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Case report

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Sigmoid Colon Schwannoma: A Case Report & Review of Literature

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Abstract

Colorectal schwannoma is a rare neoplastic growth originating from the myenteric plexus of the GI tract, which was described for the first time in 1962. It is an extremely rare presentation, with around 95 reported cases with only 28 reported cases in the sigmoid colon.

This paper presents a case report of a 55-year-old male patient with complaints of constipation, per rectal bleeding, and on-and-off abdominal pain. He was worked up coloscopically and radiologically and found to have a sigmoid colon mass with ulceration and lumen narrowing leading to colonic intussusception. Preoperative biopsy was inconclusive. The patient underwent open sigmoid resection after proper preoperative preparation, with his postoperative histopathology reported as sigmoid colon schwannoma.

Background/Introduction

Verocay, in 1910, was the first to describe a benign rare neoplasm of ectodermic origin growing from the neural sheath and characterized by the presence of Verocay corpuscles (Baig et al., 2019). This growth, later known as schwannoma, is a tumor originating from the Schwann cells of the nerve sheath of the peripheral nervous system.

The presence of schwannoma in the gastrointestinal tract -from the myenteric plexus- is not a common occurrence. It is commonly found in the stomach (80%), followed by the small intestine (10-15%), while colorectal schwannoma is extremely rare (Uhr et al., 2016).

Due to its risk of malignant transformation, resection of schwannoma is the recommended treatment (Qi et al., 2021).

Case Report

A 55-year-old male known to have diabetes, hypertension, and end-stage renal disease on hemodialysis was referred to our hospital complaining of constipation secondary to colon mass. A physical exam showed a middle-aged, vitally stable man with a soft and lax abdomen. The patient's per rectal exam was unremarkable for masses or blood. Colonoscopy was done for him, and it showed a fungating ulcerating mass 40 cm from the anal verge, and the scope could not pass. Histopathologic examination of the colonic biopsy was non-diagnostic, with only fibrin and ulcer debris present in the sample. CT chest, abdomen, and pelvis with triple contrast showed the findings of mid-sigmoid colon mass with significant lumen narrowing with no high-grade bowel obstruction, with few mildly enlarged suspicious regional mesocolic lymph nodes suspicious for malignancy. FDG PET/CT scan was also done FDG avid rounded mass at the upper sigmoid colon in keeping with known colon cancer-causing colo-colic intussusception with no evidence of obstruction and no PET evidence of nodal or distant metastasis. The patient, after this, was taken to the theater

