Boards’ Fodder celebrates 10 years!

By Dean Monti, AAD managing editor, special publications

The idea was pitched 10 years ago in a Resident/Fellows Committee Meeting in February 2002, during the Academy’s Annual Meeting in New Orleans. It was suggested that the Academy find new ways to alert dermatology residents to areas that were “high yield” for the mock and real board examinations. The answer was to provide charts of commonly asked and “highly askable” factoids relating to dermatology. Boards’ Fodder had its premiere in AAD’s Resident Roundup (now Directions in Residency) in fall 2002 with “Genes to Know,” by Benjamin Solky, MD, and Brian Selkin, MD. Since that time, Boards’ Fodder has proven to be one of the most popular features of this publication and a valuable study tool for residents for the past decade — indeed, the publication’s longest running feature.

Considering that our audience changes every four years, it is notable that new residents have continued to carry the torch forward. Dr. Solky and his colleagues contributed to the feature through its nascent years, followed by Sharon Jacobs, MD, for several years. Then, somewhere along the line, word got around: Boards’ Fodder was an opportunity for all up-and-coming residents to share information with other residents and showcase their individual resident talent in an Academy publication. As Boards’ Fodder enters a new decade, contributions to the column are no longer the voice of one resident, but many, and contributions are on the rise. To celebrate the occasion, we are presenting three times the Fodder for this special issue.

Thank you all for your contributions over the years, which are helping us build an archive of study materials for current and future residents. As we forge ahead, Directions in Residency is working toward ways to bring this feature to you in new formats and, with your help, more often.

additional resources

More resources than ever to help with boards study

If there’s one thing we’ve discovered over the years, it’s that residents can’t get enough boards study materials. Recently the Resident/Fellows page at aad.org was updated and includes a new section, “Board prep,” containing boards study resources.

The AAD offers a variety of resources to help you prepare.

- View the Boards’ Fodder section in current and past issues of Directions in Residency, a quarterly publication for residents.
- Listen to Dialogues in Dermatology, the Academy’s monthly audio subscription program in which leading dermatologists discuss current clinical issues.
- Read Derm Clips, a monthly publication that includes evidence-based summaries of clinical content from a broad range of medical journals.
- Watch Boards Blitz for two hours of board prep insight, tips, and resources that you can view while studying.
- Review the AAD’s online Medical Student Core Curriculum to cover the basics of dermatology when preparing for the boards.
- Explore peer-recommended boards study tools that include books, study guides, websites, and quizzes.
- Board prep courses offered at AAD meetings.

You can also learn more about boards prep on the American Board of Dermatology site, www.abderm.org.

Directions in Residency is also looking for new ways to bring board materials to you. If you have not already done so, please take the Directions in Residency reader survey (see page 8).
### Oral Disease, Part 2

