American College of Mohs Surgery

Diagnostic Quality Control Program 2010

(Review of Answers)

Question 1

A flesh-colored plaque on the right ala of an otherwise healthy 57 year-old male is referred for Mohs surgery.

The correct diagnosis is:

a. Desmoplastic trichoepithelioma
b. Microcystic adnexal carcinoma
c. Keratotic basal cell carcinoma
d. Metastatic adenocarcinoma

Discussion

Question 1

Correct answer:

b. Microcystic adnexal carcinoma

Histologic features:

- Poorly circumscribed dermal neoplasm embedded in a desmoplastic stroma that invades deeply
- Superficially, the tumor is composed of small to medium-sized nest and cords often containing keratinizing cysts lined by squamous epithelium. Nests and cords consist of two components (horn cysts and ductal structures) present in varying amounts. Some tumor foci may exhibit a syringomatous appearance.
- Deeper, the tumor is made up of cells arranged in smaller strands, cords and nest of tumor cells infiltrating into the deep muscle tissue. Perineural/intraneural invasion is frequently seen in MAC
- Tumor cells are cytologically bland with rare mitosis.
- Pancytokeratin, AE1/AE3, and cytokeratin-1 positively highlight tumor cells. EMA and CEA highlight ductal structures or intracytoplastic lumen formation
- CK 15 can be a useful marker in distinguishing MAC from infiltrative BCC and SCC with ductal differentiation.
- CD5 positivity in deeper sections would favor MAC over desmoplastic trichoepithelioma

Differential diagnosis:

- Desmoplastic trichoepithelioma
- Infiltrative basal cell carcinoma
- Metastatic adenocarcinoma
- Eccrine carcinoma

Clinical concerns:

- Tendency for perineural and intraneural invasion, local recurrence common, but rare metastasis
- Can be confused with infiltrative basal cell carcinoma, metastatic tumors of glandular origin and desmoplastic trichoepithelioma

References:

Hoang MP, Dresser KA, Kapur P, et al. Microcystic adnexal carcinoma: an immunohistochemical reappraisal. Mod. Path. 2008 Feb; 21(2):178-85.

Practical Dermatopathology. Edited by R. Rapini 2005 Elsevier Mosby

Question 2

The following Mohs section was taken from the scalp of a 59 year-old female with a history of a nodular basal cell carcinoma

The correct diagnosis is:

a. Nodular basal cell carcinoma
b. Eccrine spiradenoma
c. Subcutaneous lymphoid hyperplasia
d. Merkel cell carcinoma
e. Malignant eccrine spiradenoma

Discussion

Question 2

Correct answer:

b. Eccrine spiradenoma

Histologic features:

- Sharply demarcated nodules of basaloid cells in dermis or subcutaneous tissue ("blue balls")
- Almost never any connection to the epidermis
- Basaloid cells are of two types: the outer cells are small with compact nuclei and the inner cells are larger with more cytoplasm and lighter staining.
- Areas of eosinophilic hyaline deposits are seen between thin anastomosing cords
- Basaloid cells tend to be arranged in rosettes in rich vascular stroma
- Sparse small sweat ductal lumina usually present

Differential diagnosis:

- Nodular basal cell carcinoma
- Nodular hidradenoma (eccrine acrospiroma)
- Cylindroma

Clinical concerns:

- Solitary, often painful, intradermal nodule with bluish discoloration present often on the head and neck area
- Rare malignant variants have been reported. Histologically malignant eccrine spiradenomas demonstrate pleomorphic cells with hyperchromasia, increased numbers of mitotic figures, and often a loss of the two cell populations. Because the benign form can be seen within the carcinomatous variants, the entire specimen should be evaluated carefully.
- May mimic basal cell carcinoma

References:

Practical Dermatopathology. Edited by R. Rapini 2005 Elsevier Mosby.

Skin Cancer. Edited by K. Nouri 2008 McGraw-Hill.

Question 3

The following Mohs section was taken from a 77 year-old male with a 1.1 cm eroded, red nodule on the right upper forehead. The immunostaining profile for this tumor is as follows: S100 (-), CD68 (+), HMB-45 (-), and vimentin (+).

