

## 1. Answer: C

Atypical fibroxanthomas typically present on sun-exposed skin of the head and neck in older individuals. Areas of prior radiation exposure and individuals with a history of prior transplant may also be prone to developing these tumors. These tumors present as rapidly growing solitary red nodules with or without ulceration, clinically often confused with squamous cell carcinomas or pyogenic granulomas and rarely melanoma. Atypical fibroxanthomas, first described by Helwig in 1963, are generally regarded as a more superficial and indolent form of malignant fibrous histiocytoma.

There is no single immunohistochemical marker that defines atypical fibroxanthomas. Although CD68, CD99 and vimentin are typically positive, these are neither absolutely sensitive nor specific. Despite this, numerous markers must be applied to suspect atypical fibroxanthomas to reliably rule out other spindle cell neoplasm. These typically include HMB-45 or MART-1, cytokeratins, and CD31 to rule out melanoma, spindle squamous cell carcinoma, and angiosarcoma respectively. Exclusion of leiomyosarcoma may be complicated as focal SMA positivity is commonly observed in AFX. The use of HMB-45 and MART-1 to rule out melanoma may be problematic in cases of AFX with osteoclast-like giant cells, in which HMB-45 and MART-1 have been reported to be positive

### Reference:

1. Perrett CM. Cerio R. Proby CM. Harwood CA. Atypical fibroxanthoma in a renal transplant recipient. *Histopathol* 2005;47(3):326-7
2. Diaz-Cascajo C. Weyers W. Borghi S. Pigmented atypical fibroxanthoma: a tumor that may be easily mistaken for malignant melanoma. *Am J Dermatopathol* 2003;25(1):1-5
3. Monteagudo C. Calduch L. Navarro S. Joan-Figueroa A. Llombart-Bosch A. CD99 immunoreactivity in atypical fibroxanthoma: a common feature of diagnostic value. *Am J Clin Pathol* 2002;117(1):126-31
4. Crowson AN. Carlson-Sweet K. Macinnis C. Taylor JR. Battaglia T. LaMar WL. Minor D. Sutter S. Hill T. Clear cell atypical fibroxanthoma: a clinicopathologic study. *J Cutan Pathol* 2002;29(6):374-81
5. Huether MJ. Zitelli JA. Brodland DG. Mohs micrographic surgery for the treatment of spindle cell tumors of the skin. *J Amer Acad Dermatol* 2001;44(4):656-9

## 2. Answer: B- Juvenile Xanthogranuloma

**Clinical Presentation:** Juvenile xanthogranulomas (JXG) represent the most common non-Langerhans cell histiocytosis. The disease typically affects children with a median age of onset of 2 years. The lesions tend to be solitary but may be multiple with a predilection for the head and neck. When occurring in younger children, they tend to be multiple and have a significant male predominance. Clinically the lesions present as reddish brown papules or nodules typically less than 1-2 cm in diameter. As the lesions age, there is a tendency to become more yellowish. 4% of JXG patients may have a systemic form in which the liver, spleen, lungs, CNS and eyes may be affected. When affecting the eyes, JXG may cause spontaneous hyphenate

**Histological:** JXG appears as a circumscribed nodular infiltrate of the papillary and reticular dermis composed of a dense population of histiocytes. Accompanying inflammatory cells including lymphocytes, plasma cells, and rare neutrophils can be seen in the peri-lesional or perivascular areas. The characteristic cell found in the lesion is the Touton giant cell. The cell consists of a multinucleate giant cell with the nuclei forming a ring surrounding a central eosinophilic amorphous center, the nuclei in turn are surrounded by a pale, xanthomatized foamy rim of cytoplasm. These cells may predominate in more recent lesions

The diagnosis of Spitz nevus is typically dependent upon the observation of vertically arranged nests of spindle cells at the dermoepidermo-junction. These nests are often seen in the presence of Kamino bodies; eosinophilic globules located over spindle cell nests at the dermoepidermo-junction. Touton giant cells are not a histologic feature of Spitz nevi

The diagnosis of eruptive xanthoma is difficult to exclude, however the presence of Touton giant cells is not compatible with the diagnosis of eruptive xanthoma. In addition, eruptive xanthomas occur in large number in a widespread distribution and typically occur in older individuals.

