Question 16 – Week of November 19

Which of the following statements regarding Peutz-Jeghers syndrome is incorrect?

A. It is autosomal dominantly inherited.
B. The classic mucocutaneous melanin pigment spots are seen in the axilla and groin.
C. A high risk for both GI and non-GI cancers is known to be part of this condition.
D. The polyps are most commonly found in the small intestine.
E. The polyps exhibit an arborizing pattern of growth with muscularis mucosae extending into branching fronds of the polyp.

Answer: B

Explanation: Peutz–Jeghers syndrome (PJS) is an autosomal dominantly inherited syndrome of histologically distinctive hamartomatous polyps of the GI tract and characteristic mucocutaneous pigmentation. Its incidence is estimated at 1 in 120 000 births. A high risk for both GI and non-GI cancers is known to be part of this condition. The mucocutaneous melanin pigment spots are seen in more than 95% of cases. They are 1–5 mm in diameter and most commonly occur in the perioral and buccal areas (94%). The GI polyps occur in 88%–100% of patients. Their frequency by segment is: stomach, 24%; small bowel, 96%; colon, 27%; and rectum, 24%. They are nondysplastic, have normal overlying epithelium specific to the GI site in which they are found, and exhibit an arborizing pattern of growth with muscularis mucosae extending into branching fronds of the polyp.

Reference:

Yamada pg. 1653