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Quiz in Dermatology

Quiz questions from tumors of skin and appendages

Parambath Nimitha[1]

[1]Department of Dermatology, Malabar Hospital, Kozhikode, Kerala, India.

***Corresponding author:**
Parambath Nimitha,
Department of Dermatology,
Malabar Hospital, Kozhikode,
Kerala, India.

nimitha90@gmail.com

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Quick Response Code:

1. Identify the autosomal dominant (AD) condition characterized by follicular atrophoderma of hands and feet since birth, multiple basal cell carcinomas (BCC) since adolescence, facial hypohidrosis, and sparse hair.
2. Which is the most common mutation associated with familial melanoma?
3. What temperature must be achieved with cryotherapy for the adequate treatment of a small and superficial squamous cell carcinoma (SCC)?
4. What is the name given to the AD variant of keratoacanthoma seen in children?
5. SCC of ……. (body region) shows the greatest risk of metastasis.
6. Which body site is the most common location for fibroepithelioma of Pinkus variant of BCC?
7. According to the reflectance confocal microscopy, retraction artifact seen in histopathology of BCC occurs due to……
8. Name the only US FDA (Food and Drug Administration, The United States of America) approved oncolytic viral therapeutic agent and the malignancy for which it is used.
9. Dermoscopy showing well-focused, arborizing vessels, multiple blue-gray globules, leaf-like structures, large blue-gray ovoid nests, and spoke-wheel areas in the absence of pigment network on a pigmented papule on the face of an elderly male is diagnostic of …….
10. What is the most likely diagnosis [Figure 1]?
11. What is the relation between the size of a congenital melanocytic nevus and its risk of developing a melanoma?
12. Which human papilloma virus (HPV) serotype is most commonly associated with periungual SCC?
13. Name the syndromes associated with multiple pilomatricomas.
14. What is the overall risk of developing cutaneous and systemic malignancies in an organ transplant recipient?
15. What are the histological features which differentiate microcystic adnexal carcinoma from desmoplastic trichoepithelioma?
16. According to the American Academy of Dermatology (AAD) and National Comprehensive Cancer Network recommendations, the surgical margin required for melanomas that measure more than 2 mm in thickness is……
17. Identify the mutation associated with the familial form of this disease [Figure 2].
18. Which of the following is a systemic association of eruptive vellus hair cyst?
    a. Renal failure
    b. Hyperuricemia

c. Cardiac failure
d. Bronchial asthma

19. Tent sign and teeter totter sign are seen in…….
20. The most likely appendageal tumor that manifests as multiple, eyelid cysts in a patient with Schopf-Schulz-Passarge syndrome?
21. A 21-year-old male presented with a slow growing flesh-colored, smooth, mobile, firm, and well-circumscribed nodule on the scalp [Figure 3]. Histopathology showed a dermal cyst lined by stratified squamous epithelium without granular layer and compact keratin as content. Diagnosis would be…….
22. A 50-year-old male presented with segmentally arranged firm, painful swellings on the left, lower anterior chest wall [Figure 4]. He had pain which increased on exposure to cold. What are the systemic associations that one must look for in the given clinical presentation?
23. Presence of multiple trichodiscomas and fibrofolliculomas should arouse the suspicion of .....malignancy.
24. Which immunosuppressant drug is most commonly associated with sudden eruption of sebaceous hyperplasias in renal transplant recipients?
25. Which is the systemic condition that is associated with clear cell syringomas?
26. Which specific histopathological feature acts as a clue in diagnosing Gardner's syndrome in patients with epidermoid cysts?
27. …….is the most common location for Woringer-Kolopp disease.
28. CD34 positive and factor XIIIa negative dermal tumor showing t(17;22)(q22;q13) translocation suggests…….
29. Painless and firm, solitary nodule on the face of an elderly man, which stains positive for neuron specific enolase is suggestive of……
30. In this biopsy confirmed BCC patient [Figure 5], what would be the treatment of choice?