Langerhans cell histiocytosis, although sharing the histiocytic appearance of the cells, typically has associated infiltrating eosinophils. In addition, the clinical picture of Langerhans cell histiocytosis is typically one of an erythematous, scaling, seborrheic dermatitis-like rash affecting the intertriginous areas. Touton giant cells are also not a feature of Langerhans cell histiocytosis.

3. Answer: A, Oxalosis

Cutaneous oxalosis is a very rare occurrence, but may occur after ingestion of significant quantities of automotive anti-freeze (Polyethylene glycol). Its presentation in the skin is unique in that it is the only intra-vascular deposition which polarizes. Cutaneous oxalosis may also be seen in cases of Celiac disease and Cohn's disease. Renal failure is a common manifestation of oxalosis.

Although calciphylaxis bears a striking similarity of oxalosis by H&E stain, the presence of polarizable crystals rules this out. As calciphylaxis is often seen in the setting of renal failure, care must be taken to differentiate these two entities  
There is no such thing as embolic gout

Thrombosis cutis medicamentosa is an uncommon cutaneous manifestation of improper subcutaneous injection technique, typically seen in the administration of depot-medrol or long acting antipsychotics or vitamin K. The medication is directly injected into a vascular space, resulting in a hypertonic shock to the vessel wall, with subsequent thrombosis. The histologic picture of this entity is essentially that of common vascular thrombosis.

4. Answer: C-Inflammatory breast carcinoma

Cutaneous mastocytosis/mastocytoma is classically well demarcated papules and nodules which never form plaques. Typically cutaneous mastocytomas will urticate upon rubbing. Mastocytomas classically arise in younger individuals. The cells within the dermis demonstrate significant pleomorphism, which is not a feature of mastocytoma.

Leukemia cutis shares many features with cutaneous mastocytosis, in that it presents as papules and nodules and never as widespread plaques, however the infiltrate is composed of malignant hematopoietic elements. The images shown show tumor cells forming cords and nests with poorly formed glandular structures

Although inflammatory breast carcinoma in males is uncommon, it remains an important diagnosis to recognize. Clinically, the lesions present as indurated, erythematous, variably scaling plaques with well defined borders. When the inflammatory condition encompasses the entire chest wall, the term carcinoma en curasse may be utilized. Histologically the infiltrating cells form cords and nests and form pseudoglandular structures. Nests of tumor cells may be seen occluding lymphatic structures.

Large cell transformed MF may present as plaques, and such cells are often large with significant pleomorphism, however there is typically a history of long-standing mycosis fungoides in these patients. In addition, the cells of MF are never seen forming the pseudo-glandular structures that are evident in this case  
There is no such entity as segmental lamellar ichthyosis

5. Answer: D-Follicular mucinosis

Alopecia areata is an idiopathic condition typified by non-scarring, inflammatory alopecia. Clinically the disease presents as asymptomatic well demarcated areas of hair loss with retention of the follicular orifices. Histologically, alopecia areata shows peribulbar lymphocytic infiltrates sometimes likened to a "swarm of bees". Trichotillomania is a form of compulsive disorder often described in children. The condition results in compulsive pulling and plucking of hairs. Clinically, trichotillomania can resemble alopecia areata, however in trichotillomania, minute hair shafts within the follicular orifices may be observed. These regrown hairs are too small to be manually plucked. Alopecia areata lacks these hairs. Histologically, trichotillomania shows pigmented casts within the follicles, termed trichomalacia. Folliculitis decalvans is an idiopathic scarring inflammatory alopecia typified by acute and chronic inflammatory infiltrates and dermal fibrosis.

Follicular mucinosis is an inflammatory non-scarring alopecia typified by distention of the follicle by mucinous deposition. There is a well established association between follicular mucinosis and mycosis fungoides, and some believe that follicular mucinosis may represent a distinct form of cutaneous T cell lymphoma. This theory is countered by the fact that many cases of follicular mucinosis occur in children and are self limited and never eventuate into lymphoma of any kind.

