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Quiz in Dermatology

Quiz questions from skin and systemic diseases

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1. First described by two legendary dermatologists from Madras, presenting with pigmented macular lesions on palms and soles; on histopathology, reveals localized areas of fingerlike prolongation of rete ridges with increased pigmentation of basal cell along with small islands of whorled and wavy fibers with spindle-shaped nuclei. What sign or what disease are we talking about?
2. Insomnia, tachycardia, and a pattern of vascular mottling together constitute Marshal-White syndrome. A similar pattern of mottling when seen with cyanosis and urticaria-like eruption is termed as BASCULE syndrome. What is in the B?
3. Identify this syndrome characterized by a triad of discolored nails, pulmonary manifestations, and lower limb lymphedema.
4. Considered as a rare differential diagnosis for pustular psoriasis, this condition often presents within the first few weeks of life with fetal distress, pustular rash, and joint inflammation. If you know the gene mutation, you can "name" the condition. If you know the name, you can definitely identify the mutation. {Hint: Anakinra}
5. Dr. HS Plummer and colleagues described concave, distal onycholysis and longitudinal striations with flattening of the nails as a pathognomonic sign for "X." Although not considered pathognomonic anymore; in which systemic condition do you see these nail changes?
6. If considered as a spectrum of diseases, these psychocutaneous conditions can be represented by literary characters on either end. The more common and less dramatic variant is often related to one of the merry men from the folklore of Robin Hood [Figure 1], while the rare variant is as famous as the maiden on the high tower [Figure 2]. Identify both conditions.
7. "Autoimmune polyendocrinopathy candidiasis ectodermal dystrophy" syndrome is caused by mutation of which gene?
8. This deficiency disorder can present as white nail beds and hypopigmentation of skin and hair which resolve with appropriate supplementation. However, the most characteristic finding is endemic cardiomyopathy, which is often fatal and is mainly seen in endemic areas of Keshan County of Northeast China. Identify this deficiency disorder.
9. Elderly, diabetic male presented with a long history of asymptomatic, multiple, minute, hyperkeratotic papules grouped in a miniature "cobblestone" pattern on the dorsum of the distal phalanges, knuckles, and the interphalangeal joints. What are these papules known as?
10. Bywater's lesions are punctate, tender, purpuric papules on the distal digits associated with which rheumatological condition?
11. A 20-year-old male presented with a history of dyspnea and palpitations since 6 years of age. Chest radiograph revealed right ventricular and atrial enlargement along with a

Figure 1: The Merry man from the folklore of Robin Hood. Image used under creative commons license (https://sherwoodforesthistory.blogspot.com/2011/09/friar-tuck.html?m=1).

Figure 2: Maiden on the high tower. Image used under creative commons license (https://pixabay.com/illustrations/rapunzel-tangled-island-plats-5996180/).

prominent pulmonary trunk. An intern with Sherlock-like observation skills alerted the attending physician of a typical pigmented skin lesion. What was the patient subsequently diagnosed with?

12. Sneddon's syndrome is a non-inflammatory, thrombotic vasculopathy characterized by cerebrovascular manifestations and extensive skin lesions due to permanent impairment of peripheral blood flow. What cutaneous lesion do we expect to see in Sneddon's syndrome?
13. Coronavirus disease-2019 (COVID-19) has proved to be equally intriguing for dermatologists with its myriad cutaneous presentations. However, the fluorescence of hair and nails under Wood's lamp has been attributed to the use of a specific drug for COVID-19. Identify this drug.
14. "X" is a rare, multifocal, vascular anomaly syndrome characterized by skin, soft tissue, gastrointestinal, and venous malformations leading to severe anemia. Octreotide, antiangiogenic agents, laser photocoagulation, and aggressive surgical excisions have been tried with mixed results. However, a mammalian target of rapamycin (mTOR) signaling pathway inhibitor paved a new way for management. Identify this syndrome and the new treatment modality.
15. A triad of fibrofolliculoma, trichodiscoma, and acrochordon with a possible background of benign or malignant renal growth is commonly known as …….
16. Schimmelpenning syndrome presents with extensive, linear lesions in a blashkoid fashion and associated neuroectodermal abnormalities. What characteristic cutaneous finding of Schimmelpenning can also be linked with patch testing, anetoderma, and pityriasis lichenoides?
17. The term "knife-cut" sign is used to describe the cutaneous, linear, erosive lesions seen in the "X disease." Histopathological analysis of skin biopsy will show non-caseating granulomatous inflammation similar to the gastrointestinal "X disease." In which systemic disease do we see a knife cut sign?
18. In Greek mythology, "X" is a prophetic sea God, who can foretell the future, but, will change his shape to avoid doing so. An adjective derived from this feature of "X," means "mutable" or "capable of assuming many forms." "X" syndrome, thus named because of its "polymorphous" nature, is a severe disorder of asymmetric and disproportionate overgrowth of body parts. Specific features include cerebriform connective tissue nevus, thin limbs, lipomas, and lung cysts. Identify this Greek sea God.
19. Pigmentary abnormalities of the hair, skin, and eyes along with congenital sensorineural hearing loss, and Hirschsprung disease are characteristic features of which syndrome?
20. An internet slang used for a tough, intimidating person can also be used to describe a syndrome mostly seen in the context of gastric bypass surgeries. It usually presents as fever, malaise, and polyarthralgia followed by erythematous, macular or papulovesicular eruptions. Identify the slang or the syndrome.
21. First described in 2000, nephrogenic systemic fibrosis (NSF) is a systemic disorder, characterized by fibrotic and sclerotic-myxedematous skin lesions occurring in end-stage chronic kidney disease patients. In 2006, it was linked with previous exposure to certain contrast media. The United States of America Food and Drug Administration mandated black box warnings about NSF and other restrictive policies about their use leading to a drastic decline in the incidence. Which specific contrast media was implicated in NSF?

