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Case Report

Schwannoma of the colon: a case report and literature review

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ABSTRACT

Tumours that originate from the Schwann cells are known as schwannomas. Schwannomas are generally rare, but the rarest are perhaps those of the colon and rectum. The areas of the body that ordinarily experience more schwannomas include such organs as the head, neck, arms, and limbs. Although they are mostly usually harmless, malignant degeneration may appear without surgery. The current case study is a male, 40 years of age, who reported abdominal pain and recurrent vomiting and was admitted for elective Esophagogastroduodenoscopy (EGD) and colonoscopy. There was no family history of inflammatory bowel disease or cancer, and he had no prior abdominal surgeries. A physical examination revealed mild tenderness in the right lower quadrant. Results of the examinations indicate normal mucosa and vasculature in visualized parts. One polyp ascending around 1 cm was removed completely by cold snare after elevation by an adrenaline base secured by one clip. Another polyp in the transverse colon, around 1 cm, was removed completely by a hot snare base secured by one clip. Following a routine biopsy test that revealed a polypoid lesion consisting of bland spindle cells with an elongated shape. Upon histological testing, a schwannoma was found. Immunohistochemistry revealed that the tumor was negative for EMA, cluster of differentiation (CD)117, calertinin, and synaptophysin, and positive for S100 and vimentin, with a Ki-67 proliferative index of 3%. There was no lymph nodes implicated. Schwannomas may present difficult biopsy investigations, and a reliable diagnosis of this sickness requires immunohistochemistry. Unlike gastrointestinal stromal tumors, Schwannomas contain vimentin and the S100 protein but are negative for CD117. The likelihood of tumor aggressiveness increases significantly in cases recording a Ki-67 score of ≥5%. As a result, the recommended treatment for schwannomas is oncological drastic surgical excision.

Keywords: Immunohistochemistry, Schwannoma, Colon, Tumor

INTRODUCTION

Schwannomas are a type of tumor that originates from Schwann cells. Although schwannomas can develop in almost any part of the body, particularly along the peripheral nerves, they hardly ever grow in the colon or rectum.[1] The tumors are more common in the extremities, the cephalic region, and the neck. As of 2020, only about 96 cases of primary colorectal schwannomas unrelated to von Recklinghausen's disease had been reported worldwide.[2] Of the 96 cases, almost 97% of them were established to be benign.[2] While discomfort, exhaustion, and fever are common non-specific symptoms of schwannomas, most of them are asymptomatic and non-malignant.[3] Signs of intestinal blockage and rectal bleeding can also happen on occasion.[4] Although most initial schwannomas are benign and asymptomatic, there is a chance of malignant degeneration, especially if the tumor is big. The preferred course of treatment is radical excision with disease-free margins because if left

untreated, the likelihood of recurring locally or becoming malignant increases significantly.[5,6] Different surgical approaches can used in treating the tumors depending on various factors, such as the size and location of the tumor and its histopathological pattern.[7] The suitability of radiation and chemotherapy is still unknown, so those treatments are not recommended.

CASE REPORT

The current case involves a 40-year-old medically free male. He reported a history of abdominal pains and recurrent vomiting for 1 month and was subsequently admitted to the admitted to Gastroenterology department for an elective Esophagogastroduodenoscopy (EGD) and colonoscopy. The patient reported that he had never undergone any abdominal surgeries. Additionally, he reported that he did not know any family member who has previously had inflammatory bowel disease or tumors. However, upon physical examination, results showed that right lower quadrant had slight tenderness. A C-reactive protein level of 45.90 mg/l was observed in the laboratory, other tests yielded no noteworthy results.

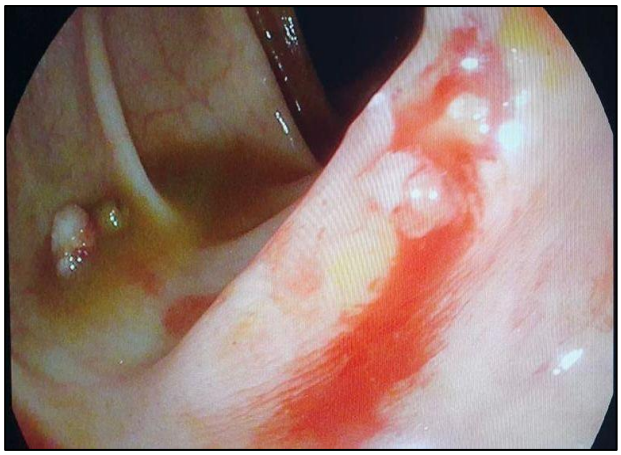

Figure 1: A sessile polyp observed on the ascending colon during colonoscopy.

Upper gastrointestinal endoscopy for the esophagus part showed GERD grade B with a small tongue-like projection biopsied for histopathology. Other parts were unremarkable. On colonoscopy One polyp in ascending around 1 cm removed completely by cold snare after elevation by adrenaline base secured by one clip. Another 1 cm polyp was removed from the transverse colon by a hot snare base secured by one clip.