## 6. Answer: D- Sclerotic fibroma

Dermatofibrosarcoma protuberans is a slow growing, locally aggressive neoplasm typically occurring on the trunk and proximal extremities. Histologically, these tumors show a bland storiform arrangement of plump spindle cells with a minimum of pleomorphism or mitoses. The spindle cells infiltrate through the subcutis and surrounding adnexal structures. Necrobiotic xanthogranuloma is an uncommon cutaneous condition typically associated with underlying gammopathy. It presents with verruciform yellow to orange plaques and is histologically typified by cholesterol clefts, Touton giant cells and zone of necrobiotic collagen.

Neurothekoma or myxoid nerve sheath tumor is an uncommon tumor, which demonstrates lobules of delicate spindle cells in a reminiscent storiform pattern embedded within a myxoid stroma. The prominent sclerosis evident in this case is not a feature of neurothekoma. Pleomorphic fibromas closely resemble soft fibromas in overall architecture, however are distinguished by the presence of large stellate atypical appearing fibroblastic cells.

Sclerotic fibromas are a benign soft tissue tumor of uncertain etiology. The prevailing opinion regarding these tumors is that they represent a morphologic variant of dermatofibromas. This idea has been challenged by some case reports of sclerotic fibromas recurring as sclerotic fibromas at sites of previous incomplete excision, suggesting that the tumors represent distinct lesions, unassociated with dermatofibromas. Histologically these lesions have a striking “wood grain” appearance with storiform bundles of collagen with interspersed bland fibroblastic cells. Sclerotic fibromas have been reported in association with Cowden’s disease.

## 7. Answer: A- Glomangioma

Targetoid hemosiderotic hemangioma, also known as hobnail hemangioma typically presents as a dark brown to violaceous nodules. Histologically, these lesions demonstrate a proliferation of ectatic thin walled vessels lined by hobnail endothelial cells. An associated lymphocytic inflammatory infiltrate is commonly seen in association, as is the presence of hemosiderin. Similarities to entities such as Kaposi’s sarcoma may be striking.

Kaposi’s sarcoma is a low-grade malignant vascular neoplasm associated with HHV8. Histologically, Kaposi’s demonstrates a proliferation of bland appearing spindle cells interspersed with thin, slit-like vascular spaces. Hemorrhage and hemosiderin deposition are often seen. An inflammatory infiltrate, dense in plasma cells is often present. Fragments of red blood cells may often be seen within the cytoplasm of swollen endothelial cells.

Glomangiomas are uncommon dermal tumors comprised of the rare population of vascular associated modified smooth muscle cells called glomus cells. These cells, responsible for cutaneous thermoregulation via dilation or constriction of superficial dermal arterioles. Clinically, the lesions present as solitary or multiple red to purple soft nodules. The lesions may on occasion be painful, owing to thrombosis of the delicate vascular spaces. Histologically, glomus cells have bland round to oval nuclei with granular chromatin and indistinct nucleoli and indistinct eosinophilic cytoplasm. Mitotic figures and atypia are not evident. The presence of mitoses, atypia and necrosis should arise the suspicion of glomangiosarcoma.

Hidradenoma papilliferum is a benign tumor of the eccrine/apocrine glands. Typically identified on the anogenital region of females, the histology consists of a well circumscribed cystic cavity filled by delicate papillary folds lined with single to multiple columnar to cuboidal cells.

Hemangiopericytomas represent a somewhat poorly defined group of tumors which are typified by proliferations of pericytic cells demonstrating a very characteristic branching “stag-horn” vascular pattern. As this entity is further investigated, it is becoming apparent that the term hemangiopericytoma encompasses other entities such as myopericytomas.

8. Answer: C- Exogenous ochronosis

Exogenous ochronosis is an uncommon reaction to prolonged exposure to highly bleaching agents, typically containing hydroquinones. Although uncommon in the US secondary to FDA regulation of the length of use and concentration of quinones within topical preparations, the use and concentrations of the drug are not regulated in other countries. The mechanism by which the pigment is formed is unclear however is thought that hydroquinone inhibits the function of homogentisic acid oxidase leading to an accumulation of homogentisic acid in the dermis which then polymerizes to form the ochronotic pigment. Histologically, exogenous ochronosis shows yellow to brown discolored and thickened elastic fibers.