*by Helena Pasieka, MD*

<table>
<thead>
<tr>
<th>CLINICAL</th>
<th>PATHOLOGY</th>
<th>TREATMENT</th>
<th>ASSOCIATIONS</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>SALIVARY GLAND DISEASES</strong></td>
<td></td>
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<tr>
<td>Mucocele</td>
<td>Painless submucosal swelling. Color ranges from clear/blue/ colorless depending on depth of lesion. Lower labial mucosa most common, but can occur anywhere where there are minor salivary glands.</td>
<td>Collection of mucus surrounded by macrophages and granulation tissue. Look for the inflamed minor salivary gland as a clue.</td>
<td>Superficial mucoceles often resolve w/o intervention by spontaneous rupture. However, most require surgical excision.</td>
</tr>
<tr>
<td>Cheilitis glandularis</td>
<td>Ranges from slight hypertrophy of lower lip to nodular enlargement with eversion. Most commonly in adult men.</td>
<td>Localized dense accumulations of inflammatory cells within and around the mucous glands in a background of actinic cheilitis.</td>
<td>Vermilionectomy of the lower lip, with or without cosmetic debulking is the standard of care. Injections of corticosteroids can provide symptomatic relief.</td>
</tr>
<tr>
<td>Sjögren’s syndrome</td>
<td>Slow onset of variable degrees of eye and oral dryness. Increased dental caries and difficulty wearing dentures. Increased candidiasis.</td>
<td>Salivary glands with focal aggregates of &gt;50 lymphocytes adjacent to normal-appearing acini. In the parotid gland characteristic epimyoepithelial islands are seen.</td>
<td>Symptomatic care and management of the associated complications. Salivary stimulation (sugarless gum, hard candies, pilocarpine) or artificial saliva.</td>
</tr>
<tr>
<td>Salivary gland tumors</td>
<td>Submucosal, painless, rubbery firm swelling often noted on the posterior hard palate or anterior soft palate.</td>
<td>Varies depending on the specific type of salivary tumor.</td>
<td>Benign salivary gland tumors are conservatively excised. Malignant tumors are more extensively removed +/- radiotherapy as adjunct.</td>
</tr>
<tr>
<td><strong>HEMATOLOGIC/ONCOLOGIC DISEASE</strong></td>
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<tr>
<td>Chemotherapy-induced mucositis</td>
<td>Multiple oval or irregularly shaped ulcers, usually on the gingivae, lateral tongue, or buccal mucosa. Usually appear 4-7 days after chemotherapy administered.</td>
<td>Lichenoid mucositis.</td>
<td>Usually resolves in 2-3 weeks cessation of chemotherapy. Palifermin (keratinocyte growth factor) may reduce severity for those on high-dose chemotherapy. Meticulous oral hygiene and symptom management.</td>
</tr>
<tr>
<td>Leukemia</td>
<td>Many oral manifestations, most commonly bruising or hemorrhage related to thrombocytopenia. Also, pallor of anemia, increased viral, fungal and bacterial infections related to leukopenia. Diffuse, firm, non-tender gingival enlargement can be caused by infiltration of the gingival connective tissue by leukemic cells.</td>
<td>Infiltration of leukemic cells into gingival connective tissue (most commonly with monocytic or myelomonocytic leukemias).</td>
<td>Multi-agent chemotherapy and peripheral blood stem cell or bone marrow transplantation are most commonly used to treat acute leukemia.</td>
</tr>
<tr>
<td>Lymphoma</td>
<td>Slow growing, painless, soft or rubbery, with purplish swelling. Most commonly on the palate and the buccal vestibule. Overlying telangiectasia sometimes seen. Ulceration possible, mimicking SCC.</td>
<td>Infiltration of atypical lymphocytes, usually of B-cell type.</td>
<td>Chemotherapy and/or monoclonal antibodies (e.g. rituximab). HIV, other immunosuppression, older age.</td>
</tr>
<tr>
<td>Melanoma</td>
<td>Most commonly on hard palate, maxillary attached gingiva. Has same features as cutaneous melanoma.</td>
<td>Proliferation in epithelium and infiltration of the connective tissue by atypical melanocytes, with or without melanin production. Can use Melan-A or S100 to stain.</td>
<td>Wide surgical excision with +/- margins. Sentinel lymph node biopsy for prognostication. Minimal radial growth phase on mucosa, so they differ from cutaneous melanomas in that they present in the vertical growth phase. Chemotherapy and XRT of little utility. Worse prognosis than cutaneous lesions, w/ 5-year survival rate of ~15%, and median from dx of &lt; 2 years.</td>
</tr>
</tbody>
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*Helena Pasieka, MD, is a second year resident in the department of dermatology at Johns Hopkins University.*
Oral Disease, Part 2 (continued)

by Helena Pasieka, MD

**MANIFESTATIONS OF SYSTEMIC DISEASE**

<table>
<thead>
<tr>
<th>CLINICAL</th>
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<th>TREATMENT</th>
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<tbody>
<tr>
<td>Amyloidosis</td>
<td>Firm macroglossia, often with scalloped edge. Xerostomia or dysgeusia can be seen before the onset of tongue enlargement.</td>
<td>Homogeneous eosinophilic accumulation. (+) congred stain.</td>
<td></td>
</tr>
<tr>
<td>Pernicious anemia</td>
<td>Gradual onset of smooth, ‘beefy-red’ tongue, will-defined areas of erythema which can coalesce into diffuse dorsal tongue involvement causing a smooth, beefy-red appearance.</td>
<td>Absence of filiform papillae.</td>
<td>Vitamin B12 injections provide rapid improvement.</td>
</tr>
<tr>
<td>Crohn’s disease</td>
<td>Linear fissures +/- ulcers of the vestibule or “cobblestone” ulcers of the buccal mucosa. Sometimes scarring. Can also have cheilitis granulomatosa both clinically and histologically.</td>
<td>Non-necrotizing granulomatous inflammation.</td>
<td>Respond to therapy for bowel lesions. Topical or intralational corticosteroids also effective.</td>
</tr>
<tr>
<td>Pyostomatitis vegetans</td>
<td>“Snail track” arrangement of multiple tiny, creamy-yellow pustules set against a bright erythematous background. Fragile pustules lead to shallow erosions and ulcerations. Labial, gingival and buccal mucosa are most commonly involved; tongue is usually spared.</td>
<td>Intra- or subepithelial microabscesses containing eosinophils and neutrophils.</td>
<td>Management of the underlying GI disease often results in improvement of oral lesions. Inflammatory bowel disease.</td>
</tr>
</tbody>
</table>

