

# Colon Schwannoma : A Case Report

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## Abstract

Schwannoma is a common soft tissue tumor, arises from Schwann cell, which cover the peripheral nerves. Gastrointestinal schwannomas are rare and colon schwannomas are extremely rare. We report a case of ascending colon schwannoma with the initial presentation of chronic diarrhea. This 65 year-old men had chronic water diarrhea for six months. Colonoscopy examination showed a submucosal tumor with ulceration and it protruded into ascending colon. The initial biopsy pathology showed necrotizing inflammation. Another two colonoscopic biopsy also failed to prove the nature of the submucosal tumor. Finally, he received right hemicolectomy with the tumor completely resected. The pathological diagnosis of this tumor was benign gastrointestinal schwannoma. ( J Intern Med Taiwan 2009; 20: 255-259 )

**Key Words :** Schwannoma, Submucosal tumor, Neurilemmoma

## Introduction

Schwannoma, also termed neurilemmoma, arises from Schwann cells, which cover the peripheral nerves. It commonly occurs in the peripheral nerve of the limbs or body, spinal cord, and the central nervous system. Occasionally, Schwannomas can occur elsewhere, including subcutaneous tissue, the retroperitoneum, mediastinum, vulva, breast, larynx, and foot. There has been a relatively small number of gastrointestinal schwannomas reported<sup>1</sup>. The incidence of submucosal schwannoma has been reported to

range from 0.4% to 1% of all submucosal tumor of gastrointestinal tract<sup>2,3</sup>. In particular, schwannoma occurring in the large intestine is extremely rare. The incidence of colon schwannoma is equal among men and women. Worldwide distribution had not been reported. Rare genetic factors had been identified except for those with Neurofibromatosis<sup>4</sup>. Intracranial schwannoma had been reported in Taiwan<sup>5</sup>, but to our knowledge, few gastrointestinal schwannoma has been reported.<sup>2</sup> We herein report a case of colon schwannoma and review the previous reports of benign schwannoma of the colon.

## Case report

The 65-year-old man visited our gastro-intestinal (GI) clinic with the chief complaint of chronic watery diarrhea for six months. The character of bowel habit were yellowish watery diarrhea, 7~8 times per day, and associated with dark or bloody stool intermittently. Postprandial abdominal fullness, abdominal cramping before diarrhea, intermittent nausea and body weight loss 10 Kg in one month were also noticed. He had no systemic disease except for duodenal ulcer with bulb deformity. Physical examination and digital examination were unremarkable. Laboratory test revealed hemoglobin 13.2 gm/dl, albumin 3.54 gm/dl and the stool occult blood was positive. Colonoscopy was arranged and a large submucosal tumor was found in his ascending colon (Figure 1). Then, he was admitted for further workup. The initial colonoscopic biopsy showed necrotizing inflammation and he received two more times colonoscopy for biopsy. However, we still failed to prove the nature of the tumor (not showed). Lower GI series revealed an eccentric polypoid mass at the posterior aspect of the distal ascending colon (Figure 2). Multi-detector computed tomography (CT) of the abdomen was performed. Pre-enhancement image showed an isodense polypoid lesion over ascending colon (Figure 3a). After contrast enhancement, it revealed a polypoid lesion over ascending colon with endoluminal protrusion. Homogeneous enhancement with central necrosis and peripheral thin layer enhancement of mucosa were also noted (Figure 3b,3c,3d). Submucosal tumor was diagnosed and the differential diagnosis included gastrointestinal stromal tumor(GIST), lymphoma, leiomyoma, leiomyosarcoma, neurogenic tumor. Neither obvious infiltration to surrounding organ, hepatic metastatic lesions, nor intra-abdominal lymphadenopathy were found. His serum carcinoembryonic antigen (CEA) was

