Question 13 – Week of February 1

This autosomal dominant syndrome is associated with cutaneous sebaceous neoplasms and multiple primary malignancies, most commonly adenocarcinoma of the proximal colon.

A. Gardner's syndrome
B. Peutz-Jeghers syndrome
C. Cronkite-Canada syndrome
D. Osler-Weber Rendu syndrome
E. Muir Torre syndrome

Answer: E

Muir Torre syndrome is likely part of the Lynch II syndrome, associated with a high prevalence of sebaceous neoplasms. It is also associated with multiple polyps of the intestines, as well as urogenital carcinomas. Gardner's syndrome, or familial adenomatous polyposis, is an autosomal dominant syndrome associated with a mutation in the APC gene. Findings are of multiple intestinal polyps along with cutaneous epidermoid cysts, jaw osteomas, and supernumerary teeth. Peutz-Jeghers syndrome includes GI hamartomas and mucocutaneous pigmentation. These patients are at risk for multiple malignancies, including duodenal carcinoma and ovarian tumors. Cronkite-Canada syndrome is a rare syndrome of GI polyposis, mucocutaneous hyperpigmentation, alopecia, malabsorption, and nail dystrophy. Osler-Weber Rendu syndrome, also known as hereditary hemorrhagic telangiectasia, is characterized by telangiectasias, AVM's, and aneurysms of the skin, lung, brain, and GI tract.