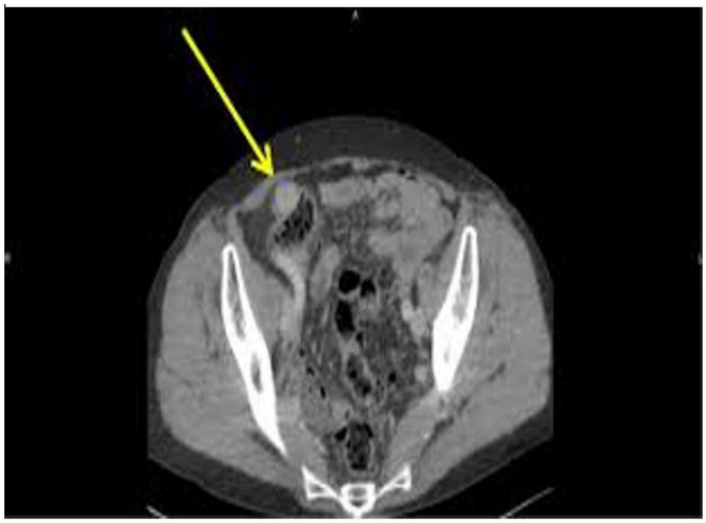

Figure 1: *CT scan showing presence of colon schwannoma.*

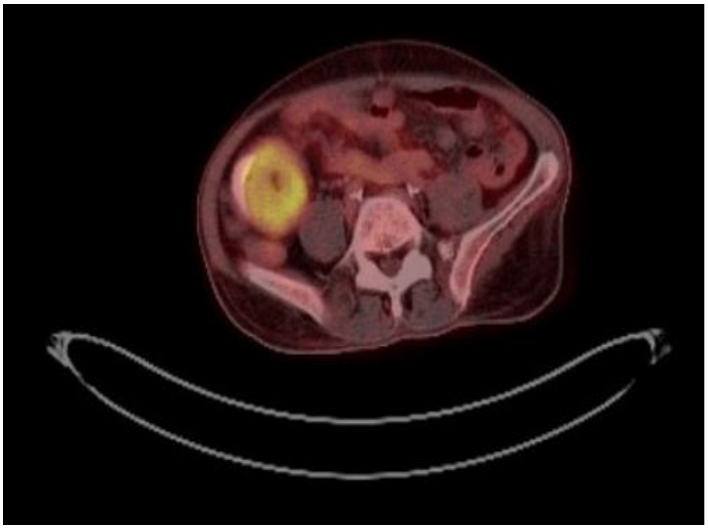

Figure 2: *FDG PET/CT scan showing presence of that same mass as FDG avid lesion*

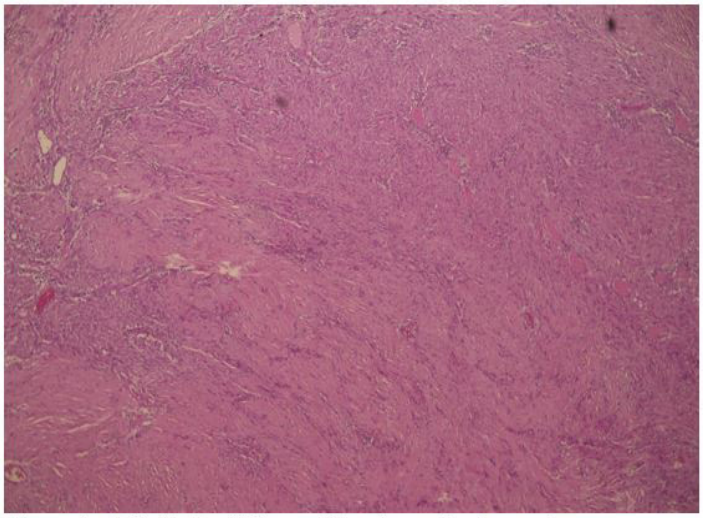

Figure A: *Spindled cellular proliferation with alternating hypocellular and hypercellular zones (40x magnification, H&E).*

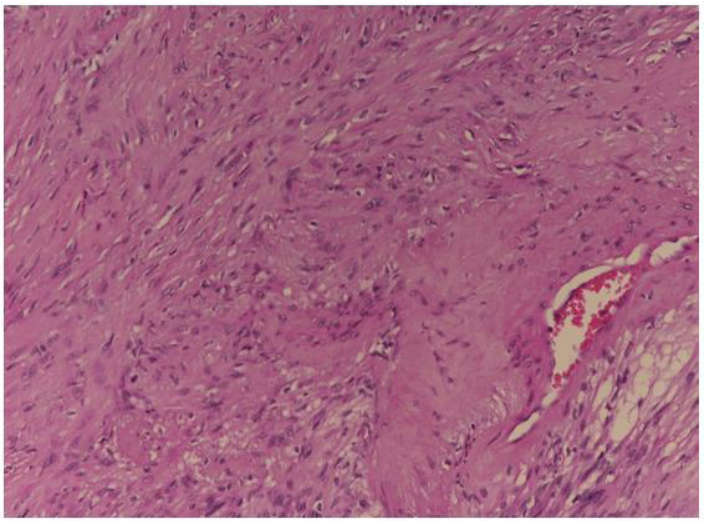

Figure B: *Thickened hyalinized blood vessel is appreciated (right side) (200x).*

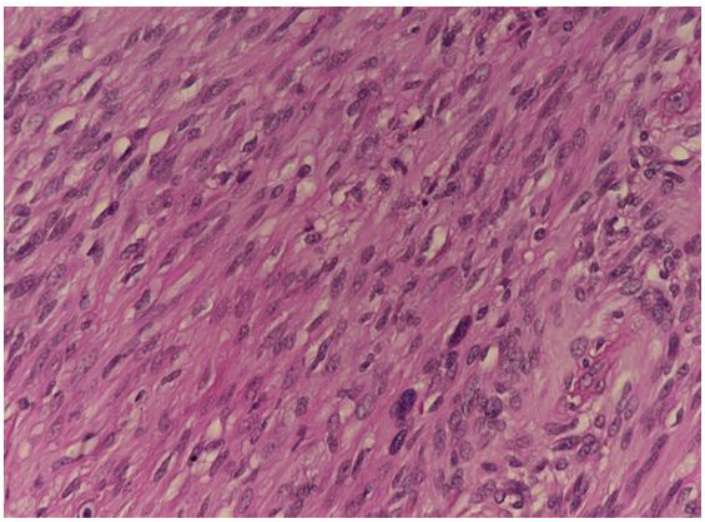

Figure C: *The neoplastic cells show eosinophilic cytoplasm and contain elongated wavy nuclei with fine open chromatin and indistinct nucleoli (400x).*

