Cutaneous manifestations of gastrointestinal disease

Part I

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Learning Objectives
After completing this learning activity, participants should be able to identify cutaneous signs of polyposis and nonpolyposis disorders and their molecular/genetic associations to initiate prompt work-up, surveillance, and prevention of colorectal carcinoma.

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Cutaneous findings are not uncommonly a concomitant finding in patients afflicted with gastrointestinal (GI) diseases. The dermatologic manifestations may precede clinically evident GI disease. Part I of this 2-part CME review focuses on dermatologic findings as they relate to hereditary and nonhereditary polyposis disorders and paraneoplastic disorders. A number of hereditary GI disorders have an increased risk of colorectal carcinomas. These disorders include familial adenomatous polyposis, Peutz–Jeghers syndrome, and juvenile polyposis syndrome. Each disease has its own cutaneous signature that aids dermatologists in the early diagnosis and detection of hereditary GI malignancy. These disease processes are associated with particular gene mutations that can be used in screening and to guide additional genetic counseling. In addition, there is a group of hamartomatous syndromes, some of which are associated with phosphatase and tensin homolog (PTEN) gene mutations, which present with concurrent skin findings. These include Cowden syndrome, Bannayan-Riley-Ruvalcaba syndrome, and Cronkhite–Canada syndrome. Finally, paraneoplastic disorders are another subcategory of GI diseases associated with cutaneous manifestations, including malignant acanthosis nigricans, Leser–Trélat sign, tylosis, Plummer–Vinson syndrome, necrolytic migratory erythema, perianal extramammary Paget disease, carcinoid syndrome, paraneoplastic dermatomyositis, and paraneoplastic pemphigus. Each of these disease processes have been shown to be associated with an increased risk of GI malignancy. This underscores the important role of dermatologists in the diagnosis, detection, monitoring, and treatment of these disorders while consulting and interacting with their GI colleagues. (J Am Acad Dermatol 2013;68:189.e1-21.)

Key words: Bannayan-Riley-Ruvalcaba syndrome; Cowden syndrome; Cronkhite–Canada syndrome; cutaneous manifestations; gastrointestinal disorders; hamartomatous polyposis syndrome; hereditary and nonhereditary gastrointestinal malignancies; Lynch syndrome; Muir–Torre syndrome; paraneoplastic syndrome; Peutz–Jeghers syndrome.

Alimentary tract disease may present with cutaneous symptoms, either primarily or secondarily caused by an underlying systemic disorder. Because the gastrointestinal (GI) and cutaneous systems have closely linked developmental origins, concurrent pathologic disease frequently presents in unison. This review focuses on cutaneous manifestations pertaining to GI disorders. Part I focuses on the cutaneous manifestations of hereditary GI malignancy and paraneoplastic syndromes. Part II focuses on GI inflammatory bowel disease and vascular and connective tissue disorders as they relate to GI disease.

CAPSULE SUMMARY

- Hereditary gastrointestinal diseases and paraneoplastic syndromes manifest distinct cutaneous features.
- Hereditary malignancies include nonpolyposis and polyposis colorectal cancer, hamartomatous polyposis, and Cronkhite–Canada syndrome.
- Paraneoplastic syndromes do not have direct malignant infiltration, but primary tumor and cutaneous progression is typically parallel.
- Identification of these cutaneous findings is necessary for timely diagnosis, surveillance, and treatment of the underlying gastrointestinal pathology.

CUTANEOUS MANIFESTATIONS OF HEREDITARY GASTROINTESTINAL CANCERS

See Table I. Colorectal cancer (CRC) affects 130,000 people annually in the United States. Almost one-third of CRCs are familial, some of which have a genetic basis. Lynch syndrome (LS) or hereditary nonpolyposis colon cancer (HNPCC), familial adenomatous polyposis (FAP), Peutz–Jeghers syndrome (PJS), and juvenile polyposis syndrome (JPS) are the 4 main familial CRC syndromes.[1]

The early diagnosis of these syndromes is imperative so that appropriate surveillance and prevention can be initiated. Familial

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