

Question 18 – Week of March 8

All of the following are associated with Cowden's disease except:

- A. Trichilemmomas
- B. Multiple gastrointestinal hamartomas
- C. Gingival cobblestoning
- D. Epidermoid cysts
- E. Sclerotic fibromas

Answer: D

Cowden's disease is a multiple hamartomatous syndrome inherited in an autosomal dominant fashion. Mucocutaneous findings are of oral mucosal papules, most prominent on the gingiva. Trichilemmomas are benign follicular tumors usually located on the face. Sclerotic fibromas are firm nodules, that are usually solitary and benign. If 2 or more of these lesions are present, the incidence of Cowden's is nearly 100%. GI polyps are present in approximately 30% of patients, and can present at any site in the GI tract. Epidermoid cysts are seen in patients with Gardner's syndrome.