The correct diagnosis is:

a. Dermatofibroma sacrcoma protuberans
b. Atypical fibroxanthoma
c. Leiomyosarcoma
d. Spindle cell melanoma

Discussion

Question 3

Correct answer:

b. Atypical fibroxanthoma

Histologic features:

- The tumor appears to arise from the reticular dermis and subcutaneous tissues as a cellular proliferation of bizarre spindle cells, epithelioid cells, or multinucleated giant cells
- It distends but does not involve the epidermis,
- Often there is background solar elastosis
- Severe pleomorphism, hyperchromatism, many very atypical mitoses
- Tumor cells have abundant eosinophilic and large hyperchromatic nuclei
- Malignant cells often have multiple, atypical nuclei
- The atypical, hyperchromatic nuclei are dispersed in between the collagen bundles
- Positive staining for vimentin and histiocyte stains (CD68), negative staining with S-100 and pancytokeratin

Differential diagnosis:

- Spindle cell squamous cell carcinoma
- Spindle cell melanoma
- Superficial portion of a malignant fibrohistiocytoma
- Leiomyosarcoma

Clinical concerns:

- In adult cases, skin underlying developing AFX lesions may be considered locally immunosuppressed. Recent reports showed an increased incidence of AFX in patients with AIDS and in patients who are immunosuppressed because of organ transplantation
- Local recurrence not uncommon after excision, MMS treatment of choice, metastasis a rare event

References:

Ang GC, Roenigk RK, Otley CC, Kim Phillips P, Weaver AL. More than 2 decades of treating atypical fibroxanthoma at mayo clinic: what have we learned from 91 patients?. *Dermatol Surg*. May 2009;35(5):765-72.

Practical Dermatopathology. Edited by R. Rapini 2005 Elsevier Mosby.

Question 4

The following Mohs section was taken from a 65 year-old male with a 1.8 cm painful red-brown plaque on the scalp. The immunostaining profile for this tumor is as follows: S100 (-), actin (+), HMB-45 (-), vimentin (+), and cytokeratins (-).

The correct diagnosis is:

a. Spindle cell squamous cell carcinoma
b. Epithelioid sarcoma
c. Cutaneous leiomyosarcoma
d. Desmoplastic melanoma

Discussion

Question 4

Correct answer:

c. Cutaneous leiomyosarcoma

Histologic features:

- Nodular growth pattern with high cellularity and densely packed oval and spindle-shaped cells arranged in longitudinal and transverse intersecting fascicles
- The more spindle-shaped cells exhibit cigar-shaped nuclei with eccentric vacuoles and eosinophilic cytoplasm
- Frequent mitoses, often atypical mitoses are characteristic
- Some tumors exhibit a diffuse growth pattern with a low cellularity and more well-differentiated smooth muscle cells and less conspicuous mitotic figures

Histologic concerns:

- Immunostains are helpful in confirming the diagnosis. Smooth muscle actin and vimentin are almost always expressed
- Some tumors do not express desmin, even though it is traditionally thought of as a distinctive marker for smooth muscle differentiation

Differential diagnosis:

- Spindle cell squamous cell carcinoma
- Desmoplastic malignant melanoma
- Epithelioid sarcoma

Clinical concerns:

- Although the tumor has been reported to have a high recurrence rate, there are no clear cut cases of metastasis, most metastatic leiomyosarcomas are of the subcutaneous type
- Treatment with wide local excision or Mohs micrographic surgery effective

References:

Mohs Micrographic Surgery. 2nd edition. Edited by S. Snow and GR Mikhail 2004. The University of Wisconsin Press.

Practical Dermatopathology. Edited by R. Rapini 2005 Elsevier Mosby.

Question 5

This patient is sent for Mohs surgery to remove his basal cell carcinoma. **The slide from their last layer indicates:**

a. There is tumor still present at the margin.
b. There is a congenital type nevus present, but no basal cell carcinoma.
c. There is a coincidental sebaceous adnexal tumor that does not require another layer.
d. There is a benign nevus and basal cell carcinoma present. Another layer is needed.

Discussion

Question 5

Correct answer:

b. There is a congenital type nevus present, but no basal cell carcinoma.

