A fair-skinned seven-year-old girl presented to her physician because her parents were concerned about a lazy eye. She had no eye trauma, pain, or vision changes.

Physical examination revealed an asymmetric, hyperpigmented lesion that was isolated to the iris of her left eye (Figure 1) and was not present the year before. The examination was otherwise normal.

**Question**

Based on the patient’s history and physical examination findings, which one of the following is the most likely diagnosis?

- A. Iris freckle.
- B. Iris melanoma.
- C. Iris nevus.
- D. Primary iris stromal cyst.

See the following page for discussion.
The answer is B: iris melanoma. Iris melanomas are a type of uveal melanoma. They are uncommon and account for approximately 5% of uveal melanomas.1 The lesions are asymptomatic and typically identified during a routine examination with a primary care physician. Iris melanomas are the most common primary malignancy of the iris.2 The mean age of presentation is 40 years. There is no predilection for sex, but they are more common in persons with fair complexions. Iris melanomas can be circumscribed or diffuse. Circumscribed melanomas arise in the inferior iris in approximately 80% of cases. Iris melanomas are unilateral and most are shades of brown, although they may also be yellow or tan.1,2

Using an ophthalmic microscope, an ophthalmologist can definitively diagnose an iris melanoma by identifying prominent tumor vascularity and tumor seeding within the anterior chamber or increasing size over time.2,3 Biopsies may be performed if further confirmation is needed. Treatment is limited for iris melanomas. Small, circumscribed lesions may be closely observed with serial examinations. Discrete lesions that show growth over time should be excised surgically. Large or diffuse tumors often require enucleation. Alternative treatments include plaque brachytherapy and proton beam therapy. Iris melanomas have malignant potential with a 2.6% rate of metastasis.2

Iris freckles are flat, discrete areas of light- to dark-brown pigmentation on the surface of the iris. They are common and have no known malignant potential. Unlike iris melanomas and nevi, iris freckles do not distort the architectural appearance of the stroma.3

Iris nevi are solid brown lesions that often simulate iris melanoma. These nevi penetrate into the iris stroma, causing distortion of the stroma. This can result in ectropion (outward turning of the lid margin) and corectopia (displacement of the pupil). The mean tumor diameter is 3 mm. They are most common in white females and persons with light-colored irises.4

Primary stromal cysts of the iris are rare. Most appear as dome-shaped translucent masses that arise from the peripheral or midportion of the iris. They may appear lobulated. Most cases are reported in the first year of life, and they are considered congenital.1

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References