Onchocerciasis and chromoblastomycosis are both infectious diseases and although cutaneous onchoceromas may show delicate curved organisms in the superficial papillary dermis, there is also an accompanying inflammatory infiltrate.

Mal de Meleda is a rare mutilating palmo-plantar keratoderma lacking any distinct histopathologic changes.

Henna tattoos are hyperpigmented tattoos that are the result of the application of Henna extract, often doped with para-phenylenediamine ( a common black hair dye). The resulting pigment deposition is entirely limited to the stratum corneum, and does not extend into the dermis. Permanent tattooing has resulted from individuals developing severe, acute allergic dermatitis to lawsone (2-hydroxyl-1,4-naphthoquinone), the naturally occurring pigment in henna.

9. Answer-B: Dermatofibrosarcoma protuberans

Low grade fibromyxoid sarcoma is a rare soft tissue neoplasm typified by bland, desmoid-like fibrous stroma with abrupt intervening areas of loose myxoid stroma. The cellularity is less pronounced compared to DFSP, and typically has a more delicate loose vascular pattern.

Dermatofibrosarcoma protuberans is a rare soft tissue tumor that demonstrates a locally aggressive behavior pattern but with little metastatic potential. Clinically DFSPs present as slightly erythematous plaque-like dermal nodules that may eventuate into large fixed, firm, multinodular tumors with extension into the subcutis. The tumors are nonpainful and rarely ulcerate. Fibrosarcomatous or malignant fibrous histiocytoma-like areas within the tumors have been reported in 10-15% of lesions. Whether this degeneration affects the overall behavior of the tumor is debated. The histology of dermatofibrosarcoma protuberans is characterized by a dermal proliferation of bland monomorphic spindle cells typically arranged in a whorled and storiform pattern. The tumor is atypical in that the absence of numerous mitoses, nuclear atypia, and necrosis belie its malignant potential. Often the spindle cell proliferation can be seen dissecting through subcuticular fat. In 5% of cases the tumor may be associated with a proliferation of melanin containing spindle cells. These pigmented DFSP tumors are referred to as Bednar tumors. Dermatofibrosarcoma protuberans is typically distinguished from other spindle cell neoplasms by its positive staining for CD34, a human hematopoietic progenitor cell antigen. This is in contrast to dermatofibromas in which the spindle cells stain positively for factor XIIIa and negatively for CD34. There are rare case reports of CD34 negative DFSP reported in the literature; however, these cases are thought to represent less than 10-20% of reported cases. It has been well documented that DFSP tumors which have undergone fibrosarcomatous transformation often lose CD34 positivity. Isolated cases of DFSP occurring in the setting of nuchal-type fibromas, spindle cell lipomas, and breast carcinoma have been reported. The differential diagnosis of DFSP is broad and includes the majority of the spindle cell neoplasms. The most common mimic of DFSP is the dermatofibroma. Other tumors which may simulate DFSP are diffuse neurofibromas, nodular fasciitis, nerve sheath tumors, leiomyomas and low-grade fibrosarcomas. Pathophysiology: The cell of origin for dermatofibrosarcomas is generally regarded to be fibroblastic. Cytogenetic analysis of the tumor cells in 1990 revealed the presence of a supernumerary ringed chromosome harboring a t(17;22)(q22;q13) translocation which resulted in a fusion between the PDGFB gene and the COL1A1 gene promoter. This has been speculated to result in autocrine stimulated proliferation of tumor cell. This discovery has also led to the development of new, targeted therapies directed at blocking tumor growth stimulation.

Dermatofibromas are very common cutaneous neoplasms composed of haphazardly arranged dermal spindle cells. The lesions can be distinguished from DFSP by the lack of the prominent storiform pattern seen in DFSP. In addition, dermatofibromas typically do not infiltrate into the subjacent subcuticular fat lobules.