**HIV**

| Kaposi’s sarcoma | Multiple violaceous macules, plaques, or nodules. Most commonly on palate, but can be anywhere in mouth. | Infiltration of the dermis w/slit-like vascular spaces, and dilated vessels. Many extravasated RBCs and proliferating spindle cells seen. | May improve or resolve with improved immune status (i.e., initiation of HAART). |
| Oral hairy leukoplakia | White, shaggy, corrugated protrusions on lateral tongue. Cannot be dislodged with tongue depressor. | Irregular keratin projections, parakeratosis, acanthosis, and groups of pale epithelial cells. | Antiretroviral therapy may lead to regression. Epstein-Barr virus in HIV. Predictor of rapid decline and progression to AIDS. |
| Candidiasis | Thick white or cream-colored deposits on tongue or posterior oropharynx w/“cottage cheese-like” appearance. Can be dislodged with tongue depressor. Also can have tissuing at the corners of the mouth. | Pseudohyphae and budding yeast sometimes seen in H&E. More easily seen with PAS or GMS stain. | Improvement of immune status. Anti-candidal treatment, such as clotrimazole troches or PO fluconazole. |
| HSV | Grouped vesicles on erythematous base, becoming ulcerated. Can coalesce into larger lesions. Look for scalloped border. | Epidermal necrosis and ballooning degeneration. Infected cells are multinucleated w/glassy nuclear appearance. | Suppressive therapy with antiviral medications. Severely immunosuppressed may need IV acyclovir. |
| CMV | Ulcerations anywhere in mouth. Appear like aphthae, may be slightly larger. | Vascular dilatation with large cytoplasmic endothelial cells. *Owls eye* appearance due to halo around intranuclear inclusion bodies. | Antiviral drugs (ganciclovir and valganciclovir). Development on mucosal surface usually a sign of disseminated disease. |

**SYNDROME ASSOCIATIONS**

| Odontogenic keratocysts of the jaw | Often incidentally noted radiolucent or mixed radiolucuent/radiopaque lesions of the mandible. Asymptomatic; rarely mild facial swelling and discomfort. | Lining of stratified squamous epithelium with basal layer of the epithelium exhibiting palisaded cuboidal to columnar cells. The luminal surface often with a corrugated morphology and parakeratosis. | Opinions differ and range from wide local excision to marsupialization to curettage. Neviod basal cell carcinoma syndrome mutations in PTCH (Hedgehog signaling pathway). |
| Multiple osteomas of the jaw | Asymptomatic facial deformity. | Same as typical solitary osteomas. | Surgical removal, screening for malignancy. Often the earliest marker of Gardner syndrome occurring >80% around puberty. |
| Multiple endocrine neoplasia syndrome type 2B | Multiple mucosal neumomas involving lips and tongue are often the first dermatologic manifestation. | “Plexiform neumomas” of hyperplastic bundles of nerves surrounded by a thickened perineurium. | Screening for malignancy: Medullary thyroid carcinoma with pheochromocytoma in 50% of cases, and digestive neurofibromatosis. |

