EUROPEAN JOURNAL OF BIOMEDICAL AND PHARMACEUTICAL SCIENCES
http://www.ejbps.com

ISSN 2349-8870
Volume: 8
Issue: 3
121-127
Year: 2021

SPECTRUM OF DIFFERENT SKIN LESIONS AT VARIOUS STIES- A SHORT SERIES OF CASE REPORTS

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Article Received on 31/12/2020 **Article Revised on 21/01/2021** **Article Accepted on 10/02/2021**

ABSTRACT
The spectrum of skin disease vary according to age, gender and other factors. Skin biopsies are needed to establish diagnosis. There are various skin lesions having similar clinical manifestations, hence histopathological examination is necessary for diagnosis and treatment. In the present article, various case reports of skin lesions occurring at various sites have been described with clinical and histopathological diagnosis.

KEYWORDS: Various skin lesions, histopathology, diagnosis.

INTRODUCTION

Skin is the largest sensory organ of the human body, comprising of epidermis, dermis, and subcutis. It is a complex organ with multiple functions, mainly acts as a barrier against various harmful environmental agents, and also maintains homeostasis. It is most exposed to ultraviolet rays of sunlight and is susceptible to a wide spectrum of disorders ranging from inflammatory conditions to neoplastic lesions. These skin disorders usually arise from the normal histological constituents of the skin.

Skin disorders are influenced by multiple factors such as environment, economy, literacy, region, ethnic groups, genetic factors and social customs. Occasionally it may be a manifestation of systemic diseases. Pattern of skin diseases varies from country to country and even region to region within a country. Majority of skin lesions are diagnosed by clinical history but some lesions require histopathological examination mandatory for diagnosis and treatment.

CASE No. 1
DOWLING DEGOS DISEASE

A 42 year old male patient presented with multiple hyperpigmented macular lesions over face, neck and axillary region with presence of pits since 4-5 years. Biopsy was taken from lower axillary region and sent for HPE. H&E section showed small horn cysts in the epidermis with thin epithelial strands extending into superficial dermis from epidermis resulting in antler-like pattern. Dermal perivascular lymphocytic infiltrate was present (Fig 1,2). The histological diagnosis of Dowling Degos disease was made.

Discussion

Dowling Degos disease is a rare reticulate pigmentary disorder inherited as autosomal dominant trait described by Dowling and Freudenthal characterized by numerous asymptomatic, symmetrical, progressive, small, round, pigmented macules over groin, face, neck, arms and trunk, scattered comedo-like lesions and pitted acneiform scars.[1] Most common presentation is hyperpigmented macules in a reticulate pattern in the flexures. Associated features include comedo like papules, perioral pitted scars, epidermoid/ trichilemmal cysts.[2] Histopathology is diagnostic.

3 genes have been shown to be associated with Dowling Degos disease. Flexural Dowling Degos disease was caused by loss of mutations affecting the KRT-5 gene region encoding the initial part of keratin.[3] Generalized Dowling Degos disease was found to be associated with mutation in POFUT1, which encodes protein O-fucosyltransferase and POGLUT1 which encodes for protein O-glucosyltransferase.[4]

There seems to be no definite cure for Dowling Degos disease, many treatment options like depigmenting agents such as hydroquinone, systemic and topical retinoids, lasers have been beneficial to some extent.[5]

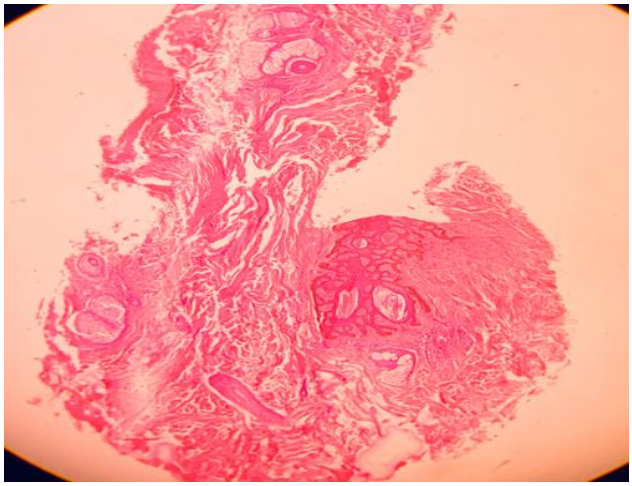

Fig 1. Dowling Degos disease, H&E 10X.