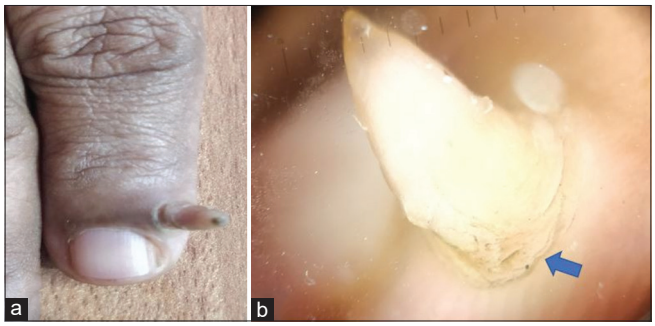

Figure 1 (a): Skin-colored and well-defined papule with a characteristic hyperkeratotic collarette at its base; (b): the dermoscopic view (Heine Delta 20T, Heine Optotechnik, Herrsching, Germany, polarized, ×16) shows a central pale-yellow core surrounded by a hyperkeratotic, white, collarette (arrow).

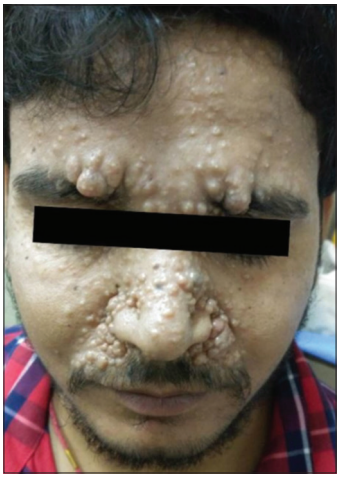

Figure 2: Midface and eyebrows show symmetrically arranged round-to-oval, skin-colored, and smooth-surfaced papules. Medial eyebrow involved in the form of pedunculated papulo-nodules.

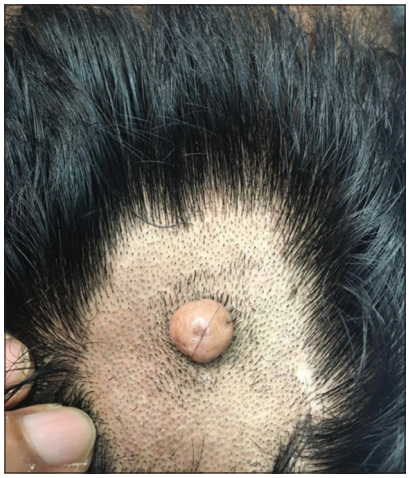

Figure 3: Flesh-colored, smooth, mobile, firm, and well-circumscribed nodule on the scalp.

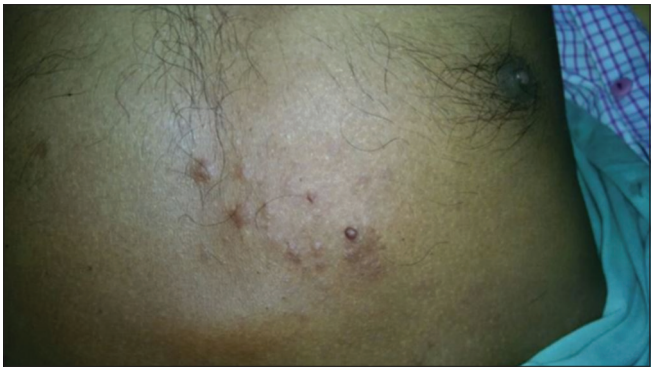

Figure 4: Multiple and segmentally arranged skin-colored to erythematous papules involving the left anterior chest wall.

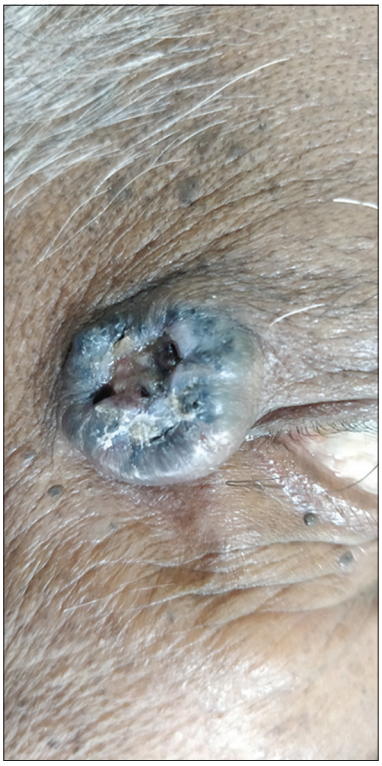

Figure 5: Well-defined 2.5 × 2 cm hyperpigmented, ulcerated plaque involving the lateral canthus; surface shows scales and adherent crusts.

