Pediatric Ulcerated Nodular Scalp Lesion

DISCERNING DERMATOLOGY

Miriam Kravitz, FNP-BC

Because primary care family nurse practitioners frequently lack access to dermatology specialists, pediatric patients presenting with atypical lesions can be a challenge. For example, Sean was 34 months old when his parents brought him to a clinic with concerns about a scalp lesion. They reported first noticing a small, red, smooth papule when he was about 1 year old. Since their pediatrician assured the couple that "it was nothing" to be worried about, the slowly enlarging nodule had not been further discussed.

It had slowly evolved over time from red to orange-brown and now appeared more yellow. The lesion had ulcerated in its center, which was a disturbing development. Sean showed no signs of discomfort or injury. Both his personal and family health histories were unremarkable. The 1.5-cm, smooth, orange-yellow nodule with central ulceration was buried beneath his thick, curly hair and appeared non-tender to palpation and well demarcated, with no surrounding erythema. He was a smiling, active, well-developed, otherwise well-appearing youngster.

Fortunately, in this case, the differential included relatively few possibilities. On close examination with a simple polarized dermascope, the lesion lacked either the distinctive pattern of a dermatofibroma or the pigmented network or symmetric peripheral pigmented globules of the common Spitz nevus.[1] Rubbing the lesion with a gloved finger produced no urtication, which eliminated a diagnosis of mastocytoma.

After a thorough examination revealed no other lesions, his eyes and vision were evaluated for any sign of ocular involvement. With all systemic findings being negative and signs limited to this solitary lesion, Sean's parents were informed that no biopsy was required to diagnose this common juvenile xanthogranuloma (JXG). The lesion was expected to recede over the next several months without any treatment; however, precautionary referral to a pediatric ophthalmologist was recommended to help rule out a less benign form of JXG. His parents were provided with educational materials and advised to keep monitoring for the possible development of multiple lesions or systemic illness.

Sometimes present at the time of birth, JXGs are non-Langerhans cell histiocytic tumors that may increase in size and number until the child is 2 or 3 years old and usually spontaneously regress by age 6. Epidemiologic data indicate that JXGs are 40% more common in males, with 83%-89.5% being solitary lesions and 10.5%-17% occurring as multiple lesions.[2,3] Lesions in infancy begin as firm, smooth or mildly scaled, round, tan or red papules on the head, neck, or upper trunk and evolve into smooth, dome-shaped nodules or plaques.[4] They may also present in the mouth or other mucous membranes and rarely occur in adults.

JXGs typically become orange-brown to yellow as they progress. They may increase in diameter up to 2.0 cm, with central ulceration commonly occurring in JXGs $\geq$ 1 cm. Although the site of former JXGs may remain discolored or atrophic after regression, systemic involvement is rare, especially in cases of a solitary JXG.[5] Excisional or deep shave biopsies are indicated only in cases requiring diagnostic clarification, symptomatic or multiple skin lesions, or lesions with associated systemic complaints.[5]

Dermatopathology reports confirming early JXGs will describe the dermis containing abundant histiocytes, inflammatory cells derived from monocytes that function as antigen presenting

cells and phagocytes. At later stages Touton giant cells develop, demonstrating multiple nuclei in a wreath-like pattern with foamy peripheral cytoplasm. As JXGs become orange-brown to yellow, the color change is caused by lipidization of the cytoplasm of their histiocytes.[6] Pathology may also report lymphocytes and eosinophils demonstrating inflammation, while fibroblasts and fibrosis indicate lesions in regression.[6] Immunohistochemical staining is positive for factor XIIIa, CD68, HAM56, CD163, fascin, HLA-DR, and CD14 and negative for S100 and CD1a, which can assist clinicians in determining more difficult or confusing diagnoses.[5,7,8]

Ocular exam revealing glaucoma, iris color changes, iris lesions, iris prolapse, or blood within the anterior eye chamber (hyphema) are the most common signs of extracutaneous involvement. Symptoms such as eye redness, photophobia, or complaints of eye irritation could be JXG related. Ocular involvement occurs in less than 0.5% of JXG patients and has been noted only in cases involving multiple lesions.[5] Ocular JXGs require early detection and treatment to prevent blindness.