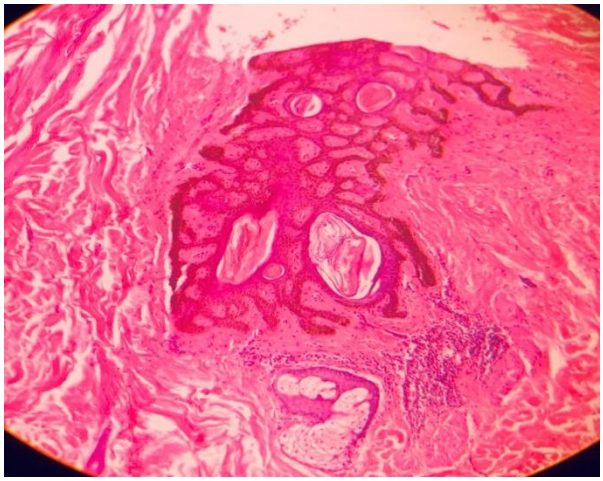

Fig 2. Dowling Degos disease, H&E 40X, Horn cysts in epidermis with antler -like pattern.

CASE No. 2
MACULAR AMYLOIDOSIS

A 45 year old female presented with hyperpigmented macular patches on back with pruritus. Initially it started with pinpoint dark spots which later coalesced to form macular lesion measuring 4x3 cm, increasing gradually in size (Fig 3). Extensor surfaces of both forearms were also involved. Biopsy was taken and submitted for HPE. H&E section showed mild thinning of epidermis and mild hyperkeratosis. Focal disruption of the basal layer with increase in melanophages was seen, perivascular lymphocytic infiltrate. Congo red stain showed presence of amyloid deposits in subepidermal zone. CO2 laser and local steroid was used alongwith homeopathy treatment but with no good results.

Discussion

Macular Amyloidosis also known as Friction Amyloidosis, is the most subtle form of cutaneous amyloidosis, characterized by greyish -brown pruritic macules in a rippled pattern, which coalesce into patches.[6] It is most commonly seen in the interscapular area, extremities (shin and forearm). Also seen in clavicles, breast, face, neck and axilla.[7] Risk factors such as race (high incidence in Asians, Middle East and South America) but rarely in European and North America. Most cases are sporadic, female preponderance with F:M ratio 7.3 :1, Patients occasionally complain of pruritus. Sometimes pruritus is minimal to moderate and occasionally severe, mainly in high humidity. Mechanical trauma induced by nylon fibres and bristles, exposure to ultraviolet rays could be etiological factor.

There are two theories explaining the origin of amyloid protein in the skin – fibrillary body theory and secretory theory. The fibrillary body theory states that damaged keratinocytes undergo filamentous degeneration by apoptosis and transformation by dermal fibroblasts and histiocytes and are converted into amyloid which deposits in the papillary dermis.[8] Secretion theory describes the deposition of amyloid from the degenerated basal keratinocytes at the dermoepidermal junction which drops into the papillary dermis through the damaged lamina densa of basal layer.[9]