**Sources:**
Special thanks to Dr. Gary Warnock.
### Inborn Errors of Metabolism

**by Kristina Burke, MD, and Erin Adams, MD**

<table>
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<th>DEFECT</th>
<th>SKIN FINDINGS</th>
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<tr>
<td><strong>Alkaptonuria</strong>&lt;br&gt;(Endogenous ochronosis)&lt;br&gt;AR</td>
<td>Homogentisic acid oxidase&lt;br&gt;Disorder of phenylalanine and tyrosine metabolism. Homogentisic acid accumulates</td>
<td>Blue-grey pigmentation of nose, ears, axillae, genitalia and cartilage. Blue sclera (Osler’s sign), dark urine (pH &gt;7), black cerumen</td>
<td>- Large joint arthropathy&lt;br&gt;- Intervertebral disc calcification&lt;br&gt;- Mitral/aortic valve disease&lt;br&gt;- Nephrolithiasis</td>
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<tr>
<td><strong>Fabry’s disease</strong>&lt;br&gt;(Angiokeratoma Corporis Diffusum)&lt;br&gt;XLR</td>
<td>α-galactosidase A&lt;br&gt;(glycolipids accumulate in skin, heart, kidneys)</td>
<td>Angiokeratomas (esp lower extremities, scrotum, penis, and lower trunk), wool-like cornal opacities, edema, hypohidrosis</td>
<td>- Renal failure, cardiovascular events, strokes&lt;br&gt;- Acroparesthesias and painful crises&lt;br&gt;- Maltese cross in urine&lt;br&gt;- Enzyme replacement available</td>
</tr>
<tr>
<td><strong>Fucosidosis</strong>&lt;br&gt;AR</td>
<td>α-L-fucosidase</td>
<td>Angiokeratomas, coarse features, facial dysmorphism</td>
<td>- Mental retardation (MR), neurologic deterioration</td>
</tr>
<tr>
<td><strong>Gaucher disease</strong>&lt;br&gt;AR</td>
<td>Acid-β-glucosidase (glucocerebrosidase)&lt;br&gt;leads to accumulation of glucocerebroside in histiocytes (Gaucher’s cells)</td>
<td>Type 1: diffuse hyperpigmentation, petechiae, pungueulae of sclera&lt;br&gt;Type 2: congenital ichyiosis, collodion baby</td>
<td>ALL: hepatosplenomegaly (HSM)&lt;br&gt;- Type 1: Adult type, Ashkenazi Jews, no CNS involvement&lt;br&gt;- Type 2: infantile, rapid neuro deterioration, aspiration pneumonia&lt;br&gt;- Type 3: juvenile chronic neuropathic</td>
</tr>
<tr>
<td><strong>Phenylketonuria</strong>&lt;br&gt;AR</td>
<td>Phenylalanine hydroxylase (Phenylalanine not oxidized to tyrosine)</td>
<td>Pigmentary dilution of skin, hair, eyes (fair complexion, blond hair, blue eyes), pseudosclerosis, eczematosus dermatitis</td>
<td>- MR, seizures&lt;br&gt;- Phenylpyruvic acid in urine (musty odor)&lt;br&gt;- Screened for at birth&lt;br&gt;- Dietary restriction</td>
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<tr>
<td><strong>Tyrosinemia II</strong>&lt;br&gt;(Richner-Hanhart)&lt;br&gt;AR</td>
<td>Tyrosine aminotransferase (hepatic)&lt;br&gt;TAT gene</td>
<td>Painful palmoplantar keratoderma</td>
<td>- Herpetiform keratitis, blindness&lt;br&gt;- MR&lt;br&gt;- Corneal ulcers</td>
</tr>
<tr>
<td><strong>Homocystinuria</strong>&lt;br&gt;AR</td>
<td>Cystathionine β-synthase</td>
<td>Fair complexion, malar flush, livedo reticularis, leg ulcers&lt;br&gt;Sparse, fine hair&lt;br&gt;Marfinoid habitus</td>
<td>- Thromboembolic events (50% by 30yo)&lt;br&gt;= common cause of death&lt;br&gt;- Ectopia lentis (downward)&lt;br&gt;- Osteoporosis&lt;br&gt;- MR, developmental delay</td>
</tr>
<tr>
<td><strong>Niemann-Pick disease</strong>&lt;br&gt;AR</td>
<td>Type A and B: Sphingomyelinase (SMPO1)&lt;br&gt;Type C: NPC1 and 2&lt;br&gt;A lysosomal storage disease</td>
<td>Type A and B: ochre to brownish-yellow discoloration of skin, papular lesions face and upper extremities, xanthomas&lt;br&gt;Type A: severe, CNS deterioration, HSM, failure to thrive&lt;br&gt;Type B: spares CNS, survival to adulthood&lt;br&gt;Type C: childhood, HSM, developmental delay, psychomotor deterioration</td>
<td>- All: hepatosplenomegaly (HSM)&lt;br&gt;- Type A: CNS deterioration, HSM, failure to thrive&lt;br&gt;- Type B: spares CNS, survival to adulthood&lt;br&gt;- Type C: childhood, HSM, developmental delay, psychomotor deterioration</td>
</tr>
<tr>
<td><strong>Trimethylaminuria</strong>&lt;br&gt;“Fish odor syndrome”&lt;br&gt;A lysosomal storage disease</td>
<td>Mutation of flavin-containing monooxygenase type 3 (FMO3) gene</td>
<td>Skin, urine, and sweat smell like “rotting fish”</td>
<td>- Smell due to accumulation of trimethylamine&lt;br&gt;- Avoid choline in diet</td>
</tr>
<tr>
<td><strong>Lesch-Nyhan</strong>&lt;br&gt;(juvenile gout)&lt;br&gt;XLR</td>
<td>HPRT1 gene leading to hypoxanthine-guanine phosphoribosyl transferase (HGPRT) deficiency</td>
<td>Loss of tissue around mouth and fingers (due to self-mutilation)&lt;br&gt;Tophaceous deposits (hyperuricemia)</td>
<td>- MR, choreoathetoid movements, self-mutilation&lt;br&gt;- Orange crystals in diaper</td>
</tr>
<tr>
<td><strong>Wilson’s disease</strong>&lt;br&gt;(Hepatolenticular degeneration)&lt;br&gt;AR</td>
<td>Defect in ATP7B gene (hepatic copper transporting ATPase)</td>
<td>Blue lunulae, Kayser-Fleischer rings (copper deposition in Descemet’s membrane), greenish discoloration of skin, face and genitalia, pretibial hyperpigmentation</td>
<td>- HSM, cardiomyopathy, renal tubular acidosis&lt;br&gt;- Progressive neurologic dysfunction (dysthria, ataxia, dementia)&lt;br&gt;- Lab: low ceruloplasmin&lt;br&gt;- Tx: penicillamine, trientine, zinc supplement</td>
</tr>
<tr>
<td><strong>Hartnup disease</strong>&lt;br&gt;AR</td>
<td>SLC6A19 gene (neutral amino acid transporter)</td>
<td>Pellagra-like dermatitis (photosensitive eruption on face, arms, neck, legs)</td>
<td>- Cerebellar ataxia, MR&lt;br&gt;- Tends to improve with age&lt;br&gt;- Defect in tryptophan transport</td>
</tr>
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Inborn Errors of Metabolism (continued)