22. A French tire company is often associated with a benign, hamartomatous condition presenting with numerous, deep, gyrate symmetrical skin folds over extremities. Identify this French tire giant and the condition.
23. Flaky paint dermatosis is seen in which nutritional deficiency disorder?
24. "X" community, one of the main ancestral groups of a certain religion, was mainly settled along the banks of the river Rhine in Germany. The following dermatological conditions are most commonly seen in the descendants of this group (non-exhaustive list). Identify these syndromes and the community.

A syndrome
- Short stature
- Photosensitivity/ malar rash
- Mild immunodeficiency
- Insulin resistance
- Increased susceptibility to cancers

A storage disorder
- Glucocerebrosidase deficiency
- Hepatosplenomegaly
- Anemia/ thrombocytopenia
- Diffuse pigmentation, easy tanning

An angioproliferative sarcoma
- Viral infection
- Dark blue or purple red skin lesions

25. Presence of follicular and non-follicular pustules over the intertriginous area, scalp, and the periorificial areas of the head in the setting of an autoimmune condition like systemic lupus erythematosus is known as .....
26. A young bodybuilder on an ill-advised raw egg diet presented with erythematous and scaly periorificial dermatitis around the eyes, nose, and mouth. What supplementation would you start him on?
27. A 38-year-old female presented with 2-year history of erythema multiforme, urticarial rashes, and dermographism occurring 1 week before menstruation which resolve within 2 days of menstruation. Which cyclical autoimmune disorder will you consider?
28. A 45-year-old woman presented with a few tender, non-edematous, and ecchymotic patches over her thighs. She has been experiencing similar recurrent episodes for the past 2 years. She gave no preceding or concomitant history of injury, drug intake, malnutrition, or external bleeding manifestation. Her hematological workup was unremarkable. After a thorough evaluation, she was diagnosed as a case of Diamond-Gardner syndrome. What crucial history would have helped arrive at this diagnosis?
29. Pinch purpura is the characteristic dermatological finding of which systemic condition?
30. Strasbourg diagnostic criteria for this syndrome involve the occurrence of chronic episodes of a primary dermatological lesion as an obligate criterion for diagnosis. Identify the primary dermatological lesion and the associated autoinflammatory syndrome.

Obligate criteria
- A primary dermatological lesion
- Monoclonal immunoglobulin (Ig)M or IgG

Minor criteria
- Recurrent fever
- Objective findings of abnormal bone remodeling with or without bone pain
- A neutrophilic dermal infiltrate on skin biopsy
- Leukocytosis and/or elevated C-reactive protein

Definite diagnosis	Probable diagnosis
Two obligate criteria plus two minor criteria (if IgM) or three minor criteria (if IgG)	Two obligate criteria plus one minor criterion (if IgM) or two minor criteria (if IgG)