Histologic features:

- This slide shows the presence of nests of bland blue round cells starting in the superficial dermis and extending to the deep reticular dermis.
- The cells are larger at the top of the specimen but shrink with increasing depth. There is a lace like downward growth of epidermal strands and a horn cyst. The cells wrap around adnexal structures in places. These features are commonly associated with congenital nevi.
- There is no metachromasia around the nests, no retraction artifact and no peripheral palisading. Sebaceous glands are present, but are not atypical or compatible with a sebaceous adnexal tumor. Higher powered views will confirm the presence of nevoid like cells, consistent with benign nevus. Features of basal cell carcinoma are not seen.

Differential diagnosis:

- Acquired nevus. These are typically restricted to the papillary dermis, though the only way to truly know if something is congenital is by patient history.

Clinical concerns:

- At low power and a quick glance nevi stained with toluidine blue can sometimes simulate basal cell carcinoma. A more careful look will usually discern the two.

References:

Lever's Histopathology of the Skin, 10th edition, Lippincott Williams & Wilkins, Philadelphia 2008.

Question 6

This patient is undergoing slow Mohs surgery for his periocular 0.7 mm depth malignant melanoma. Provided is the slide from his last layer which is stained with micropthalmia transcription factor (MITF).

Which of the following is the most correct choice regarding this slide?

a. Invasive malignant melanoma is still present within these sections. Another layer is required.
b. There is no evidence of residual melanoma. A benign nevus is present and does not require an additional layer.
c. Melanoma in-situ is still present along with a follicular adnexal neoplasm. Another layer is required.
d. Melanoma in-situ is present along with a benign nevus. Another layer is required.

Discussion

Question 6

Correct answer:

b. There is no evidence of residual melanoma. A benign nevus is present and does not require an additional layer.

Histologic features:

- There are benign, small cells located along the dermal epidermal junction. There are no nests there, no cells above the basal layer and no atypical melanocytes.
- In the dermis there are nests of bland nevoid cells in the superficial epidermis. No atypical features are recognizable, compatible with a diagnosis of benign nevus. Hair follicles are present, but have no features of adnexal neoplasms.

Differential diagnosis:

- Clark's nevus could be considered if there were atypical features, but there are none present.

Clinical concerns:

- With a history of a malignant melanoma in the area where one is operating attention needs to be paid to possible dermal tumor involvement. However, benign nevi are often located in close approximation with melanomas and should not be misinterpreted.

References:

Lever's Histopathology of the Skin, 10th edition, Lippincott Williams & Wilkins, Philadelphia 2008.

Question 7

A 58-year-old solid organ transplant patient is sent for Mohs surgery for a forehead SCC in-situ. The included slide is from the first layer.

Which of the following is correct?

a. The layer is clear. The wound can be closed, but the patient should return routinely for skin cancer screening.
b. The layer is clear. The wound can be closed, and the patient does not need routine skin cancer screening due to the low probability of developing future malignancies.
c. There is evidence of SCC in-situ at the margin; a superficial additional layer should be obtained.
d. There is evidence of invasive SCC. An additional layer which includes subcutaneous tissue and, if necessary fascia should be obtained.

Discussion

Question 7

Correct answer:

d. There is evidence of invasive SCC. An additional layer which includes subcutaneous tissue and, if necessary fascia should be obtained.

Histologic features:

- Deep in the dermis and within the subcutaneous plane there are several islands of glassy atypical cells.
- Deep sections into the block reveal that these appear keratinocytic in morphology. Thus the diagnosis of invasive SCC is established. There is inflammation adjoining a nearby nerve, but no tumor cells visible there. No evidence of epidermal disease is seen.
- This tumor is well differentiated as witnessed by the formation of keratin.

Differential diagnosis:

- At quick glance one might mistake these tumor islands for eccrine glands or follicular hamartomas, but the presence of atypical glassy keratinocytes with formation of horn cysts is diagnostic of SCC.

Clinical concerns:

- This case illustrates the importance of scanning the entire slide even if the biopsy demonstrates only superficial disease. When there are questions regarding the type of cells or neoplasms encountered, deeper cuts into the block, as was the case with this tumor, will often clarify the diagnosis.

