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Lobulated intradermal nevus: A rare case report

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Abstract

Intradermal nevi are common benign proliferations of uniform melanocytes that are located initially at the dermal-epidermal junction and over time tend to migrate into the dermis and regress with subsequent morphological changes. At the end stage of this process, all the Naevus cells are completely detached from the overlying epidermis and is called intradermal nevus. We report a case of intradermal nevi in a 36 year old female presented to the Dermatology OPD with clusters of grapes like growth over the scalp for the last 5 years and confirmed with clinical and histopathological features.

Keywords: Intradermal nevi, corymbiform nevi, melanocytic nevi, lobulated intradermal nevus, surgical removal

Introduction

Intradermal (or dermal) naevi are skin-coloured, dome shaped nodules or papules that can be greater in size than junctional naevi. Their surface is usually flat but can also appear papillomatous. They are seen primarily on the head, neck and shoulders. One or a small number of hair shafts may project from the naevus surface. 15-30% of melanomas arise in association with already existing naevi, naevi turning malignant is a very rare event^[1, 2]. An intradermal naevus has identical features to the dermal component of a compound naevus, with melanocytes slowly suffer the loss of their ability to produce melanin as they move forward from the upper to the deeper dermis.

Case report

An otherwise healthy 36 year old female presented to the Dermatology OPD with clusters of grapes like growth over the scalp for the last 5 years.

The growth started to occur on a skin lesion that was already present since 3 years of age.

The growth is gradual in onset and progressive in nature. History of itching and pain were denied.

There is no history of bleeding on touch. History of treatment for the same was denied.

On further questioning, the patient revealed having multiple dark coloured skin lesions over the left arm since birth.

Past medical history, family history and drug history were insignificant

On examination, hyperpigmented, lobulated, smooth surfaced, non-tender, soft nodules with a bluish hue present over the left temporal region (Fig 1). Few hair strands seen emerging from the lesion (Fig 2). Two Hyperpigmented velvety plaques embedded with hair seen over the left forearm (Fig 3). Nails were normal. Skin elsewhere normal.

A biopsy was taken and the histopathology examination revealed thinned out epidermis. The dermis showed a pigmented lesion composed of uniform round cells arranged in nests, cords and sheets extending deep down to subcutaneous fat. The cells in the deep dermis are spindle shaped with oval nucleus, no melanin pigment and are exhibiting neurotisation (Fig 4 & 5).

A diagnosis of Lobulated Intradermal Nevus with Neurotization was made after the clinical and histopathological examination.

The patient was reassured about the condition and was referred to Plastic surgery for further management.



Fig 1: Hyper pigmented, lobulated, smooth surfaced, non-tender, soft nodules with a bluish hue present over the left temporal region



Fig 2: Few hair strands seen emerging from the lobulated lesion



Fig 3: Two Hyper pigmented velvety plaques embedded with hair seen over the left forearm

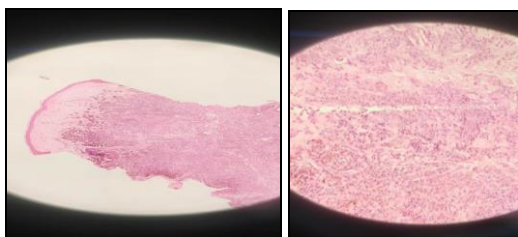


Fig 4 & 5: Histopathological examination revealed thinned out epidermis with dermis showing a pigmented lesion composed of uniform round cells arranged in nests, cords and sheets extending deep down to subcutaneous fat. The cells in the deep dermis are spindle shaped with oval nucleus, no melanin pigment and are exhibiting neurotisation

Discussion

Melanocytic nevi are benign neoplastic proliferations consisting of melanocytes. If they proliferate at the dermo-epidermal junction known as junctional naevus and over a period of time they slide down to dermis while one of the component being attached with the basal layer known as compound naevus.