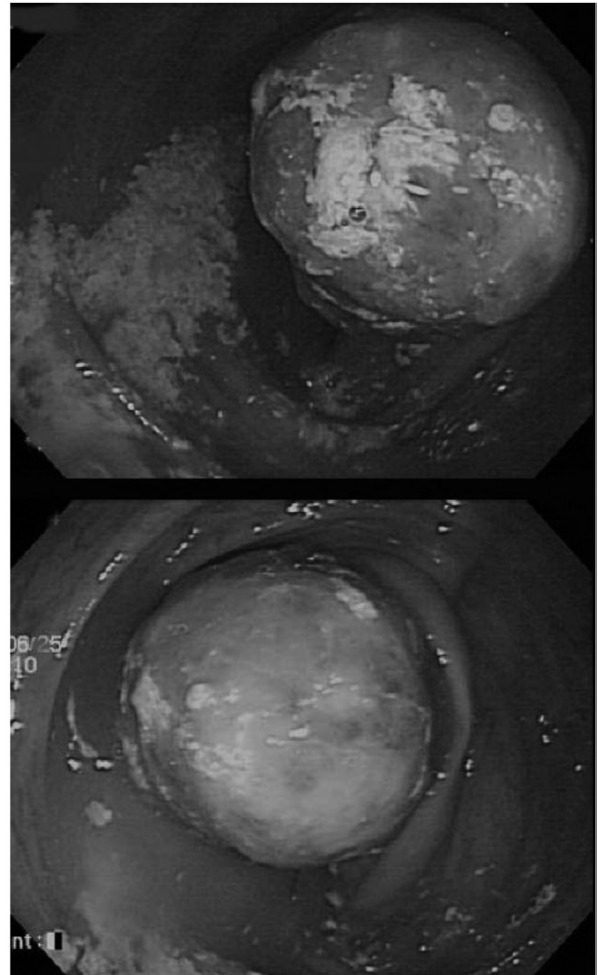


Fig.1. Colonoscopy showed an intraluminal tumor covered with necrotic tissue in the ascending colon. The size is approximately 3.5 cm in diameter.

4.65 ng/ml. Surgical intervention was suggested and right hemicolectomy with regional lymph node resection was done. Grossly, the tumor was polypoid and measures 3.1 x 2.8 cm in diameter. On cross section, the tumor was grayish to white in color. Microscopically, haematoxylin & eosin-stained sections of ascending colon tumored show a well-circumscribed tumor composed of blank looking spindle cells with palisading nuclei. Lymphoid cuffing over vessel and lymphoid infiltration in the tumor were also noted. (Figure 4A). Immunohistochemical studies was positive for S-100 protein (Figure 4D), negative to smooth



Fig.2. Barium enema revealed eccentric polypoid mass like lesion at posterior aspect of the distal ascending colon.

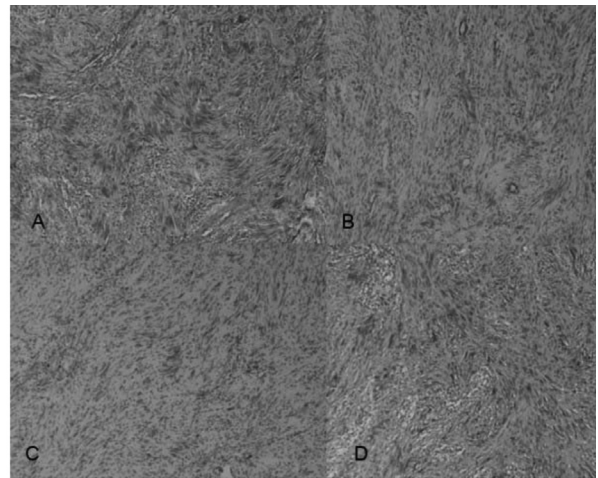


Fig.4 Tumor cell showing the palisading of tumor cell nuclei under H&E stain (A). Immunohistochemical stain demonstrating diffusely positive cytoplasmic reactivity to S-100 protein(D), but weak to smooth muscle actin(B) and CD117(C)

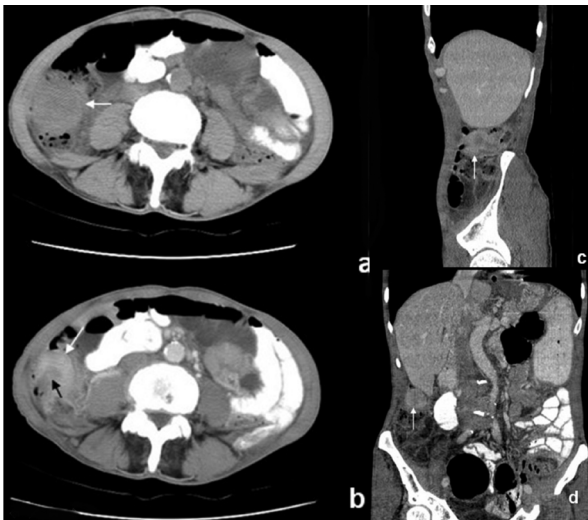


Fig.3. Abdominal CT showed a polypoid lesion over ascending colon(3a), Enhancement CT image showed homogenous enhanced polypoid lesion over ascending colon with central necrosis(3b, black arrow) and peripheral thin layer enhancement of mucosa(3b, white arrow). No obvious infiltration of the tumor to surrounding organ and no obvious near lymphadenopathy noted.

muscle actin and CD117 (Figure 4B & 4C). Therefore, schwannoma was diagnosed. Twenty-

seven regional lymph nodes were resected and the pathology showed reactive hyperplasia. The postoperative course was smooth and we followed up this patient for one year. He was symptom-free and there was no evidence of recurrence or metastasis.