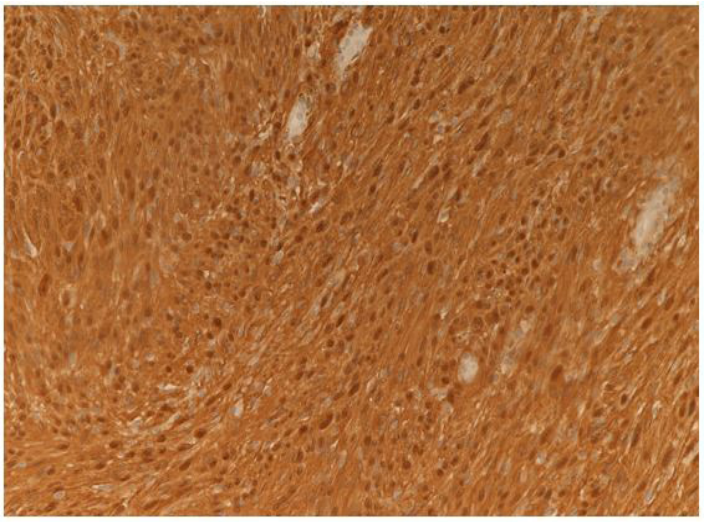

Figure D: *The tumor cells are diffusely positive for S100 immunohistochemical stain (200x).*

Figure 3: *A to D shows the microscopic features of the tumor.*

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for anterior resection anterior resection with end colostomy and mucus fistula as primary anastomosis was not feasible due to poor nutritional status and co-morbidities. Post-operatively, the patient developed pseudomembranous colitis, for which he started oral and rectal vancomycin. He stayed stable throughout the postoperative recovery period and was discharged home in good condition. The sigmoid resection specimen demonstrated a 5 x 4.5 x 3.0 cm mass associated with mucosal ulceration. Histologic sections of the colonic mass revealed spindle cellular proliferation with alternating hypocellular and hypercellular areas (Wang et al., 2016). There are occasional thickened and hyalinized blood vessels seen throughout the mass. The tumor cells show eosinophilic cytoplasm and contain elongated wavy nuclei with fine open chromatin and indistinct nucleoli. There is no evidence of increased mitotic activity, necrosis, or marked cytological atypia. Immunohistochemical stains show that the neoplastic cells are diffusely positive for S100 and negative for DOG-1, CD117, desmin, and smooth muscle actin. The overall morphologic and immunophenotypic findings support the diagnosis of colonic schwannoma (Hsu et al., 2007; Wang et al., 2010; Chayanupatkul et al., 2018; Ferreira Cardoso et al., 2019).

Discussion

According to Bohlok et al. (2018), 95 cases of colorectal schwannomas have been reported distributed from the most common location to the least; cecum and right colon (30.5%), sigmoid (28.1%), rectum (21.1%), left colon (8.3%) the transverse colon (5.3%) and the appendix (1.15) with slight female predominance (58%) and a median age of 61.2 years.

The presentation of colorectal schwannoma varies on its presentation. It can be incidentally diagnosed in an asymptomatic patient during screening colonoscopy or with a wide range of symptomatic presentations, including abdominal pain, change in bowel habit - constipation, diarrhea or change in stool diameter, per rectal bleeding, tenesmus, decrease in appetite or weight loss (Zippi et al., 2013; Pansari et al., 2020). It can also present with a mass effect complication, e.g., intestinal obstruction or intussusception. Two reported cases where schwannoma was diagnosed during workup with a synchronous colorectal adenocarcinoma (world journal of surgical oncology page 97 & Page 77)

Due to its wide range of presentation, the workup of colorectal schwannoma follows a seemingly similar route to the workup of any colorectal mass. On colonoscopy, schwannoma varies in its gross appearance as a polypoidal lesion or a submucosal mass with normal overlying mucosa, which might mandate the use of endoscopic ultrasound and deeper biopsy or FNA to reach an accurate diagnosis (Wilde et al., 2010). However, the mucosal covering might be ulcerating with bleeding/sloughing/lumen compromisation mimicking malignancy.