Clinically, different types of melanocytic nevi have been identified, namely smooth or flat lesions, papillomatous, dome shaped plaques and pedunculated lesions. When nevi cells occupy the dermis alone, with absence of junctional cavity the term intradermal nevus has been administered [1]. The predisposing factors of the naevus include individuals with Turner syndrome, history of atopy, chemotherapy given for leukemia in childhood or involving organ transplantation individuals. Studies have shown that sunlight is associated with increased naevus cases in children. Genetics also play a role in intradermal naevus like involvement of somatic BRAF mutations which is indicative of activation of RAS/MAPK kinase pathway in the naevogenesis [2, 3].

One of the type of lobulated intradermal nevus which is specifically characterised by aggregation of lobules depicting a bunch of grapes is known as Corymbiform nevi, which was first coined by Löffler and Effendy [7].

Remarkably, all the cases presented in the past including ours was seen in females, we are not sure of whether there appears to be a relationship in consider to female hormones which is yet to be clarified. One of the characteristic feature that bear a resemblance to our case was the age of presentation which was mainly in the age group varying from 32 to 40 years. Other characteristic features were presence of skin colored lobules as well as pigmented lobules that were distributed in multiple sizes throughout the size of the nevi, with some of them exhibiting comedo-like plugging. In our case, this u feature of comedo-like plugs in corymbiform nevi is an distinctive feature and hence being reported for the same [3, 4].

Intradermal nevi are raised, fleshy and moderately pigmented papules or plaques. Lesions differ in size from a few millimeters to a centimeter. Intradermal nevi are reddish brown or black, but after certain period become lighter or skin-colored with time. These are seen in a vast morphologic variations, including flat or smooth lesions, slightly raised lesions often with a elevated center and a flat periphery, papillomatous lesions, dome-shaped papules and pedunculated lesions. The different morphology like shape, size or color of the lesion mull over the transformative process in which the nevi extend downward with increasing age and nevus cells degenerate or they get replaced by collagen, fat, and fibrous tissue. Melanocytic nevi are conditioned to change with increasing age in both clinical and histopathologic findings [4, 5].

"Lobulated intradermal nevus" was the name given by Cho *et al.* who suggested that their cases showed an specific form of regressing melanocytic nevus as lipomatous changes which are generally seen in a reverting intradermal nevus in patients older than 50 years and that these changes in young to middle aged females is a rare entity [6].

Our case is identical with cases described by Cho *et al.* in 1991 and others mentioned in the literature.

The lesions clinically showed lobulation and histologically showed well-known fat cell infiltration within the nests of nevus cells, neuroid differentiation of nevus cells is clearly seen and can see presence of dermal fibrosis. Presence of

lobulations in an intradermal nevus generally indicates the aging of the melanocytic nevus but not necessarily in all cases.

This condition needs to be distinguished from other similar-looking nevi entities, such as nevus lipomatous superficialis and cerebriform nevus, based on the clinical and histopathologic findings^[5].

Nevus lipomatous superficialis histopathological picture shows strands of fat cells and it is most commonly seen over the lower back.

It doesn't show any aggregation of nevus cells.

Cerebriform nevus exhibits single or collection of nevus cells having multiple amounts of melanin and neuroid transformation but does not exhibit fatty degeneration or fibrosis^[7].

Generally most of the melanocytic nevi regress eventually and the naevi number also decreases with increasing age.

No treatment is required as such for these cases. Surgical removal is performed only for cosmetic concern of the patient, sometimes it has the potential risk of scarring, post-inflammatory hyper or hypopigmentation with the surgical removal.

In conclusion we agree to the former opinion and suggest that lobulation is a rare clinical manifestation which is associated with common Histopathologic findings of aging melanocytic nevi.

And we also think that lobulated intradermal nevi are both clinically and histopathologically heterogeneous entities. Their clinical and histopathologic changes are variable in degree but on a collinear spectrum.

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