by Dr. Burke, MD and Dr. Adams, MD

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<tr>
<td>Prolidase deficiency</td>
<td>Deficiency of the enzyme prolidase</td>
<td>Skin fragility, lower extremity ulceration, telangiectasias, poliosis</td>
<td>Mental deficiency, recurrent infections, syndromic facies</td>
</tr>
<tr>
<td>Citrullinemia</td>
<td>Type 1: argininosuccinic acid synthetase (ASS1 gene)</td>
<td>Resembles zinc deficiency</td>
<td>Clears with arginine supplementation</td>
</tr>
<tr>
<td></td>
<td>Type 2: SLC25A13 gene</td>
<td>Erythematous, erosive, scaling patches perioreally, lower abdomen, and diaper area</td>
<td>Procainamide supplements</td>
</tr>
<tr>
<td>Farber disease</td>
<td>A lysosomal storage disease</td>
<td>Periarticular swelling, rubbery SQ nodules</td>
<td>Onset first month of life, death by age 2</td>
</tr>
<tr>
<td>Adrenoleukodystrophy</td>
<td>(Schilder’s disease) X-linked</td>
<td>Hyperpigmentation, mild ichthyosis, sparse hair with trichorhexis nodosa-like features</td>
<td>Progressive demyelination of cerebral white matter</td>
</tr>
<tr>
<td>CADASIL</td>
<td>NOTCH 3 gene</td>
<td>Findings on skin biopsy (eosinophilic granular material in arterial walls)</td>
<td>Depression, migrane headaches, multiple cerebral infarcts leading to early dementia, Most common hereditary stroke disorder</td>
</tr>
<tr>
<td>Lafora disease</td>
<td>(Lafora progressive myoclonic epilepsy)</td>
<td>EPM2A- encoding laforin EMP2B – encodes a ubiquitin ligase</td>
<td>Progressive epilepsy syndrome, dementia and ataxia, Best site to biopsy = axilla (Lafora bodies around eccrine ducts)</td>
</tr>
<tr>
<td>Alagille syndrome AD</td>
<td>JAG 1</td>
<td>- Xanithomas, jaundice - Unusual facies</td>
<td>Congenital intrahepatic biliary hypoplasia / cholesterol and pruritis, Hyperlipidemia, Buttery-shaped vertebra</td>
</tr>
<tr>
<td>Sitosterolemia AR</td>
<td>“phytosterolemia” ABCG5 (encoding sterolin-1) or ABCG8 (encoding sterolin-2)</td>
<td>Tuberous and tendinous xanithomas during the first decade of life</td>
<td>Elevated plasma levels of plant sterols, Arthritis, premature vascular disease, high risk of fatal cardiac events during teenage years</td>
</tr>
<tr>
<td>Hunter syndrome XLR</td>
<td>A lysosomal storage disease</td>
<td>Deficiency of iduronate-2-sulfatase</td>
<td>Dysotosis multiplex</td>
</tr>
<tr>
<td>Tangier Disease AR</td>
<td>(Familial α-lipoprotein deficiency) ATP-binding cassette (ABCA1) transport protein: almost complete absence of plasma HDL and massive deposition of cholesterol esters in tissues</td>
<td>Torsils are yellow and enlarged. Maculopapular eruption over trunk and abdomen</td>
<td>HSM, lymph node enlargement, peripheral neuropathy, corneal infiltration in adults, Premature coronary artery disease</td>
</tr>
</tbody>
</table>