Answers.

1. Bazex-Dupré-Christol syndrome
   Other syndromes associated with BCC are Gorlin syndrome/nevoid BCC syndrome, multiple hereditary infundibulocystic BCC, Rombo syndrome, Brooke-Spiegler syndrome, Oley syndrome, Schopf-Schulz-Passarge syndrome, and xeroderma pigmentosum.[1]
2. Cyclin-dependent kinase inhibitor 2A (CDKN2A) gene located on chromosome 9p21. The most common cause of inherited melanoma is mutations in CDKN2A gene.[2]
3. For destroying malignant tumors, the tissue should be frozen to –50°C for at least two freeze-thaw cycles. Benign lesions can be destroyed at temperatures around –25°C for keratinocytic tumors and –5°C for pigmented lesions.[3]
4. Ferguson-Smith type of keratoacanthoma
   These are usually multiple in number and are self-healing. Another variant is Grzybowski, which is a non-familial disorder in which patients develop generalized eruptive keratoacanthomas after the age of 40 years.[4]
5. Ear
   The approximate rate of metastasis from ear is 14%, which is slightly higher than the metastasis rate of 11% from the lip.[5] Size greater than 2 cm, perineural invasion, history of immunosuppressive therapy, histological types (such as "acantholytic," "spindle," "verrucous," "cutaneous SCC with single cell infiltrates," and "desmoplastic"), >2 mm Breslow's depth, and Clark's grade > IV are the other factors which predispose to metastasis.[6,7]
6. Lumbosacral area.[8]
7. Cleft formation between BCC nests and stroma was regarded as a fixation artifact occurring due to shrinkage of mucin during tissue fixation and staining. However, *in vivo* reflectance confocal microscopy and *ex vivo* confocal laser scanning microscopy also detect the gaps. Hence, the new theory suggests degradation of extracellular matrix during tumor growth as the actual reason for the cleft formation[9]
8. Talimogene laherparepvec in metastatic melanoma.
   This technique involves intra-tumoral injection of oncolytic virus which specifically and effectively destroys malignant cells without much immune-related adverse events.[10]
9. BCC
   The dermoscopic features of BCC include absence of pigment network and presence of at least one of the following – ulceration, blue-gray globules, maple-leaf like structure, blue-ovoid nests, arborizing vessels, and spoke-wheel structures.[11-13]
10. Acquired digital fibrokeratoma
    It is a solitary acral lesion with hyperkeratotic collarette [denoted by the arrow in Figure 1 (b)] which characteristically lacks nerve twigs and bone on pathologic examination.[14]
11. Risk of melanoma is directly proportional to the size of the congenital melanocytic nevus. Risk of melanoma increases with the size of the nevus. For small and medium-sized congenital melanocytic nevi, the risk of developing melanoma is < 1% whereas for giant congenital melanocytic nevi (more than 20 cm in projected adult size), the risk is likely <5%.[15]
12. Most common HPV subtype associated with periungual SCC is HPV 16, other subtypes are 2, 11, 18, 26, 31, 34, 35, 58, 56, and 73.[16]
13. Curschmann Steinert myotonic dystrophy, Rubinstein-Taybi syndrome, Turner syndrome, and Gardner's syndrome. Usually, syndromic association is seen, if the number of pilomatricomas is six or more.[17]
14. Four-fold
    Organ transplant recipients have 3–4 times higher risk of developing malignancies in comparison to the general population.[18] Cutaneous malignancies are the most common. The risk is in the following order: SCC (65-fold) > BCC (10-fold) > melanoma (3.4-fold)).[19]
15. Perineural invasion, deep subcutaneous infiltration, ductal differentiation, and presence of mitotic figures differentiate microcystic adnexal carcinoma from desmoplastic trichoepithelioma.[20] Both come under the category of paisley tie tumors. Hence, a superficial biopsy is inadequate to diagnose microcystic adnexal carcinoma.
16. 2 cm
    The current surgical margins are 0.5 cm for melanoma *in situ*. The recommendation is wide local excision