Answers

1. Premalatha-Yesudian sign
   Premalatha-Yesudian sign refers to palmar melanotic macules seen in neurofibromatosis.[1]
2. Bier spots
   Bier spots are physiologic anemic macules mostly seen on extremities. They are thought to be an exaggerated physiologic vasoconstrictive response of the small cutaneous vessels to hypoxia-induced by venous stasis or venous hypertension.[2]
3. Yellow nail syndrome
   Yellow nail syndrome is associated with FOXC2 gene mutation.[3,4]
4. DIRA (deficiency of interleukin 1 receptor antagonist)
   DIRA is an autosomal-recessive condition due to pathogenic variants in IL1RN, the gene encoding the IL-1RA (interleukin 1 receptor antagonist). Treatment with recombinant IL-1RA and anakinra results in marked improvement.[5]
5. Hyperthyroidism
   Plummer's nails are seen in patients with hyperthyroidism. The hyponychium commonly traps dirt, giving the nail a dark appearance and the name "dirty nails".[6]
6. Trichotillomania and Rapunzel syndrome
   Friar Tuck (from Robin Hood folklore) sign is seen in trichotillomania. In severe conditions, individuals often consume the plucked hair (trichophagia) resulting in trichobezoars. If these trichobezoars extend to the duodenum; they are then called as the Rapunzel syndrome.[7,8]
7. AIRE (autoimmune regulator) gene[9]
8. Keshan disease or selenium deficiency[10]
9. Huntley's papules[11]
10. Rheumatoid arthritis
    Bywater's lesions include nailfold thromboses and purpuric papules on the distal digits. Histology reveals small-vessel leukocytoclastic vasculitis.[12]

11. Watson syndrome
    "Elementary, my dear Watson" as Sherlock Holmes would say. Café-au-lait macules with pulmonary stenosis (right ventricular and atrial enlargement) are typical of Watson syndrome, an allelic variant of neuro fibromatosis type 1.[13]
12. Livedo racemosa
    Sneddon's syndrome is a rare, non-inflammatory, thrombotic vasculopathy characterized by the combination of cerebrovascular disease with livedo racemosa. Livedo racemosa is a permanent reddish-blue mottling of the skin in an irregular, large, and reticular pattern consisting of broke, circular segments, while the more common livedo reticularis consists of fine, regular, complete network, and resolves on warming.[14]
13. Favipiravir[15]
14. Blue rubber bleb nevus syndrome and mTOR inhibitor sirolimus[16,17]
15. Birt-Hogg-Dubé syndrome[18]
16. Nevus sebaceus of Jadassohn[19,20]
    Joseph Jadassohn, a German dermatologist, is considered as the father of patch testing and was the first to report anetoderma and pityriasis lichenoides.[21]
17. Metastatic Crohn's disease
    The cutaneous, linear, erosive flexural lesions seen in metastatic Crohn's disease are described as knife-cut sign.[22]
18. Proteus – Greek sea God
    Protean – mutable/versatile. Proteus syndrome is caused by a variant in a growth regulatory gene called AKT1 that occurs after fertilization of the embryo (somatic mutation).[23]
19. Waardenburg syndrome Type 4[24]
20. BADAS
    BADAS (Bowel-associated dermatosis-arthritis syndrome) is an uncommon neutrophilic dermatosis characterized by arthralgias, fever, myalgias, and malaise as well as cutaneous eruptions on the extremities and trunk. Badass, the noun, is used to refer to a tough, uncompromising, or intimidating person.[25]
21. Gadolinium-based contrast media[26]
22. Michelin tire; Michelin tire baby syndrome
    The term was derived from the cartoon mascot of the Michelin tire company.[27]
23. Kwashiorkor
    Other signs seen with Kwashiorkor are flag sign (hair) and broomstick appearance (eyelashes).[28]
24. Bloom syndrome, Gaucher's disease, and Kaposi sarcoma
    Most commonly seen in Ashkenazi Jews.[29]
25. Amicrobial pustulosis of folds
    Amicrobial pustulosis of folds is a rare manifestation of autoimmune conditions, which on histopathology, reveals spongiform, subcorneal pustulosis associated with mainly neutrophilic dermal infiltrate.[30]
26. Biotin supplementation
    Avidin in raw egg forms a strong bond with free biotin, thus causing acquired biotin deficiency. Cooked eggs do not lead to biotin deficiency as avidin loses its affinity for biotin on heating.[31]
27. Autoimmune progesterone dermatitis[32]
28. Psychosocial history
    Psychogenic purpura, Gardner-Diamond syndrome or autoerythrocyte sensitization syndrome, is a rare condition characterized by spontaneous development of painful, edematous, ecchymotic skin lesions following episodes of severe stress and emotional trauma.[33]
29. Primary systemic amyloidosis[34]
30. Urticaria and Schnitzler syndrome
    Schnitzler syndrome is an autoinflammatory syndrome, which presents with low-grade fever, recurrent urticarial rash, and muscle, bone, and/or joint pain with enlarged lymph nodes.[35]

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Figure 1 image used under creative commons license (https://sherwoodforesthistory.blogspot.com/2011/09/friar-tuck.html?m=1). Figure 2 image used under creative commons license (https://pixabay.com/illustrations/rapunzel-tangled-island-plats-5996180/).

Declaration of patient consent

Not required as there are no patients in this article.

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Conflicts of interest

Dr. Soumya Jagadeesan is on the editorial board of the Journal.

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