References:

In 2013, the In-Training Examination will be given on Thursday, February 21 in US and Canadian dermatology training programs, and on Monday, February 25 in overseas international dermatology training programs. Information will be emailed to training programs in the fall and the deadline for registering online is November 15. For more information, call the Board office at 313-874-1089.
Race for the Case
By Karolyn Wanat, MD

A 44-year-old woman presented to a community STD clinic with an asymptomatic but growing lesion that occurred after shaving in the suprapubic area. She was otherwise completely healthy. A biopsy was performed.

1) What is the diagnosis?
2) What is the most important histopathologic feature (shown above)?
3) Are there any laboratory abnormalities in these patients?
4) What genodermatoses can these be associated with?

Respond today with the correct diagnosis to Allison Evans, staff editor at the AAD, at aevans@aad.org, and you might win a Starbucks gift card and get your photo in Directions.

In the last issue, we presented a 57-year-old Caucasian man with history of hypertension controlled with medication who presents with abrupt onset of a new pruritic dermatitis. The questions and correct answers are:

1) Diagnosis and what would you expect to see on pathology?: Drug-induced subacute cutaneous lupus erythematosus; on pathology, one would expect to see an interface dermatitis with baso-vascular change, possibly eosinophils

2) Most common medication associated with this eruption? Hydrochlorothiazide

3) HLA most commonly associated with this condition? HLA-DR3

4) In the majority of cases, what is the auto-antibody most commonly associated with this? SSA, anti-Ro

Congratulations to Van Hoang, MD, winner of the summer 2012 Race for the Case! She’s a third-year dermatology resident at the Montefiore Albert Einstein Dermatology Residency Program in New York. Other than studying dermatology factoids (which she obviously excels at!), she enjoys doing yoga, running in Central Park, and trying new restaurants in New York City. Congratulations and thanks for racing with us!

Help make us ‘appy: AAD seeking new mobile apps
The Academy would like to hear from residents about mobile apps that you use professionally or recommend as health care resources for patients, as well as app ideas or apps in development. Please visit www.aad.org/app-info to share the name of the app, what type of app it is (iPhone, iPad, Android), and how you use it professionally.
Eponyms in Dermatology