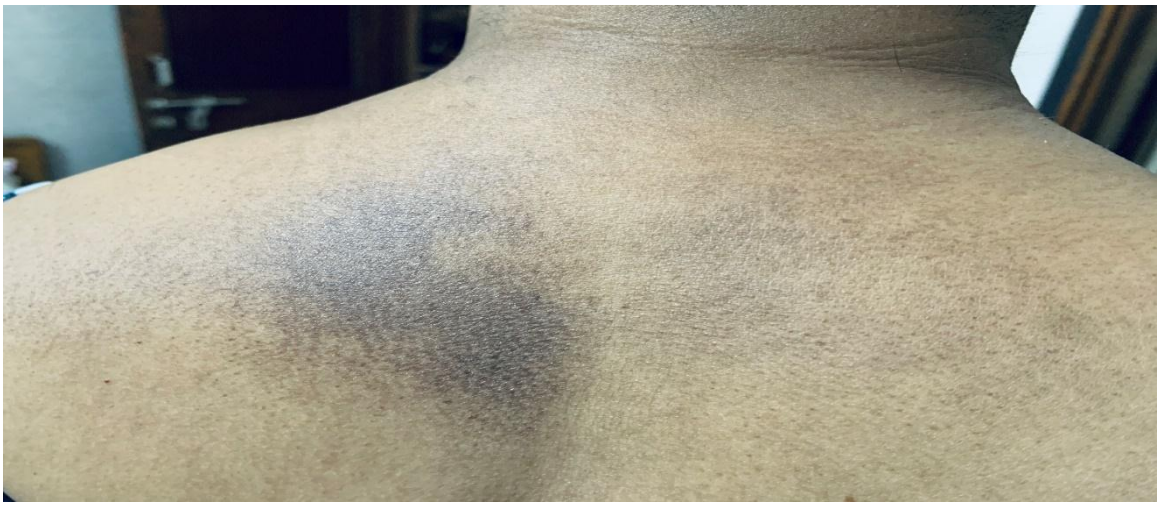

Fig 3: Macular amyloidosis.

CASE NO. 3
ICHTHYOSIS VULGARIS
A 42 year old male presented with dry scaly skin over buttock with exacerbation during winter season since 4 years. Skin biopsy was taken from buttock and sent for HPE. H&E section showed increase in stratum corneum, at place or absent granular layer with mild inflammation. Histological diagnosis of Ichthyosis vulgaris was made.

Discussion
Ichthyosis Vulgaris / Fish scale disease is an inherited skin disorder in which dead skin cells accumulate in thick, dry scale on your skin's surface, can present at birth, usually appear during early childhood. It is autosomal dominant disease, worsens in cold, dry environment, appears on elbows and lower legs.

CASE NO. 4
DIFFUSE ALOPECIA AREATA
A 21 year old female presented with ill defined diffuse hair loss over entire scalp since 4-5 years . Biopsy from fronto parietal region was taken and sent for HPE (Fig 5). H&E section showed thinning of keratinized epidermis. Underlying dermis showed pilo sebaceous glands. Few mono nuclear inflammatory cells are seen in peribulbar region. Miniaturized follicles are seen in superficial dermis (Fig 6,7). The histological diagnosis of Diffuse Alopecia areata (chronic stage) was made.

Discussion
Diffuse alopecia is common complaint mainly affecting females, due to female androgenetic alopecia, telogen effluvium, diffuse alopecia areata.

Telogen effluvium- begins with a sudden increase in hair loss and maintenance of frontal hair density (10). Surgery, fever, childbirth, iron deficiency, stress,chronic disease, dietary changes esp. protein and iron restrictions are the triggering factors. Microscopically total number of follicles and the terminal : villus (T:V) ratio does not change in chronic TE (normal 7:1).[11]

In diffuse alopecia areata, hair thinning is subtly distributed throughout the scalp triggered by psychological stress or systemic diseases.[12] Histologically there are three stages : acute, subacute and chronic.

In acute stage, the first histological sign is a lymphocytic infiltrate around the bulb of the terminal hair located in the hypodermis, necrosis of matrix cells in the hair bulb and presence of remains of melanin within the follicular epithelium due to necrosis, transformation of rounded shape of hair bulb to golf-club shape.[13], In subacute stage, there is increase in catagen follicles, telogen germinal units show distortion or dystrophy, prominent follicular tracts, with lymphocytic inflammatory infiltrate with absence of peribulbar inflammation.[14]

In chronic stage, there is alopecia totalis and universalis with inversion of T:V ratio, large numbers of miniaturized hairs (nanogen), lymphocytes around the miniaturized follicles, melanophages.[14]

