

Not-So-Mellow Yellow

Is it the liver? Cholesterol? Genetics? Whatever the cause, a yellowish cast to a lesion, skin, or eyes is a signal for investigation.

Match the diagnosis to the photo by letter

- a. Necrobiotic xanthogranuloma
- b. Nevus sebaceous
- c. Primary biliary cirrhosis
- d. Adult-onset xanthogranuloma



1. An otherwise healthy 13-month-old boy presents with a well-circumscribed, 3×4-cm, yellow-orange plaque with a verrucous velvety surface on the right side of his posterior scalp. Present since the child's birth, the lesion was initially a flat and smooth hairless plaque. Over time, however, it has thickened and become more noticeable, with a bumpy, greasy consistency.



2. A 40-year-old man presents with tender lesions on his back, abdomen, and thighs of 10 years' duration. His medical history is remarkable for follicular lymphoma treated with chemotherapy and a monoclonal gammopathy of uncertain significance diagnosed 5 years after the onset of skin symptoms. Physical exam reveals numerous irregularly shaped yellow plaques on the back, abdomen, and thighs with overlying telangiectasia. A single lesion is noted to extend from a scar.

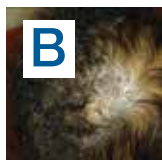


3. A 64-year-old woman presents with complaints of itchy skin and fatigue. The clinician notes that the patient has scleral icterus and jaundice. The patient denies drinking alcohol or using illegal drugs and has no history of hepatitis. On examination, no liver or spleen abnormalities are detected. The clinician orders tests of liver enzymes, hepatitis serologies, and antinuclear and antimitochondrial antibodies.



4. A 66-year-old woman with a history of type 2 diabetes and mild dyslipidemia presents with lesions over the eyelids and cheeks of 10 years' duration. Systemic review is unremarkable. There is no family or personal history of atopy, asthma, or other dermatologic disorders. Physical exam reveals confluent yellowish plaques and nodules over the periorbital regions, as well as yellowish plaques over the neck and back. The lesions are firm to palpation, and the epidermis appears unaffected. The ophthalmic examination is normal, and other mucosal surfaces are unaffected.

ANSWERS



Diagnosis: This solitary scalp lesion is a typical characteristic of **nevus sebaceous (NS)**. Also known as *NS of Jadassohn*, this benign congenital hamartoma of the sebaceous gland frequently involves the scalp and/or face. The classic NS lesion is solitary and appears as a well-circumscribed, waxy, yellow-orange or tan, hairless plaque. Clinically, NS lesions vary in size from 1 cm to several centimeters; they tend to grow proportionately with the child until puberty, when they become notably thicker, greasier, and verrucous or nodular under hormonal influences. The yellow discoloration of the lesion is due to sebaceous gland secretion, and the characteristic color usually becomes less evident with age.

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Diagnosis: Necrobiotic xanthogranuloma (NXG) is a multisystem non-Langerhans cell histiocytic disease characterized by infiltrative plaques and ulcerative nodules. Lesions may appear red, brown, or yellow with associated atrophy and telangiectasia. Although 80% of cases involve periorbital locations, as seen in this patient, the trunk and extremities also may be involved. Researchers have described a predilection for granuloma formation within preexisting scars. Characteristic systemic associations have been reported in the setting of NXG: More than 20% of patients may exhibit hepatomegaly. Hematologic abnormalities, hyperlipidemia, and cryoglobulinemia also may be seen.

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Diagnosis: Lab testing revealed elevated liver enzymes, particularly serum alkaline phosphatase and γ -glutamyl-transpeptidase, and positive antinuclear and antimitochondrial antibodies. A liver biopsy confirmed **primary biliary cirrhosis**. This disease of unknown etiology is characterized by inflammatory destruction of the small bile ducts and gradual liver cirrhosis. It is much more common in women than in men (9:1 ratio). Regardless of the histologic stage of their cirrhosis or their eventual need for liver transplantation, patients with primary biliary cirrhosis typically are treated with ursodeoxycholic acid (13 to 15 mg/kg/d)—a treatment option shown to significantly reduce the incidence of liver transplantation. Bile acid sequestrants (eg, cholestyramine) can be used to treat pruritus.

Originally published in *The Journal of Family Practice* (2014;63[7]). For more information, see <https://bit.ly/2Sw0004>. Text for this case courtesy of Richard P. Usatine, MD. Photo for this case courtesy of Javid Ghandehari, MD. This case was adapted from Smith M, Mathia A. Liver disease. In: Usatine R, Smith M, Mayeaux EJ, et al, eds. *The Color Atlas of Family Medicine*. 2nd ed. New York, NY: McGraw-Hill; 2013:377-385. To learn more about *The Color Atlas of Family Medicine*, see www.amazon.com/Color-Family-Medicine-Richard-Usatine/dp/0071769641/. You can now get the second edition of the *Color Atlas of Family Medicine* as an app by visiting usatinemedia.com.



Diagnosis: Adult-onset xanthogranuloma (AXG) is a rare disease entity, usually manifesting in the third to fourth decades of life. The condition typically presents as a red to yellow-brown nodular cutaneous lesion located on the scalp, face, neck, trunk, or limbs. Most cases (70% to 89%) involve a solitary lesion, but in rare cases (such as that presented), lesions can manifest in multiples or even be disseminated. The lesions of AXG demonstrate a time-dependent progression both clinically and histologically: Early lesions comprise dense monomorphic nonlipid histiocytic inflammatory infiltrates and appear more erythematous. In mature lesions, seen in the case patient, the infiltrate is predominantly composed of lipid-laden histiocytes with associated Touton giant cells; they appear more yellowish on clinical presentation. **CR**

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