## 10. Answer: D- Psoriasis

Inflammatory linear verrucous epidermal nevus is a rare congenital lesion characterized clinically by linear scaling erythematous plaques, typically unilateral. Histologically, ILVEN demonstrates a papillomatous acanthotic epidermis with prominent alternating hyperorthokeratosis and parakeratosis. A mild to moderate perivascular and lichenoid lympho-histiocytic infiltrate is typically present. Spongiform microabscesses and intracorneal microabscesses are rarely seen. Focal mild spongiosis with exocytosis of lymphocytes may be seen.

Fungal Id reactions represent wide-spread allergic responses to fungal antigens. Clinically these lesions present as nummular scaling and weeping plaques over areas distant to the site of primary fungal infection. Histologically, they demonstrate features of a spongiotic dermatitis, with intercellular edema and microvesicle formation.

Chronic eczematoid dermatitis demonstrates many of the same features as the above mentioned entity, however the chronic nature of the lesion typically is reflected by the presence of acanthosis and less prominent spongiosis. The histologic sections shown in this case demonstrate the classic features of psoriasis. Clinically psoriasis presents as erythematous, variably pruritic, heavily scaling plaques distributed symmetrically over the extensor surfaces of the body; most commonly the elbows and knees. Other commonly affected areas include the mid back, natal cleft, and scalp. Histologically, psoriasis demonstrates acanthosis and confluent parakeratosis. One of the most striking features is the distinct absence of a granular layer, as shown in slide 2. Neutrophilic microabscesses in the granular/spinous layer, and in the stratum corneum are also classic findings. Dilated thin-walled vessels in the dermal papillae are often present. The diagnosis of psoriasis typically requires the exclusion of a dermatophyte infection.

## 11. Answer: A- Granuloma annulare

Granuloma annulare is an idiopathic condition that typically affects children on the dorsa of the hands and feet. Clinically, the lesions are typified by aggregated, discrete, non-scaling papules arranged in an annular configuration with central clearing. Histologically, the lesion demonstrates the features of a palisaded histiocytic reaction pattern. In the central area of the lesion, there is tinctorial alteration of the collagen fibers; the so-called necrobiosis effect. In addition there is prominent mucin deposition within the lesion. Although the present example is the most classic, other varieties exist; these include interstitial GA, deep GA, and perforating GA. The condition is benign and typically self-limited. The only association reported is with diffuse GA and diabetes. This association however, is not well established.

Although rheumatoid nodules demonstrate the above mentioned palisaded histiocytic reaction pattern mentioned above, and does shown prominent alteration of the collagen, there is typically no mucin present. In addition, the center of rheumatoid nodules is typified by fibrinoid necrotic material, which is described in the literature as having a "brick-red" color. These lesions occur predominantly over the extensor surfaces of the upper extremities, and occur in individuals with long-standing, active rheumatoid arthritis.

Epithelioid sarcoma is an uncommon malignant soft tissue neoplasm, often mistaken for rheumatoid nodule upon superficial examination. Epithelioid sarcoma, however, is distinguished by the presence of prominent nuclear pleomorphism, cellular atypia, mitosis and necrosis.

Necrobiosis lipoidica is a common idiopathic condition most commonly presenting on the lower extremity of individuals with glucose intolerance. Clinically, the lesions present as well circumscribed, non-scaling, slightly atrophic, yellowish plaques with central telangiectasia and peripheral erythema. Histologically, the lesions demonstrate layers or bands of necrobiotic collagen, interspersed by zones of palisaded histiocytes. An accompanying sparse plasmacytic infiltrate is often observed.

## 12. Answer: D- Necrobiotic xanthogranuloma

Necrobiosis lipoidica is a common idiopathic condition most commonly presenting on the lower extremity of individuals with glucose intolerance. Clinically, the lesions present as well circumscribed, non-scaling, slightly atrophic, yellowish plaques with central telangiectasia and peripheral erythema. Histologically, the lesions demonstrate layers or bands of necrobiotic collagen, interspersed by zones of palisaded histiocytes. An accompanying sparse plasmacytic infiltrate is often observed. In comparison with this case, needle-like cholesterol clefts are not a feature, nor is the presence of Touton-morphology giant cells.

