JOURNAL OF DRUGS IN DERMATOLOGY
JDD
DRUGS • DEVICES • METHODS

Board Review

In partnership with Derm In-Review

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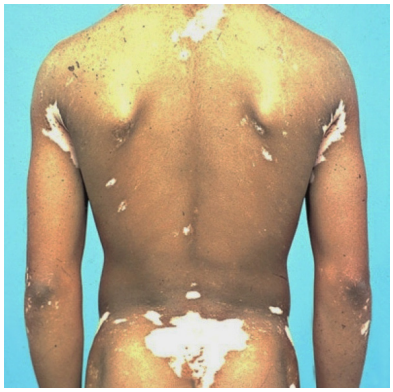
Amanda Abramson Lloyd, MD

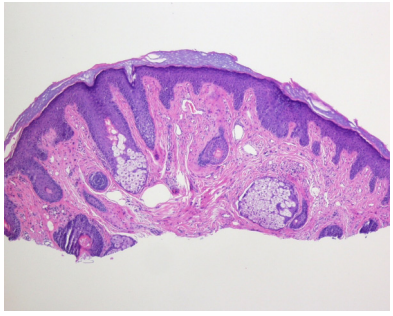
1)



A patient with a history of meningoencephalitis presents with white patches on his body as shown. During the patient encounter, you also note that the patient has trouble hearing you. What is the most likely associated ophthalmologic finding?

A. There is no associated ophthalmologic finding
B. Unilateral vision changes
C. Granulomatous uveitis
D. Dystopia canthorum
E. Heterochromic irides

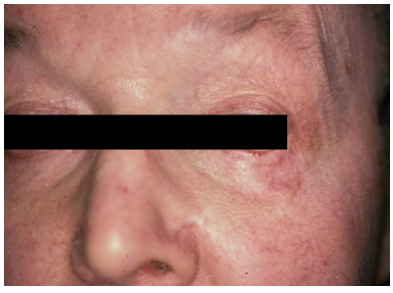
2)



A 13-year-old male presents with numerous facial lesions which have the pathology shown. On further questioning, the mother recalls that when her son was born he needed frequent echocardiograms, however, he has not needed one recently. On physical exam, small white macules are noted on the patient's bilateral tibia and on the patient's trunk. What is this patient at risk for due to the location of his likely genetic mutation?

A. Spontaneous pneumothorax
B. Pancreatic tumors
C. Epidermal cysts
D. Renal cysts
E. Macular amyloid

3)



A 78-year-old female with a history of extensive sun exposure presents with blanchable erythema, uneven pigmentation, and multiple dilated small vessels on the nose, cheeks, and periorbital skin, as shown. The patient in the photograph has a social event next week and is interested in decreasing the appearance of redness. What wavelength of laser would be most appropriate to achieve that goal?

A. 10,600 nm
B. 755 nm
C. 2940 nm
D. 1540 nm
E. 532 nm

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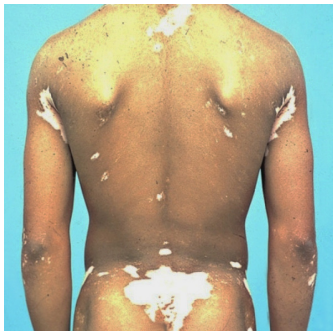
Board Review Answers

1)

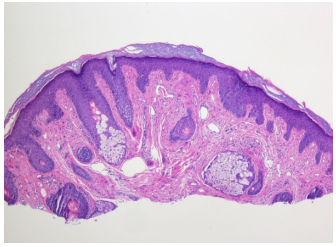


What is the most likely associated ophthalmologic finding?

C. Granulomatous uveitis

The patient has Vogt Koyanagi Harada syndrome, which has two phases. The first phase is meningoencephalitis and the second phase is the ophthalmic auditory phase where the patient experiences deafness, poliosis, alopecia, vitiligo and granulomatous uveitis. As the patient can become blind from the granulomatous uveitis, it is very important to send the patient for ophthalmologic exam. Alezzandrini Syndrome is associated with unilateral vitiligo, deafness on the ipsilateral side, and unilateral vision changes. Waardenberg syndrome is associated with dystopia canthorum, heterochromic irides and a white forelock. Deafness is most common in Waardenburg type 2. Ziprowski Margolis syndrome is associated with hypopigmented macules, congenital deafness and heterochromic irides.

2)



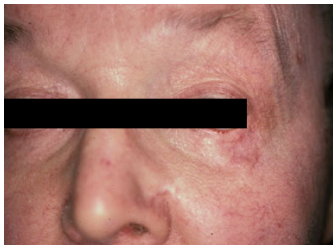
What is this patient at risk for due to the location of his likely genetic mutation?

D. Renal cysts

The pathology shows an angiofibroma with dilated capillaries close to the epidermis and a fibrous stroma. The three syndromes with numerous facial angiofibromas, include tuberous sclerosis, Birt Hogg Dube and MEN 1. The patient in the question has facial angiofibromas, was born with a cardiac rhabdomyoma that resolved with age, and hypopigmented macules and pretibial confetti macules. Thus, the most likely diagnosis is tuberous sclerosis. The two mutations for tuberous sclerosis are TSC1/hamartin and TSC2/tuberin. The tuberin gene is located very close to the gene for polycystic kidney disease; therefore, patients with tuberous sclerosis due to a tuberin mutation are at an increased risk for having multiple renal cysts.

Birt Hogg Dube is a mutation in folliculin and is associated with spontaneous pneumothorax, lung cysts and renal cell carcinoma. MEN 1 is due to a mutation in menin and is associated with angiofibromas and hypopigmented macules, but not congenital cardiac rhabdomyomas. Patients with MEN 1 also present with café au lait macules, peptic ulcer disease and pancreatic tumors including insulinomas and gluagonomas. Patients with Gardner syndrome, which is due to a mutation in the APC gene, get epidermal inclusion cysts, congenital hypertrophy of the retinal epithelium and adenomatous polyps in their GI tract. Lastly, macular amyloid is associated with MEN IIa which is due to a mutation in RET, and those patients often have parathyroid tumors and pheochromocytomas.

3)



What wavelength of laser would be most appropriate to achieve that goal?

E. 532 nm

The patient's primary concerns are redness, due to telangiectasias and small venules, and minimal downtime. The best laser in this case is the KTP 532nm laser. The pulse dye laser at 585nm is also an excellent choice to treat vascular lesions. If the venules were larger, the long pulse 1064 Nd:YAG laser would be a good choice. The carbon dioxide laser at 10,600 nm and the Erbium:YAG at 2940nm are both lasers used for ablative skin resurfacing, and although they improve redness and pigment abnormalities, among other benefits, the recovery is more involved. The Alexandrite 755nm laser, when used with Q-switched settings, is excellent for pigmented lesions, and when used in normal mode, it is good for laser hair removal. The Erbium: glass laser is 1540nm and is used for fractional non-ablative skin resurfacing.

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