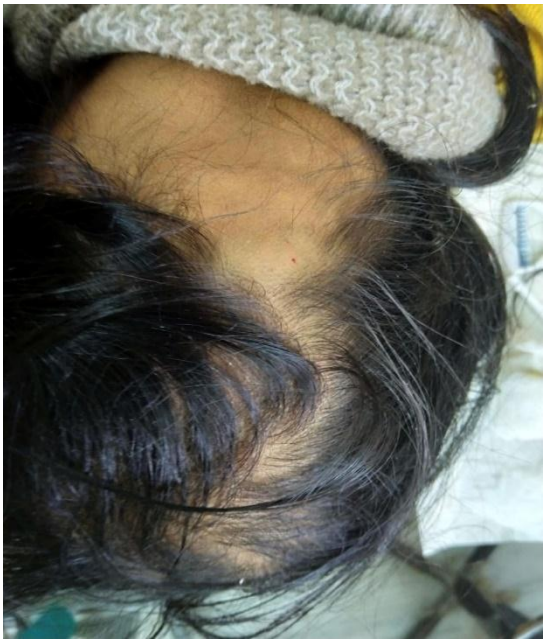

Fig 5. Clinical photo of diffuse alopecia areata.

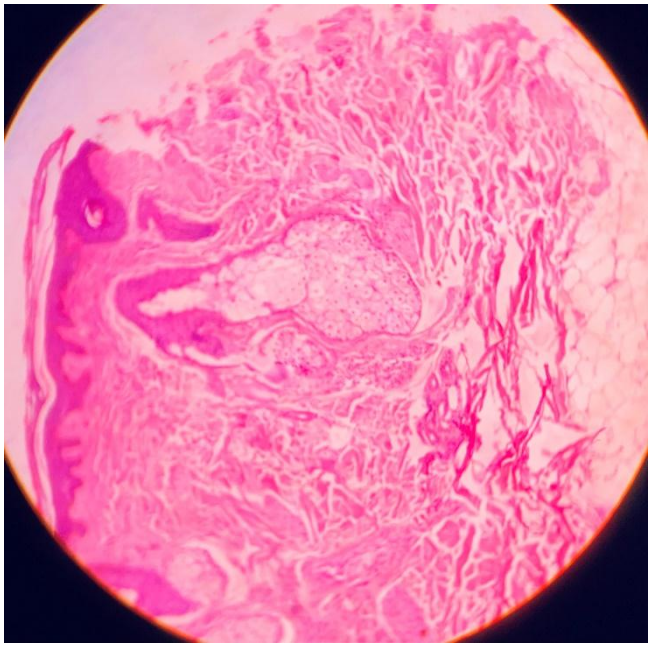

Fig 6. Diffuse alopecia areata, H&E stain, 10 X, Thinningof epidermis, miniaturized hair follicles in superfical dermis, pilosebaceous glands

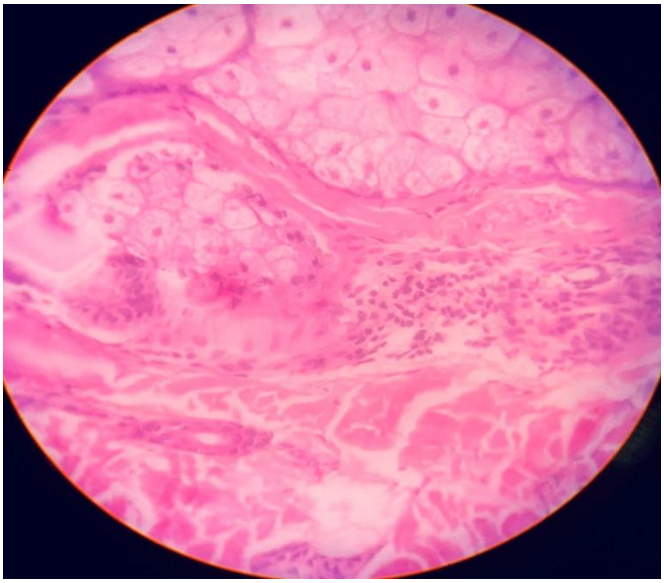

Fig 7. Diffuse alopecia areata, H&E stain 40X, Mononuclear infiltration of peribulbar region.

CASE NO. 5
GENITAL SYRINGOMA

A 26 year old female presented with multiple skin covered brownish papule of various size present over labia majora and minora with history of pruritus since 3 months. Biopsy was taken from lesion over labia minora and sent for HPE. H&E section showed epidermis with underlying numerous small ducts lined by two layers of epithelial cells embedded in fibrous stroma. Some of these ducts showed small comma like tails of epithelial cells imparting them a tadpole shape. Lumina of some of the ducts contained amorphous debris (fig 8). Histopathological diagnosis of Syringoma of Labia Minora was made.