with a negative margin of 1 cm for tumors of < 1 mm thickness, 1–2 cm for tumors 1–2 mm thick, and 2 cm for tumors more than 2 mm thick.[21]

17. Inactivating mutation in tumor suppressor gene CYLD. Brooke Spiegler syndrome shows multiple trichoepitheliomas, cylindromas, and spiradenomas on head-and-neck.[22]
18. A. Renal failure
    Late onset, eruptive vellus hair cyst may be associated with renal failure. Other associations are Lowe syndrome and hidrotic and anhidrotic ectodermal dysplasia.[23]
19. Pilomatricoma
    "Tent sign" is pathognomonic of pilomatricoma, in which stretching of skin over the lesion shows multiple facets and angles. "Teeter totter" sign can be demonstrated by pressing one edge of the lesion, which results in protrusion of the opposite edge like a teeter totter.[24]
20. Apocrine hidrocystoma
    Other features are hypodontia, hypotrichosis, nail dystrophy, palmoplantar keratoderma, hyperhidrosis, and BCC.[25]
21. Trichilemmal/pilar cyst.[26]
22. The picture depicts segmental leiomyoma. Systemic associations are uterine leiomyomatosis, and renal cell carcinoma.[27] Reed's syndrome, a genetic disorder with an AD pattern of inheritance, manifests cutaneous and uterine leiomyomas.[27] A subgroup of patients with Reed's syndrome develops renal cell cancer ("hereditary leiomyomatosis and renal cell cancer").[27]
23. Renal malignancy
    These occur as a part of Birt-Hogg-Dubé syndrome. Other important finding is spontaneous pneumothorax.[28]
24. Cyclosporine
    Mechanism postulated is hyperproliferation of undifferentiated sebocytes and differentiation arrest leading to enlargement of sebaceous glands.[29]
25. Diabetes mellitus
    Clear cells represent accumulated glycogen.[30]
26. Apart from the usual histopathological findings of epidermoid cyst, the lesions in Gardner's syndrome show the presence of hair matrix like basophilic cells indistinguishable from pilomatricoma. Since epidermoid cysts are the earliest clinical manifestations of Gardner's syndrome, early detection helps in screening for complications like intestinal polyposis.[31]
27. Hands and feet
    Pagetoid reticulosis/Woringer-Kolopp disease presents as scaly, oval plaques in acral areas.[32]
28. Dermatofibrosarcoma protuberans (DFSP)
    DFSP is CD34 positive and factor XIIIa negative, allowing its differentiation from dermatofibroma (usually CD 34 negative and factor XIIIa positive). Dermatofibroma may show a weak positivity for CD34 at the periphery. Factor XIIIa is positive in dermatofibroma, but it lacks specificity. The translocation t(17;22)(q22;q13) resulting in COL1A1-PDGFB fusion is seen in all cases of DFSP.[33]
29. Merkel cell carcinoma
    This neuroendocrine tumor has a high potential for regional and distant metastasis.[34]
30. Mohs micrographic surgery[35]

The following are the indications for Mohs micrographic surgery.

1. Large BCC (>1 cm on the face or >2 cm on the trunk)
2. Involvement of high-risk anatomic areas (ear, eyelids, lips, nose, temples [Figure 5], nipple, genitalia, hand, feet, ankle, and nail unit)
3. Aggressive histological subtype – morpheaform, infiltrating, and micronodular BCC and BCC with perineural involvement.
4. Tumors with positive margins, after conventional excision
5. Recurrent BCC
6. Immunocompromised individuals
7. BCC in association with genetic syndromes
8. BCC occurring on a prior irradiated skin

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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