### Discussion

Clinical symptoms of colon tumor depend on its location and speed of growth. The most common site of colon schwannoma reported was in the recto-sigmoid colon and the most common symptoms were constipation and difficulty in defecation<sup>6</sup>. Other symptoms included bloody stool, abdominal pain or discomfort, stool occult blood and anal pain. All of above symptoms were not specific to colon schwannoma. In general, colon schwannomas show a benign behavior. There is no mutual agreement on the histological criteriae for defining the grade of GI schwannoma and its metastatic potential is still unknown. Malignant transformation had been reported in peripheral schwannoma<sup>7</sup>. Careful observation of the patient's post-operation course is mandatory.

Histologically, schwannoma are composed of spindle-shaped cell with intertwining cyto-plasmic extensions seen on both light and electron microscopy. The typical histological features include a nuclear palisading and prominent lymphoid cuff with or without germinal centre formation. Routine hematoxylin and eosin staining does not differentiate neurogenic from myogenic tumors. Immunocytochemical studies are required to distinguish schwannoma from neurofibromas, GISTs, and leiomyomas. Cell from schwannoma are usually 100% immunoreactive with S-100 protein in contrast to 30% to 40% immunoreactivity in neurofibromas. GISTs are positive for c-kit and negative for S-100 protein, whereas leiomyomas, which are derived from a smooth muscle precursor, are positive for smooth muscle actin and desmin, and negative for S-100<sup>8</sup>.

Inagawa et al. reviewed 46 cases with large intestine schwannoma in Japan<sup>4</sup>. Immunohistochemical stainings for S-100 protein were positive in all of them. He summarized that schwannoma appears grossly as a spherical, solid, gray and well-encapsulated tumor. The tumors are typically hard and it is difficult to obtain proper biopsy samples from them. Surface ulceration is often seen. Thus, a definitive diagnosis is preoperatively made in only 7 (15%) of the cases. The mean age was 60.4 years and the average tumor size was  $4.6 \pm 2.5$ cm. Most of the tumor located in the rectum (45.7%) and the commonest initial presentations were constipation and difficulty in defecation (28.6%).

A correct diagnosis of schwannoma is often difficult before surgical intervention, because of the limited information available on such tumors. Therefore, large-forceps biopsy or aggressive surgery has often been performed<sup>6</sup>. Few case reports mentioned about endoscopic ultrasonography finding (EUS) of colon schwannoma<sup>9-11</sup>. EUS is helpful in defining the borders of the tumor and its localization within the GI wall. But there was no

conclusion about the typical EUS features of GI schwannoma because of their plexiform pattern of growth. Besides, it is common for benign schwannomas to invade several layers of bowel wall and even extend into the surrounding adipose tissue<sup>12</sup>. EUS-guided fine needle aspiration or biopsy improves the diagnosis of GI submucosal tumor. However, definitive treatment requires a complete surgical resection. Endoscopic treatment of submucosal lesions had been reported but it was limited to pedunculated and smaller (<2cm in diameter) tumors<sup>13,14</sup>. Incomplete resection could be associated with local recurrence.

In conclusion, colon schwannoma is rare colon submucosal tumor with benign behavior. Definite histological diagnosis is difficult before surgical intervention. Image study with EUS, abdominal CT and EUS-guided aspiration can increase the diagnosis rate before operation.

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## 大腸神經鞘瘤：病例報告

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### 摘 要

神經鞘瘤 (Schwannoma) 為源於包覆在神經外許旺氏細胞 (Schwann cell) 的腫瘤。腸胃道的神經鞘瘤甚為罕見，位於大腸的神經鞘瘤更為罕見。在此我們提出一個位於升結腸的大腸神經鞘瘤病例報告。病人為65歲中年男性，初期臨床表現為慢性腹瀉六個月。大腸鏡檢查發現升結腸處有一黏膜下腫瘤。腫瘤突入升結腸內，表面呈現慢性潰瘍。大腸鏡切片檢查呈現壞死性發炎 (necrotizing inflammation)。另行兩次大腸鏡檢查及切片亦無法適切的取得此黏膜下腫瘤的病理切片。最後病人接受右結腸切除術。術後病理切片證實為良性大腸神經鞘瘤。