Discussion

Syringoma is a benign adnexal tumor, first described by Kaposi & Biesiadeki in 1872.[15] originated from intraepidermal eccrine sweat glands.[16] It is commonly seen in adolescent female involving face and chest, thigh, axilla, abdomen. Genitalia being very rare site. Genital syringoma are usually asymptomatic but can present with pruritus. It can undergo cyclic changes in size and symptoms during premenstrual period, pregnancy and use of oral contraceptive, thus indicating hormonal influence of estrogen and progesterone.[17] Syringoma occurs sporadically or rarely has hereditary etiology. It is more common in patients with Marfan, Down and Ehler-Danlos syndrome.[17] Thus genital syringoma should be considered in the differential diagnosis of pruritus vulvae.

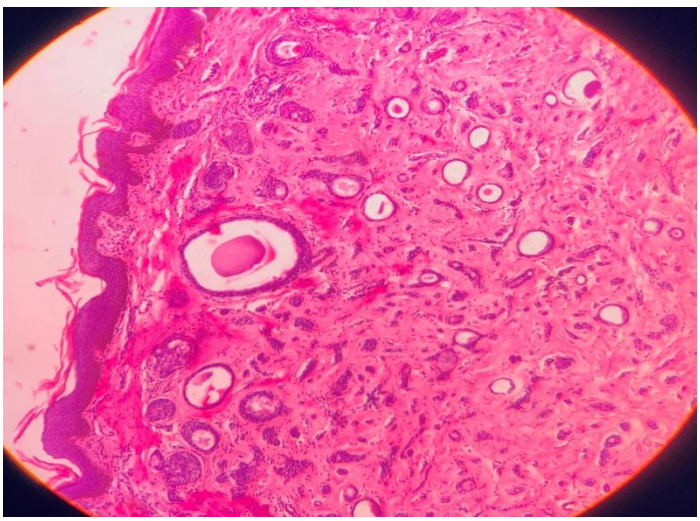

Fig 8. Genital syringoma, H&E stain, 40 X.

CASE NO 6
CEREBRIFORM NEVUS SEBACEOUS

A 12 year old female child with 3-4 well defined yellowish verrucous plaque with cerebriform pattern of size 7x4 cm (largest) presented over right temporal area of scalp since birth. Clinical diagnosis of cerebriform nevus sebaceous and verrucous epidermal nevus were made. Biopsy from right temporal area of scalp was

taken and submitted for HPE. H&E section showed acanthosis, hyperkeratosis, papillomatosis and follicular plugging in epidermis. Upper dermis showed under developed hair follicle with mature sebaceous glands (Fig 9). Histopathological diagnosis of Cerebriform nevus sebaceous was made.

Discussion

Cerebriform type of nevus sebaceous is very rare morphological variant of Nevus sebaceous of Jadassohn, an epidermal nevus. It is predominantly congenital sebaceous hamartoma with 0.3 % incidence in neonates. It is commonly seen in head and neck region as solitary lesion. Other common sites are scalp, forehead, centrofacial, periauricular and genital area. It presents at birth as single hairless yellowish plaque with smooth velvety surface, becomes verrucous and nodular at puberty indicate role of hormones. It usually occurs sporadically, but autosomal dominant transmission is seen in some reports.[18]

