there may be malignant transformation of these benign lesions, or they may mimic malignant lesions, so regular and repeated routine as well as histopathological examination is a must.
- Clonal seborrheic keratosis is a very rare variant and can be diagnosed only after histopathology and should be differentiated histopathologically from pagetoid squamous cell carcinoma in situ.

References