Histological examination of the esophageal biopsy displayed gastroesophageal junction mucosa exhibiting chronic inflammation and features indicative of reflux disease without evidence of intestinal metaplasia or dysplasia. Upon biopsy, a polyp from the ascending colon was identified as a tubular adenoma. Notably, a polyp biopsy from the transverse colon demonstrated a polypoid lesion composed of bland spindle cells with elongated, pointed nuclei. No distinct Verocay bodies or prominent nuclear palisading was observed. The stroma contained prominent collagen bands. The tumor tested positive for S-100 by immunohistochemical examination, but negative for calretinin, synaptophysin, and epithelial membrane antigen (EMA). The presence of colon schwannoma was confirmed.

DISCUSSION

Rare ectodermic cancers called schwannomas proliferate from the neural sheath.[4] They originate from the autonomous neural system in the digestive tract. In most cases, the tumors are harmless, slow-developing, and are rarely seen or detected.[8] In some cases, however, they experience malignant degeneration, and, according to Sreevathsa and Pipara, gastric schwannomas cause 0.063, 0.04, and 0.002 of all gastric mesenchymal tumors, benign gastric tumors, and gastric tumors, respectively.[3] The incidence rate of schwannomas increases between the ages of 60 and 70, affecting both genders equally, 9 with the stomach (0.83) and small bowel (0.12) being the most common.[1] Under a microscope, lobulated schwannomas with clear boundaries regularly exhibit a cyst-like structure.

Auerbach's plexus-related tumors form oval-shaped, non-pedunculated masses that extend into the intestinal lumen, while those from Meissner's plexus may have a mushroom-like shape.[10] Verocay bodies are the histological identifiers of schwannomas. Other histological identifiers include a surrounding lymphoid layer, a coil of densely packed, spindle-shaped cells.[11] Antoni A and Antoni B are the two recognized histological growth patterns, the first of which contains Verocay bodies of dense, spindle-shaped cells.[12] In contrast, although Antoni B also has spindle-shaped cells, they are loosely organized and they contain a plentiful myxoid matrix, and the presence of xanthomatous histiocytes.[12]

Given that malignant schwannomas are hardly ever seen, the number of cases reported in the literature is extremely low, with only 96 cases of primary colorectal schwannomas reported as of 2020.[2] The neck, head, arms, and limbs are the areas in which schwannomas are most commonly observed.[13] Among gastric schwannomas, the stomach is the most common site for gastric schwannomas.[1] The extreme rarity of schwannomas also means their unique biological functioning is not sufficiently understood.

Moreover, they are mostly asymptomatic, which makes it even harder to detect, study, and understand them.[14] When symptomatic, the symptoms tend to be nonspecific, including abdominal discomfort, a palpable mass, gastrointestinal bleeding, colonic obstruction, and defecation disorders.[15] Despite the mostly benign nature of schwannoma of the colon, local recurrence and distant metastasis still pose a small risk.[16] Nevertheless, dissection of the lymph nodes is not usually necessary because gastric

schwannomas rarely metastasize to the lymph nodes.[17] Therefore, although the lymph nodes are likely to enlarge, the main cause of that enlargement is inflammatory hyperplasia.[17]

The Ki-67 proliferation index (MIB-1) is an essential measure for assessing the risk posed by the tumour because a positive score (≥5%) is strongly associated with advanced malignant schwannoma.[18] Such advanced malignant schwannoma of the colon can metastasize to the liver but not to the lymph nodes.[19] The risk of metastasis increases significantly when the tumour is larger than 5cm and has a high magnification mitotic activity rate of more than five mitoses per field. In the current case, the diagnosis is based on histopathology. A biopsy of the first duodenal segment revealed a hyperplastic polyp with no evidence of cancer or dysplasia. The oesophagal biopsy showed features of reflux disease with no signs of dysplasia or cancer. A polyp biopsy from the ascending colon identified it as a tubular adenoma without dysplasia. A biopsy of the polyp from the transverse colon confirmed it to be a schwannoma.

Pre-operative biopsy exams can be challenging, and an accurate diagnosis of schwannoma is required. Immunohistochemistry for schwannomas, total surgical excision with oncological radical intent, is currently the gold standard of care.[20] Based on our experience, major surgery is the recommended course of action, even when the lymph nodes are uninvolved.

The preferred treatment is complete surgical excision with clear margins free of neoplastic involvement. Broad resections of lymph nodes are not advised because there is little chance of malignant transformation.[21] The same surgical criteria should be applied to tumors that exhibit more aggressive behaviour, although more stringent postoperative follow-up is advised. The tumour's location, size, and histological pattern all influence the surgical strategy.

A consideration for minimally invasive surgery using video laparoscopic access should be made for cases involving the proximal colon and rectum. Transanal endoscopic excision is a possible treatment for cancers in the distal rectum.[21,22] Adjuvant chemotherapy and radiation still yield inconsistent results. Thus, routine use of these treatments is not advised. Colon and rectal schwannomas are uncommon tumours that require the use of an immunohistochemistry panel to obtain a conclusive histopathological diagnosis. Radical surgical removal is the best course of action because it increases the chances of disease-free and overall survival.

CONCLUSION

In certain situations, colonic schwannoma may show in a variety of ways that resemble malignant tumours. If left untreated, it might result in serious side effects such as perforation and peritonitis that could jeopardize the critical prognosis. Total surgical resection is the standard of care. Even when partially removed out of necessity, the slow evolutionary genius of schwannoma confirms its favourable prognosis.

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