Although what appears to be a ruptured follicular structure is present in image 2, and needle-like cholesterol clefts may be seen in ruptured EIC, the broad of the lesion, it's wide-spread clinical presentation, and the presence of the Touton giant cells make this diagnosis unlikely.

Xanthoma disseminatum is an uncommon cutaneous condition characterized by the diffuse presence of xanthomatized histiocytes within the papillary and reticular dermis. In more mature lesions, a mild inflammatory infiltrate may be seen. In addition, a spindle cell storiform infiltrate, rare Touton giant cells, and scarring may be evident. The lesions of XD are typically widespread and often involve the CNS, specifically the pituitary gland. This often leads to the associated finding of diabetes insipidus in patients with XD. Again, cholesterol clefts are not a feature of XD.

Necrobiotic xanthogranuloma (NXG) is a rare condition affecting individuals with paraproteinemia, most commonly multiple myeloma. Histologically, the lesion demonstrates the triad of cholesterol clefts, zones of necrobiosis, and Touton giant cells. Although the specific pathogenesis of the lesions is unknown, it is thought to be related to the deposition and subsequent phagocytosis of deposited immunoglobulin. This theory is supported by reports of NXG resolving after treatment of the underlying gammopathy.

## 13. Answer: C-Human herpes virus infection

Bullous pemphigoid is an uncommon auto-immune blistering dermatosis which often presents with urticarial, pruritic plaques which subsequently eventuate into bullous lesions. Histologically, the blisters show a pauci-inflammatory subepidermal blister. There are often scattered eosinophils present in the dermal infiltrate, and epidermal necrosis, as may be seen in erythema multiforme/toxic epidermo-necrolysis, is not evident. The acantholysis and virocytopathic change present in the current case are not features present in bullous pemphigoid. A biopsy for immunofluorescence typically shows a linear deposition of IgG and C3 along the dermo-epidermal junction of uninvolved skin.

Pemphigus vulgaris is another rare auto-immune blistering condition which typically presents in older individuals. This disease, like bullous pemphigoid, is caused by antibodies directed at the epidermis. The target antigens however, reside within the stratum spinosum, thus the blister formation occurs intra-epidermally, rather than sub-epidermally, as is the case in bullous pemphigoid. Histologically, the blisters demonstrate acantholysis with a mild superficial lympho-histiocytic infiltrate. Eosinophils may be present, but are not required for the diagnosis. A biopsy for immunofluorescence shows a net-like deposition of IgG between the acantholytic cells.

Human herpes virus, whether HHV1, 2 or VZV, demonstrate an acantholytic blister with prominent virocytopathic changes. These include multinucleate giant cells demonstrating marginated chromatin with "steel-grey" nuclear inclusions.

Benign familial pemphigus is a rare congenital blistering condition typically affecting the flexural areas of the body. The gene defect has been traced to the ATP2C1 gene, which codes for the SPCA1 (Secretory Pathway Calcium/manganese-ATPase), and is transmitted in an autosomal dominant pattern. Histologically, benign familial pemphigus shows broad zones of acantholysis with rounded up, acantholytic keratinocytes. These broken-off keratinocytes have often been likened to a "dilapidated brick wall". Other entities which may histologically mimic the features of benign familial pemphigus include Darier's disease (follicular dyskeratosis) and Grover's disease (transient acantholytic dermatosis). Immunofluorescent examination is negative.

## 14. Answer: B-Bullous Pemphigoid

Porphyria cutanea tarda is an uncommon disease, most commonly associated in the US with liver disease related either to hepatitis or alcoholic liver disease. Clinically the lesions present as burning, tense blisters clustered over the sun exposed areas of the body, most commonly the hands, shoulders, and upper arms. The blisters rupture and drain clear fluid and heal with scarring and milia formation. Histologically, PCT demonstrates the features of a pauci-inflammatory sub-epidermal blister with festooning of the rete. The blisters are typically located over the acral surfaces. Eosinophils are rarely seen.