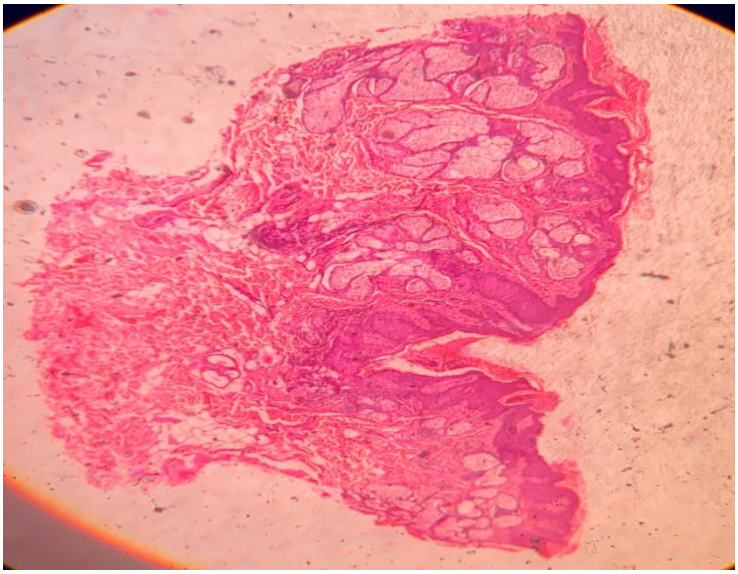

Fig 9. Cerebriform nevus sebaceous, H&E stain, 10 X.

CASE NO. 7

PEMPHIGUS VEGETANS

A 30 year old female presented with multiple painless indurated lesions over genitalia. She was under treatment for Pemphigus Vulgaris with dexamethasone pulse treatment. She also had multiple painful oral erosions. Clinical diagnosis of primary chancre or pemphigus vegetans was made and biopsy from labia majora was taken and submitted for HPE. H&E section showed hyperkeratosis, acnathosis, papillomatosis. Suprabasal blister was seen which was filled with predominantly eosinophil rich infiltrate forming microabscess in epidermis. Upper dermis show dense infiltration predominantly by eosinophils. (Fig 10,11). Histopathological diagnosis of Pemphigus Vegetans was made.

Discussion

Pemphigus is a group of Vesiculobullous autoimmune disease, most common is Pemphigus vulgaris and rarest variant is Pemphigus vegetans, representing 1-2 % of all pemphigus.[19] It is more common in female aged 30-50 years. Pemphigus vegetans forms vegetative plaques in intertriginous areas and oral mucosa. There are two forms of pemphigus vegetans, the Hallopean type and Neumann type. The Hallopeau type has an indolent course characterized by pustules and heals as vegetative plaques. Histologically it has strong eosinophilic response with eosinophilic spongiosis, intraepidermal eosinophilic microabscesses, and dense eosinophilic dermal infiltrate. The Neumann type is more severe, vegetations develop during an eruption of vesiculobullous lesions, oral mucosa is usually involved. Histologically, in this type neutrophils and lymphocytes accompany the eosinophilic response. There may be papillary dermis edema.[20] These hyperkeratotic lesions present in the intertriginous areas including groin / inguinal folds, armpits, thighs and flexural surfaces.[21] Less common areas are scalp, soles of the feet, skin graft site. It can also be seen in nasal, vaginal and conjunctiva.[22] There is association between pemphigus vulgaris and HLA-DRB1 and HLA-DQB1, thus suggesting genetic susceptibility. Also there is a hypothesis that B cells with VH1-46 heavy chain gene may be hypersensitive to desmoglein-3 increasing cellular damage.[23]