Rare non-ocular systemic symptoms of JXG, such as Bell's palsy, diabetes insipidus, and lytic lesions of the bones, are more commonly associated with Langerhans cell histiocytosis. This has led some researchers to suggest a common progenitor for both diseases.[7,8]

Other reported sites of systemic involvement include lesions of the lungs, pericardium, liver, spleen, kidney, larynx, testes and other visceral organs, as well as the central nervous system. The possibility of extracutaneous JXG should be thoroughly evaluated by computed tomography and magnetic resonance imaging in patients with multiple lesions demonstrating systemic signs or symptoms.[7] Fewer than 5% of patients with JXG lesions meet the diagnostic criteria of systemic JXG, which is determined by involvement of ≥ 2 visceral organs in addition to multiple cutaneous lesions.[2,3] In such cases specialist referral is imperative, as systemic JXG-related fatalities have been reported.[3]

Despite a documented association between JXG, neurofibromatosis 1 (NF1), and juvenile chronic myelogenous leukemia (JCML); JCML-associated risk remains low.[4,6] No complete blood count surveillance screening of patients with both JXG and NF1 is advised.[4] In those rare cases of ocular or other systemic JXG requiring specialist treatment, effective therapies include excision, corticosteroids, radiation, vinca alkaloids, and other chemotherapies.[2-4,7,8] If any questions or confusion occur, a simple biopsy of the lesion can be sent to dermatopathology.

As with most cases of JXG, Sean's was a benign lesion that regressed spontaneously. The toddler was examined and referred for appropriate ophthalmology evaluation. His parents were educated and empowered to assist with ongoing surveillance. Fortunately, neither biopsy nor treatment was required. Thanks to a basic handheld dermascope, an ophthalmoscope, and nurse practitioner knowledge and experience, this young boy, his parents, and the health care system were spared stress and an unnecessary biopsy. JNP

References

1. Marghoob AA, Braun RP, Kopf AW. *Atlas of Dermoscopy.* New York: Taylor & Francis; 2005.
2. Janssen D, Harms D. Juvenile xanthogranuloma in childhood and adolescence: a clinicopathologic study of 129 patients from the kiel pediatric tumor registry. *Am J Surg Pathol.* 200529(1):21-28.
3. Dehner LP. Juvenile xanthogranulomas in the first two decades of life: a clinicopathologic study of 174 cases with cutaneous and extracutaneous manifestations. *Am J Surg Pathol.* 2003;27(5):579-593.
4. Paller AS, Mancini AJ. *Hurwitz Clinical Pediatric Dermatology E-Book: A Textbook of Skin Disorders of Childhood and Adolescence - Expert Consult.* St Louis: Elsevier Health Sciences; 2011.
5. Bolognia JL, Jorizzo JL, Rapini RP, eds. *Dermatology.* 2nd ed. Spain: Mosby; 2008.
6. Elston DM, Ferringer T, eds. *Dermatopathology.* Philadelphia: Saunders; 2009.
7. Stover DG, Alapati S, Requeira O, Turner C, Whitlock JA. Treatment of juvenile xanthogranuloma. *Pediatr Blood Cancer.* 2008;51(1):130-133.
8. Patrizi A, Neri I, Bianchi F, eds. Langerhans cell histiocytosis and juvenile xanthogranuloma. Two case reports. *Dermatology.* 2004;209(1):57-61.

Miriam Kravitz, MSN, FNP-BC, is a board director for the National Academy of Dermatology Nurse Practitioners and a dermatology resident in the doctorate of nursing practice program at the University of South Florida in Tampa. She can be reached at miriam.kravitz@gmail.com.

1555-4155/12/$ see front matter

doi: 10.1016/j.nurpra.2012.02.012