References:

Lever's Histopathology of the Skin, 10th edition, Lippincott Williams & Wilkins, Philadelphia 2008.

Question 8

A 70 year-old female presented with a growing, ulcerated nodule on the left heel. A biopsy was performed. The most likely diagnosis is:

a. Porocarcinoma
b. Basal cell carcinoma
c. Hidradenocarcinoma
d. Basaloid squamous cell carcinoma

Discussion

Question 8

Correct answer:

a. Porocarcinoma.

Histologic features:

- Porocarcinoma may be associated with long standing eccrine poroma or hidroacanthoma simplex.
- It shows groups of monotonous basaloid (poroma) cells with focal pleomorphism, hyperchromasia and mitoses.
- Eccrine differentiation is seen in the form of ductal structures with or without cuticular material.
- Squamoid differentiation can be seen and should be differentiated from squamous cell carcinoma.

Differential diagnosis:

- Eccrine poroma
- Basal cell carcinoma, metatypical
- Aggressive digital papillary adenocarcinoma

References:

Gschnait F, Horn F, Lindlbauer R, et al. Eccrine porocarcinoma. *J Cutan Pathol* 1980;7(6):349-353.

Kolde G, Macher E, Grundmann E. Metastasizing eccrine porocarcinoma. Report of two cases with fatal outcome. *Pathol Res Pract* 1991;187 (4): 477-481.

Question 9

A 74-year-old woman presented with a long standing plaque on the forehead. A deep shave biopsy was done. The most likely diagnosis is:

a. Acantholytic squamous cell carcinoma
b. Mucoepidermoid carcinoma
c. Hidradenocarcinoma
d. Nodular melanoma

Discussion

Question 9

Correct answer:

b. Mucoepidermoid carcinoma

Histologic features:

- These are rare, aggressive neoplasms showing typical features of squamous cell carcinoma with focal glandular differentiation and hence are called adenosquamous or mucoepidermoid carcinoma.
- Characteristically, they are multifocal and some biopsies may not show epidermal attachment.
- Glandular differentiation is appreciated as intracytoplasmic vacuoles which may form small vesicles.
- Mucicarmine satin and CEA immunohistochemistry may be helpful in the diagnosis.

Differential diagnosis:

- Acantholytic squamous cell carcinoma
- Pseudovascular squamous cell carcinoma
- Sebaceous carcinoma
- Hidradenocarcinoma

References:

Landmann G, Farmer E. Primary cutaneous mucoepidermoid carcinoma. *J Cutan Pathol* 1991; 18;56-59.

Revercomb C, Reitmeyer W, Pulitzer D. Clear cell variant of mucoepidermoid carcinoma of the skin. *J Am Acad Dermatol*. 1993; 29;642-644.

Question 10

A 59 year-old white man presented with multiple papules and nodules on the face, arms and trunk. The most likely diagnosis is:

a. Basal cell carcinoma, nodular
b. Trichoepithelioma
c. Trichofolliculoma
d. Trichoblastoma

Discussion

Question 10

Correct answer:

d. Trichoblastoma

Histologic features:

- Trichoblastomas are well circumscribed but unencapsulated, nodular neoplasms spanning the entire dermis.
- They show a wide range of histologic changes made up of two main components: germinative hair bulb basaloid cell differentiation and stromal mesenchyme.
- Neoplasms with more prominent stromal component and minimal germinative epithelial component are better referred to as trichogenic fibromas.
- Trichoblastomas show more basaloid cell formation and less mature mesenchymal stromal induction. They may show focal peripheral palisading but without retraction artifact.
- Adenoid features and melanin pigmentation may be seen.

Clinical concerns:

- They may arise in association with nevus sebaceus and rarely in some autosomal dominant syndromes such as Brooke-Spiegler syndrome.

Differential Diagnosis

- Basal cell carcinoma
- So called “trichoblastic carcinoma”
- Trichoepithelioma

References:

Regauer S, Beham-Schmid C, et al. Trichoblastic carcinoma (malignant trichoblastoma) with lymphatic and hematogenous metastases. *Modern Pathol* 2000;13;673-678.

Betti B, Alessi E. Nodular trichoblastoma with adamantinoid features. *Am J Dermatopathol* 1996;18;192-195.