by Heather Kraly Orkwis, DO

Asboe-Hansen sign = extension of intact blister when pressure is applied to roof, seen in pemphigus vulgaris
Ausitz’s sign = punctate bleeding points within lesion upon scratching, seen in psoriasis
Bazex syndrome = acrokeratosis paraneoplastica (acquired)
Bazex syndrome = follicular atrophoderma, multiple BCCs, hypotrichosis, localized hypohidrosis (X-linked dominant)
Bazin’s disease = erythema induratum, associated with TB
Beckwith-Wiedemann syndrome = exomphalos-macroglossia-gigantism syndrome (p57/KIP2)
Behcet’s disease = triad of aphthous ulcers, genital ulcers, ocular inflammation (+ HLA-B51, Silk Road Disease)
Bloch-Sulzberger disease = incontinentia Pigmenti (NEMO; X-linked dominant)
Bockhart’s impetigo = follicular impetigo
Bourneville’s disease = tuberous Sclerosis (Epiloia) (TSC1, TSC2)
Bowen’s disease = squamous carcinoma in situ
Buruli ulcer = M. ulcerans (named after Buruli region of Nile River, Africa)
Busche-Lowenstein tumor = verrucous carcinoma of glans penis and prepuce (HPV 6, 11)
Busche-Ollendorff syndrome = Dermatofibrosis lenticularis disseminata, osteopetrosis (LEMD3)
Calabar swellings = localized angioedema in tissue from migrating loiasis
Carney Complex = NAME syndrome, LBAM syndrome (PKRKA1)
Carvaljal syndrome = left sided cardiomyopathy, woolly hair, keratoderma (DESMOLPLAKIN)
Cobb syndrome = Cutsomeningsopinal angitis
Civatte bodies = degenerated, apoptotic keratinocytes seen in lichen planus
Conradi-Hünermann syndrome = X-linked dominant chondrodysplasia punctata
Crowe’s sign = axillary or inginal freckling seen in neurofibromatosis
Darier disease = Keratosis follicularis (ATP2A2)
Darier’s sign = urtication following rubbing of macule/papule in mastocytosis (urticaria pigmentosa)
Dennie-Morgan lines = crescentic creases of lower eyelids due to stagnation of venous blood, seen in atopic dermatitis
Degas’ disease = malignant atrophic papulosis
Dercum disease = adiposis dolorosa, mostly obese menopausal women, consisting of multiple equisitely tender lumps
Gianotti-Crosti syndrome = Papular acrodermatitis of childhood
Goltz syndrome = Focal dermal hypoplasia (PORCN)
Gorlin syndrome = Nevoid basal cell carcinoma syndrome (PATCHED1)
Gottron’s papules = erythematous eruption over knuckles, elbows, knees, seen in dermatomyositis
Graham-Little-Piccardi-Lasseur syndrome = variant of LPP; cicatricial alopecia of scalp, non-scarring alopecia of axilla and groin, follicular lichen planus eruption
Grover’s disease = transient acantholytic dermatosis
Hailey-Halley disease = familial benign pemphigus (ATP2C1)
Heck’s disease = oral focal epithelial hyperplasia (HPV 13, 32)
Griscelli syndrome = pigmentary distution, T- and B-cell immunodeficiency, recurrent infection, progressive CNS deterioration (MICOSIN Yaj)
Hermansky-Pudlak syndrome = non-transgenerational PPK, esophageal carcinoma (TOC)
Hughes’ triad = Atrophophiloid antibody syndrome (fetal loss, thrombosis, thrombocytopenia)
Hutchinson-Gilford syndrome = Progeria (LAMIN A)
Hutchinson’s sign = pigment in paronychal area suggestive of melanoma
Janeway lesions = nonpainful hemorrhagic macules or nodules of palms and soles, seen in infective endocarditis
Kasabach-Merritt syndrome = consumptive coagulopathy within a kaposiform hemangioendothelioma at tutilged angina
Klippel-Trenaunay syndrome = angio-lymphoid hyperplasia; port-wine stain, soft tissue and bony hyper trophy, venous and lymphatic malformations
Koplik’s spots = small, white spots on erythematous buccal mucosa, seen in early measles
Kveim-Siltzbach test = skin test with human sarcoid tissue injected into a patient suspected of having sarcoidosis; positive results are a sarcoid granuloma at the site
Kyrle’s disease = chronic generalized dermatosis with papules with central keratotic plugs (DM, renal disease)
Leser-Trelat sign = abrupt onset multiple seborrheic keratoses, associated with internal malignancy
Lichtenberg’s figures = branching pattern of cutaneous marks pathognomonic for lightning injury
Louis-Bar syndrome = Ataxia-Telangiectasia (ATM)
Lovibond’s angle = 160° angle between proximal nail fold and the nail plate
Lyell’s syndrome = Toxic Epidermal Nekrolysis
Medulang’s disease = benign symmetric lipomatosis (Launois-Bensuade syndrome, horse-collar appearance)
Maffucci syndrome = superficial and deep venous malformations, enchondromas, chondrosarcoma (PHTHR1)
Majocchi’s disease = purpura annularis telangiectodes
Majocchi granuloma = deep dermoepitope infection of hair follicle
Mal de Meleda = Keratoderma palmoplantaris transgressed (SLURP1)
Marfan syndrome = tall stature, arachno dactyly, ectopia lentis, progressive aneurysmal dilatation of ascending aorta, CHF (FIBRILLIN 1)
Marjolin’s ulcer = aggressive SCC arising in site of chronic injury or burn
McCune-Albright Syndrome = Albright syndrome; “Coast of Maine” cafe-au-lait macule(s), polyostotic fibrous dysplasia, precocious puberty (GNAS1)
Milroy’s disease = congenital lower limb lymphedema (FTL4)
Montgomery syndrome = xanthoma disseminatum
Mucha Habermann disease = Pityriasis lichenoides et varioliformis acuta (PLEVA)
Muckle-Wells syndrome = recurrent fevers and urticaria, progressive deafness, secondary amyloidosis (CRYOPYRIN)
Muir-Torre syndrome = DNA mismatch repair defect, sebaceous tumors, adenocarcinoma of the colon (MLH1, MSH2)
Naxos disease = right sided cardiomyopathy, woolly hair, non-epidermolytic PPK (PLAKOLOBIN)
Netherton syndrome = ichthyosis linearis circumflexa (SPINK5)
Nikolsky’s sign = normal epidermis next circumflexa (SPINK5)
Netherton syndrome = ichthyosis linearis circumflexa (SPINK5)
Netherton syndrome = ichthyosis linearis circumflexa (SPINK5)
Olsen-Weber-Rendu syndrome = Hereditary Hemorrhagic Telangiectasia syndrome (HH1, HH2)
Papillon-Lefèvre syndrome = PPK, keratokystodontia, periodontosis (CATHESPIN C)
Parry-Romberg syndrome = acquired progressive hereditary atrophy (morphia variant)
Refsum syndrome = phytanic acid storage disease (PAHX, PEX7)
Richner-Hanhart syndrome = Tyrosinemia type II (TYROSINASE AINMOTRANSFERASE)
Ritter’s disease = Staphylococcal Scalded Skin Syndrome
Russell’s sign = dorsal hand with dry skin and calluses, seen with bulimia/purging
Schnitzler’s syndrome = nonpuritic urticaria, arthralgias, IgM monoclonal protein
Senear-Usher syndrome = pemphigus erythematosus; variant of P. foliaceous confined to seborrheic sites
Shulman’s syndrome = eosinophilic fasciitis (dry river bed)
Sjögren-Larsson syndrome = ichthyosis with erythroderma, spastic di-tetraplegia with scissor gait, mental retardation, atypical retinitis pigmentosa (FALDH)
Sneddon’s syndrome = Livedo reticularis, HTN, CVA associated with antiphospholipid antibodies
Sturge-Weber syndrome = Encephalotrigeminal angiomatosis
Urbach-Wiethe disease = Lipoid proteinosis (ECM1)
Vohwinkel syndrome = PPK mutagens, Keratoderma hereditaria mutans (CONNEEXIN 26, LORCRIN)
Von Recklinghausen disease = Neurofibromatosis I (NEUROFIBROMIN)
Well’s syndrome = eosinophilic cellulitis, ‘flame figures’ on dermapth
Zinsser-Engman-Cole syndrome = Dyskeratosis Congenita (DYSKERIN)

