A 9-year-old girl was referred to dermatology for evaluation of a persistent pinkish bump on her neck, that has been present for at least 6 months. It is not itchy or painful. It doesn’t crust, but it might have bled. She has no other similar lesions, and does not recall antecedent trauma. She is a red-headed, green-eyed girl with freckles and fair skin that burns easily in the sun. Her skin does not tan, and she describes multiple mild sunburns, and at least one very painful sunburn. She wears sunscreen on vacation and most weekends when she is going to be outdoors. She knows that her grandparents have been treated for skin cancer, and that her mother has had multiple funny moles biopsied. She does not know of any family history of melanoma.

What is your diagnosis?

A. Pyogenic granuloma  
B. Spitz nevus  
C. Juvenile Xanthogranuloma  
D. Pediatric melanoma
ANSWER and EXPLANATIONS

For teaching purposes, the answer to this case is D - Pediatric Melanoma (*Of note, Spitz nevi are much more common and this photo does look like a possible JXG)

Pyogenic Granuloma

This is a solitary, benign, red bump that develops quickly (weeks) and bleeds easily. It is vascular growth that arises spontaneously, but can be associated with trauma or medication. It is more common in children, and usually presents on the fingers or face. It is not pigmented. Treatment involves removing the lesion and cauterizing the vessels. Recurrence is common.

Spitz Nevus

This is a single, <10mm dome-shaped, firm, pink-red papule that usually develops in children under 10 years of age, generally on the head/neck or extremities. Spitz nevi that develop after the age of 20-30 years of age are more likely to develop malignancy. Histologically, it is comprised of spindled and epithelioid melanocytes. New genomic tests demonstrate an HRAS mutation in 30% of Spitz nevi, which is rare or absent in melanoma. While they were originally described as ‘benign melanoma in children’, common Spitz nevi are benign and can spontaneously regress. There is a conundrum regarding treatment among pediatric dermatologists, but at least half will recommend excision with a minimal margin. Larger lesions or ones with atypical pigment, ulceration, itch or bleeding should be removed, as atypical Spitzoid tumors or Spitzoid melanomas do occur.

Juvenile Xanthogranuloma

This is a benign, firm or rubbery, <10mm yellow-red-brown bump(s) that usually appears at birth or within the first year of life. It presents on the head/neck, arms or trunk, and will usually resolve spontaneously within 6 months-6 years. They can develop in older children or adults. While this is usually an isolated finding, look for café au lait macules to rule out neurofibromatosis. If the child is under 2 years of age and has multiple lesions, or has lesions on or around the eye, an ophthalmologic exam is warranted.

Pediatric Melanoma

Incidence: Melanoma accounts for up to 3% of all pediatric cancers. It is actually quite rare before puberty, and has an overall US childhood incidence of 5-6 per million. However, melanoma is rising about 2% per year in the 15-19-year-old age group, and about 500 cases are expected to be diagnosed in children the US this year. Treatment is delayed in up to 40% of cases of pediatric melanoma because of misdiagnosis or lack of suspicion. Melanoma is the second most common cancer in young adults aged 15-29 years, and is the most common cancer in 25-29 year olds. It is estimated that almost 145,000 new cases of melanoma (76,000 invasive melanoma cases) will be diagnosed in the US in 2016.

Causes: While the causes of pediatric melanoma are not entirely known, those that present as conventional, pigmented, adult-type melanoma demonstrate similar UV-induced DNA damage and specific UV-induced BRAF mutations. Up to 90% percent of adult-type melanomas are caused by UV exposure, and genomic studies are suggesting that UV exposure may play a role
in pediatric cases, particularly post-puberty. Just one blistering sunburn in childhood can double the risk for developing melanoma, whereas 5 severe sunburns between the ages of 15-20 years increase the risk by 80%. Just using a tanning bed before the age of 30 will increase one’s risk for developing melanoma by at least 70%. Indoor tanning is the most likely factor contributing to the increased rate of melanoma in 15-29 year olds, particularly females. Twelve US states now prohibit minors under 18 years of age from using tanning beds.

**Risk Factors:** Markers for increased risk of developing melanoma include: >50 nevi, history of multiple atypical nevi, personal or family history of melanoma or multiple atypical nevi (atypical nevi syndrome). Compounding features include: white skin, red hair, light eyes, history of significant UV exposure, history of severe sunburns. Other risk factors include: photosensitizing conditions or medications, immune suppression, syndromes of defective repair of UV-induced DNA damage (xeroderma pigmentosum), history of other malignancy, and large/giant congenital melanocytic nevi.

**Sun safety measures:** Pediatricians are in a prime position to discuss sun protection and should try to do so at every visit. Sun avoidance during peak sun hours (10am-3pm) in order to reduce skin exposure to damaging ultraviolet light is key to prevention of sunburns, skin cancer and melanoma. Parents should help children apply sunscreen every morning, and teachers should give time for kids to re-apply it before mid-day recess. Lightweight, long-sleeved shirts and longer shorts/pants will protect the body; sunglasses and brimmed-hats will shield eyes and keep sun off of faces, necks and ears. Schools should incorporate sun safety programs. They should enforce hats and sunscreen during recess and provide shaded play areas in the school yard. Routine, daily sunscreen use can decrease the risk of melanoma by up to 50%.

**Sunscreens:** Sunscreens should read broad-spectrum (UVA+UVB) with an SPF factor of at least 30. A shot glass (1 oz) volume is needed to coat the sun-exposed skin of an adult; use at least half of that for your child. Sunscreen should be applied 15 minutes before sun exposure and re-applied after sweating and swimming. Physical (mineral, inorganic) sunscreens are less irritating and best for sensitive skin; they work by deflecting UV rays as they reach the skin. When micronized, the minerals also absorb the UV radiation and absorb heat. Chemical (organic) sunscreens contain ingredients like avobenzone, which absorb, break down and dissipate UV radiation as it hits the skin. It can actually make the skin feel warmer! Since the chemical UVA coverage begins to break down after 2 hours, those sunscreens should be re-applied every 2 hours when spending time outside. Sunscreen is safe for use in children above age 6 months, but sun avoidance is always safest. Small amounts can be safely applied to exposed skin (face, dorsal hands) in infants under age 6 months (AAP approved). To date, studies have not demonstrated that oxybenzone causes endocrine disruption in humans, that retinyl palmitate induces skin cancers, or that micronized minerals (titanium dioxide, zinc oxide) can penetrate deep enough into the dermis to cause skin cancer.

**The ‘new’ Pediatric ABCDE:** Classic monitoring for adult melanoma includes the ABCDEs: A – Asymmetry of lesions, B - Border irregularity, C - Color variance or changes, D – Diameter >0.6cm or obvious growth, and E – Evolution, aka any changes over time. However, because lesions in children will usually grow with them and because they are often non-pigmented, this adult pneumonic can miss 60% of pediatric melanoma and 40% of adolescent melanoma. Instead, consider this revised ABCDE guide: A – Amelanosis (non-pigmented), B – Bleeding (or ulcerated),
Bumpy, C – Color variance, D – De novo, any Diameter. Parents should be encouraged to examine their children’s moles once a month to monitor for changes. Full skin exams should be performed by a physician (pediatrician, family physician, dermatologist) once a year, particularly if the child has many moles, has moles that are difficult to follow and/or if there is a family history of melanoma. A dermatologist should be consulted if there is a specific concern.

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