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Synchronous Primary Adenocarcinoma and Ancient Schwannoma in the Colon: An Unusual Case Report

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Key Words

Schwannoma · Ancient schwannoma · Gastrointestinal tract · Adenocarcinoma

Abstract

Gastrointestinal schwannomas are uncommon stromal tumors of the intestinal tract and colon schwannomas are extremely rare. We report a rare case of ascending colon schwannoma with associated synchronous adenocarcinoma of the sigmoid colon. A 68-year-old man presented with a 20-day history of bleeding per rectum. Colonoscopy revealed a mass of 4.2 cm in diameter with endoluminal protrusion in the sigmoid colon and a second submucosal tumor in the ascending colon. Surgical intervention was suggested and ileo-hemicolectomy was done. Microscopically, the submucosal tumor of 4 cm in diameter showed features of schwannoma with degenerative change (ancient schwannoma). Lesional cells were positive for S100p and negative for actin, desmin, CD34, CD117, and pankeratin. The mass showed features of an invasive moderately differentiated adenocarcinoma. Colon schwannoma is a rare submucosal tumor, and the incidental occurrence with adenocarcinoma has not been well described in the literature.

Introduction

Synchronous occurrence of epithelial and gastrointestinal nerve tumors in the large intestine is extremely rare [1-3]. This association has been detected incidentally at surgery or colonoscopy for other reasons. It is not known whether or not such an association is a simple incidental coexistence or whether the two lesions are connected by a causal relationship [2, 3].

We report a case of ascending colon schwannoma with associated synchronous adenocarcinoma of the sigmoid colon.

Case Report

A 68-year-old man came to the emergency department of our hospital with a 20-day history of bleeding per rectum. His past medical history included essential hypertension and hyperlipidemia. There was no family history of intestinal diseases. Physical examination of the lungs, heart, and other body systems revealed no abnormal features. Laboratory investigation including complete blood count, biochemical examination, and tumor markers (CEA, Ca19-9, and AFP) were normal except for elevated blood glucose level (142.0 mg/dl).

Computerized tomography (CT) of the pelvis showed an obvious high-density enhancing lesion in the wall of the sigmoid colon. Colonoscopy revealed a mass of 4.2 cm in diameter with endoluminal protrusion in the sigmoid colon. A second submucosal tumor was found in the ascending colon, and the biopsy failed to prove its nature. Biopsy from the mass showed an invasive moderately differentiated adenocarcinoma. Surgical intervention was suggested and ileo-hemicolectomy with regional lymph node resection was done.

Microscopically, the submucosal tumor of 4 cm in diameter was a well-circumscribed intramural tumor and showed features of schwannoma with degenerative change (ancient schwannoma). The Schwann cell nuclei were large and hyperchromatic but lacked mitotic figures. The neoplasm ulcerated the intestinal mucosa and invaded the entire thickness of the muscularis propria. Lymphoid cuffing over vessels and lymphoid infiltration in the tumor were also noted (fig. 1, fig. 2).

The immunohistochemical study showed that the lesional cells were positive for S100p and negative for actin, desmin, CD34, CD117, and pankeratin (fig. 3). Histopathology of the sigmoid colon mass showed a moderately differentiated adenocarcinoma which infiltrated the entire thickness of the large bowel wall (fig. 4). Twenty regional lymph nodes were resected and showed reactive hyperplasia.

Two years later, the patient showed no evidence of recurrence or metastasis.

Discussion

Gastrointestinal schwannomas are rare, and schwannomas occurring in the large intestine are extremely rare. The incidence of submucosal schwannoma has been reported to range from 0.4 to 1% of all submucosal tumors of the gastrointestinal tract [4–6]. The commonest initial presentations are constipation and difficulty in defecation [4]. Colon schwannomas behave in a benign fashion and malignant change is rare [1, 4, 7]. Ancient schwannomas are usually large tumors of long duration, and degenerative changes include cyst formation, calcification, hyalinization, and hemorrhage. The Schwann cell nuclei are large, hyperchromatic, and often multilobed but lack mitotic figures. These tumors behave as ordinary schwannomas [8]. It is common for benign schwannomas to invade several layers of the bowel wall and even to extend into the surrounding adipose tissue [4, 9].

Although rare, schwannomas must be included in the differential diagnosis from other intestinal mesenchymal neoplasms, such as smooth muscle tumors, neurofibromas, and GISTs.

The immunophenotype differs from that of other mesenchymal neoplasms because the spindle cells of schwannoma show strong diffuse positivity for S100p and vimentin

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and negative staining for CD117, CD34, desmin, and actin [1, 4, 7, 10]. Accurate diagnosis and recognition of this benign entity is often difficult before surgical intervention [1, 4]. Treatment of choice is radical surgery [4].

Synchronous occurrence of adenocarcinoma and gastrointestinal nerve tumor in the large intestine has not been well described in the literature. Further studies are required to clarify the molecular alterations in such cases and reveal the etiology of this association [1, 2].



Fig. 1. Ancient schwannoma. HE, ×40.



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Fig. 2. Ancient schwannoma. HE, ×200.



Fig. 3. Schwannoma, S100p. ×200.

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Fig. 4. Adenocarcinoma of the large intestine. HE, ×40.

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