References

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The ABD Board exam. For many, the last exam of our formal medical education — the last hurdle before embarking on the career we’ve dreamed of for nearly a decade or more. Upon reflection, those are probably the only positive thoughts I have about that exam .... Well, and that it’s behind me!

While the board exam is likely the furthest thing from your mind at the start of residency, as you get to know your new co-residents, new computer systems, not to mention learn an entirely new vocabulary, the seed of the board exam has already been subtly planted. As the months of that first year move along and you’re finally getting comfortable with the daily routine and developing differential diagnoses, the plant suddenly sprouts and the flowers bloom. The ABD — and likely your attendings and/or program directors — begin to not-so-gently remind you that you should be studying toward the in-service examination that will be administered in the late winter or early spring. While daunting at first, the in-service is a good way to gauge your knowledge base and provides a tangible goal for you to study and work towards as you progress through residency.

So now that the seed/plant metaphor is exhausted, let’s get to some useful “deep thoughts” for all the different years of residency from friends and colleagues who recently completed and passed the exam:

“Study regularly and maintain your sanity and you’ll be fine!”

“Make subject folders and organize your notes in advance, so you’re not doing it in the last few months of residency when all you want to just do is study…”

“Images, images, images — get a good atlas early and review the AAD slide set.”

“It’s important to see as many clinical presentations of the same diagnosis as possible...”

“Accumulate a ‘best of’ collection of dermatopathology slides, and look into attending the regional and national courses.”

“Practice the timing with the dermatopathology section — as long as it remains timed — since it can add an unnecessary dimension of stress.”

“Take advantage of journal clubs and other materials made available for reviewing the latest in the literature.”

At the end of the day, we are part of such an amazing specialty. You will pass the boards as long as you take your training and studying seriously — just don’t forget to have fun and enjoy it all! 🌼

Talk back to Directions
A special message from Jeremy Brauer, chair of the Residents/Fellows Committee.

We’re working toward improving and enhancing Directions in Residency, the official AAD publication of the Resident/Fellows Committee. That’s why we hope you take a few minutes to fill out our readership survey, which was sent to all residents last month via email.

We want Directions in Residency to reflect your preferences and reading habits, and so we need your help. With only 15 questions, this short survey will help give us information to use going forward. Additionally, because this is a publication that is both for residents and by residents, it is your opportunity to make suggestions that can influence the future of this publication.

If you have not already done so, please make sure to complete this survey, which will provide the necessary data to help improve the publication. There is also room for your comments and suggestions, and we strongly encourage them.

To access this survey, please go to www.aad.org/directionssurvey. If you have any questions or comments regarding this survey, please feel free to contact the managing editor, Dean Monti, at dmonti@aad.org. Thank you in advance for completing this survey ☺️.