The initial biopsy specimens were non-diagnostic, most likely due to the superficial nature of submitted tissue material and the submucosal nature of this neoplasm, like in any other mesenchymal tumors of the gastrointestinal tract where a deeper and targeted sampling approach results in a higher yield diagnostic neoplastic tissue. Schwannomas are characterized by biphasic growth patterns of hypercellular areas (Antoni A) of spindled tumors containing elongated and wavy nuclei admixed with loose/myxoid regions (Antoni B). However, some of the classic findings of Schwannoma described in soft tissue and central nervous system counterparts may be absent in the gastrointestinal tract tumors. The presence of lymphoid aggregates at the periphery of the tumor is a helpful diagnostic clue but not identified in our case. The gastrointestinal stromal tumors (GIST) are the most common mesenchymal gastrointestinal tumor and occasionally can share morphological similarities with Schwannoma; thus, the immunohistochemical staining panel should include GIST markers (CD117 and DOG1 stains). The diagnosis can be confirmed by diffuse positivity for S100 and SOX10 and after the exclusion of other more frequent mesenchymal tumors (such as GIST and smooth muscle neoplasms).

Radiological studies such as enhanced CT or MRI to characterize the colorectal mass with the use of contrast to delineate the tumor and lumen patency better are recommended as part of the workup, especially when malignancy is suspected with the need to rule out metastasis (Fotiadis, 2005; Cak et al., 2015). Findings on CT scans were mentioned differently among the reported cases, including a mural-originating colorectal mass with low attenuation with either smooth margins or suspicious edges, compromised lumens, thickened wall, fat stranding, and the presence of enlarged adjacent lymph nodes. All these variations add more challenges to the diagnosis of this pathology.

Moreover, a preoperative FDG-PET CT scan has been done for 5 reported cases, including our patient. Despite that, all of them were labeled as benign. Four of these cases were FDG avid lesions making it more difficult to rule out malignancy without a pathological diagnosis.

Whether colorectal schwannoma was diagnosed pre- or post-operatively, the standard treatment is radical colorectal resection depending on the tumor location, without any clear recommendations for neoadjuvant or adjuvant treatment (Kojima et al., 2020). Despite this, there have been several cases that were diagnosed preoperatively and found to have benign schwannoma treated more

conservatively by endoscopic resection (page 12), cecal wedge resection (page 84), and trans-anal with no reported recurrence, yet small sample size is very limiting.

Postoperative histopathology is the final accurate way to diagnose schwannoma. Grossly it tends to be lobulated well-defined tumors, sometimes ulcerating into the mucosa with rich staining of S-100. Histologically, there are two recognized patterns, Antoni A, with a dense growth of fusiform cells compactly arranged in palisades to form Verocay bodies, and Antoni B, in which the fusiform cells are distributed more loosely with rounded and elongated nuclei, myxoid stroma and xanthomatous histiocytes (Nonose et al., 2009).

Miettinen et al. (2001), divided colorectal schwannoma into three clinicopathological types; spindle cell, epithelioid and plexiform. All three types are positive for S-100 protein and GFAP but lack CD117 (KIT), differentiating it from GIST tumors.

Despite these histological features and types, judging a colorectal schwannoma as benign or malignant depends pathologically on its mitosis rate, atypical mitotic figures, and nuclear hyperpigmentation, Ki-67 index with a value of more than 10% considered malignant. Clinically, long-term local recurrence or distant metastasis have also been used to judge the malignant state of a colorectal schwannoma, with only 3 cases reported in the literature.

Conclusion

Colorectal schwannoma is a rare disease with a wide range of presentations but mostly benign courses. Differentiating it from other mimickers is the biggest challenge, yet excision seems to be the main treatment approach.

Conflict of interest:

There was no conflict of interest in making this case report.

Declaration:

Ethics approval and consent to participate & Consent for publication:

Patient has been consented

Availability of data and materials: Available through citation, literature review, imaging and pathology pictures

Competing interests: No competing interests

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Author's contributions: Both authors participated in writing the manuscript and revision of data

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