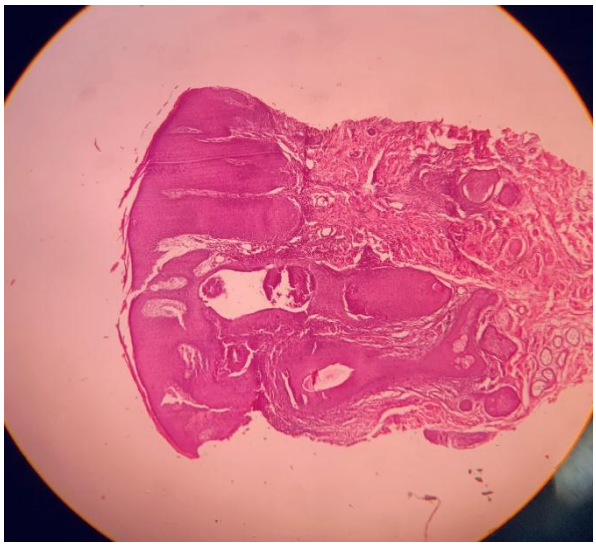

Fig 10: Pemphigus Vegetans, H&E, 10X

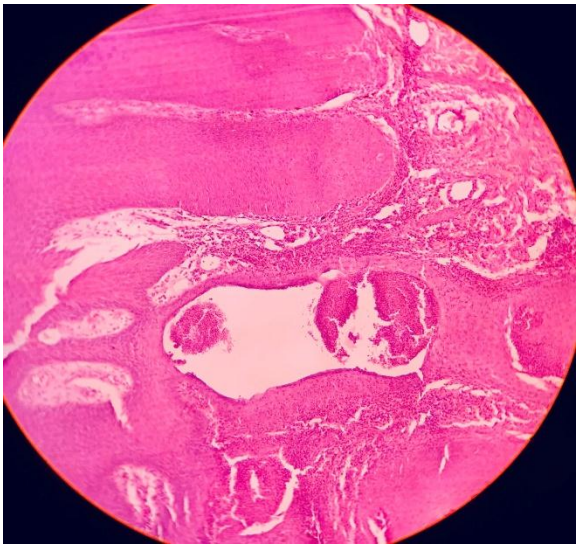

Fig 11: Pemphigus Vegetans, eosinophilic microabscess in epidermis

CASE NO. 8
ECCRINE ACROSPIROMA

A 23 year old female presented with nodular swelling 2x2 cm, firm, non tender, non motile over right elbow. Swelling was excised completely and submitted for HPE. Grossly it was lineated from the surrounding tissue and not encapsulated. Histologically, it was circumscribed, multilobular with epithelial cells in upper and middle dermis. The tumor was composed of biphasic epithelial cells surrounding stroma. Cells were round, fusiform, polyhedral. Cytoplasm scanty, oval nucleus, indistinct cell boundary, no mitotic figures seen (Fig 12). Histopathological diagnosis of Eccrine Acrospiroma was made.

Discussion

Eccrine acrospiroma are benign tumors of sweat gland origin. Tumors occur anywhere on the body, diameter measuring from 0.5 to 10.5 cm. Skin over the tumor is flesh coloured to red to blue, sometimes thickened.[24] It can occur on all areas of body, elbow is less common site. Males are affected more than females.

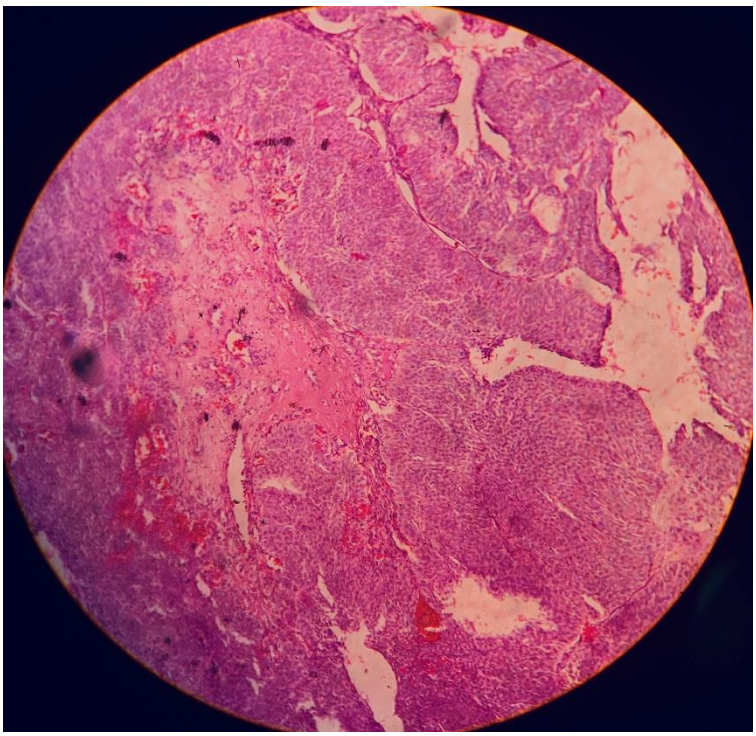

Fig 12: Eccrine acrospiroma, H&E stain, 10 X.

ACKNOWLEDGEMENT: Patient and her relatives.

SOURCE OF FUNDING: None.

CONFLICT OF INTEREST: None.

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