Bullous pemphigoid is an auto-immune bullous dermatosis caused by antibodies directed against the proteins on the surface of basilar keratinocytes responsible basement membrane anchoring. The disease typically presents as urticarial plaques that subsequently blister, forming large tense blisters that subsequently rupture and heal without scarring. Histologically, bullous pemphigoid is characterized by a pauci-inflammatory sub-epidermal blister, often with a prominent infiltrate of eosinophils. The blister roof is intact and viable, unlike cases of toxic epidermo-necrolysis or erythema multiforme. The diagnosis of bullous pemphigoid can be confirmed by immunofluorescence studies which demonstrate a linear deposition of IgG and C3 at the basement membrane.

Photo-allergic skin reactions may occur in response to a wide array of drugs and cutaneous exposures. The clinical presentation consists of well-demarcated erythematous weeping plaques, often in a unilateral or linear distribution on sun-exposed areas of the body. Histologically, photo-allergic reactions demonstrate prominent spongiosis and eosinophils. The spongiosis may lead to blister or micro-vesicle formation. These micro-vesicles occur intraepidermally, and not sub-epidermally, as is the case in bullous pemphigoid.

Polymorphous light eruption is an idiopathic hypersensitivity condition in which the individual experiences a transient allergic-like reaction to certain wave-lengths of UV light. Histologically, PMLE is characterized by papillary dermal edema, and an intense lymphocytic perivascular infiltrate. The infiltrate often extends into the deep subcutis. Eosinophils are distinctly absent, and their presence makes the diagnosis of PMLE unlikely.

## 15. Answer: C- Discoid lupus erythematosus

Central centrifugal scarring alopecia (CCSA) is a common idiopathic scarring alopecia affecting African American females. The lesions begin at the vertex and spread outward. The hair initially thins, then is lost completely. The follicular orifices are also lost. This condition is considered to be synonymous with follicular degeneration syndrome, pseudopelade of Broque and folliculitis decalvans. The histologic features are variable, depending upon the level of inflammatory activity. Consistent features include premature desquamation of the inner-root sheath of the hair follicle. This may be associated with damage to the outer root sheath, with the hair follicles eventually moving to an eccentric location, then eventually rupturing through to the outer root sheath. This progression is associated with concentric fibrosis of the follicle and a variably intense lymphocytic infiltrate. In more acute settings, the inflammatory infiltrate consists of neutrophils and histiocytes with scattered plasma cells. The process may also show evidence of bacterial folliculitis. This is regarded as a secondary phenomenon, and is not central to the pathogenesis of the disease. Alopecia areata is an idiopathic non-scarring alopecia typically affecting younger individuals. The extent of the disease can vary between limited small asymptomatic plaques on the scalp to whole body involvement. The disease is typically self limited and complete re-growth of hair is the most common outcome. Histologically, alopecia areata demonstrates the classic finding of a "swarm of bees" around the follicular base. The swarm is composed predominantly of lymphocytes. There is no evidence of vacuolar degeneration of the hair follicle. Numerous fibrous tracts consistent with catagen and telogen hairs are evident.

Discoid lupus erythematosus is a common, cutaneously limited form of lupus erythematosus. The lesions typically present as scarred, atrophic plaques with hyper and hypopigmentation. These lesions most commonly occur on the scalp, face and ears. The lesions scale and are often tender to the touch. A minority of patients with discoid lupus will have serologic markers suggestive of systemic lupus, and a small fraction of these cases may progress to the systemic form. Histologically, DLE is characterized by a "pan-dermatitis". All structures of the epidermis and dermis are affected by a predominantly lymphocytic infiltrate. There is basal vacuolopathy with associated inflammation of the hair follicles, eccrine coils, and superficial perivascular plexus. Other findings suggestive of DLE include the dilated and plugged hair follicles, and the prominent mucin deposition.

Folliculitis decalvans, as mentioned above is a form of CCSA. Typically this form is more inflammatory than pseudopelade of Broque, but less inflammatory than dissecting cellulitis. Clinically folliculitis decalvans demonstrates central scarring alopecia with peripheral inflammatory pustule formation. The lesions progress in a waxing and waning course. Histologically, Folliculitis decalvans cannot be differentiated from other forms of CCSA.