Plexiform Fibromyxoma of the Colon: A Case Report

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Abstract

Introduction: Plexiform fibromyxoma is a rare mesenchymal tumor found in the gastrointestinal tract, most commonly in the gastric antrum. Limited data exists regarding a plexiform fibromyxoma arising primarily from the colon.

Case Report: We report the first case of a plexiform fibromyxoma arising from the colon in a 43 year-old male with an unremarkable past medical and surgical history. After presenting to his primary care provider with vague abdominal pain, the patient was found to have a right upper quadrant mass on imaging that was surgically resected with a laparoscopic-assisted right colectomy for presumed adenocarcinoma of the colon. However, pathology revealed that the mass was a benign plexiform fibromyxoma.

Discussion: The differential diagnosis of plexiform fibromyxoma in the colon includes more common colon masses like adenocarcinoma as well as spindle cell lesions including Gastrointestinal Stromal Tumor (GIST), schwannoma, plexiform neurofibroma, leiomyoma, leiomyosarcoma, and inflammatory myofibroblastic tumor. Immunohistochemical staining and genetic testing can help differentiate these etiologies and better characterize management and prognosis.

Conclusion: In a patient presenting with vague abdominal pain with imaging findings of a colonic mass, it is crucial to maintain a broad differential diagnosis. In this paper, we describe the first case of plexiform fibromyxoma arising primarily from the colon.

Keywords: Plexiform fibromyxoma; Mesenchymal tumor; Spindle cells; Abnormal abdominal imaging

Introduction

First described by Takahashi and colleagues in 2007 as plexiform angiomyxoid myofibroblastic tumor, plexiform fibromyxoma is a rare mesenchymal tumor found in the gastrointestinal tract [1,2]. Nearly all cases of plexiform fibromyxoma have been reported in the antrum of the stomach [3], with scattered rare cases reported in the duodenum [4], esophagus [5], jejunum [6] and ileocecal region [7]. We here report the first case of plexiform fibromyxoma arising in the colon. This work is compliant with the SCARE Guidelines [8].

Case Presentation

This 43-year-old male presented to Beth Israel Deaconess Medical Center (Boston, MA, USA) for evaluation of right-sided abdominal pain. He did not report any fevers, chills, night sweats, unintentional weight loss, changes in bowel habits, nausea, vomiting or rectal bleeding. His past medical history is notable for hypertension, gastroesophageal reflux disease and obstructive sleep apnea. He had no history of previous surgeries. He is a never user of alcohol or recreational drugs. He is a current smoker of less than 10 cigarettes per day. The patient has no known family history of Crohn's disease, colitis or colorectal cancer. The patient's general physical exam did not demonstrate any abnormalities. There was no palpable mass found in the abdomen. The patient had a previous abdominal ultrasound 7 years prior for RUQ abdominal pain that was normal.

Work up began with an ultrasound demonstrating a cystic mass with a thickened irregular wall seen in the right abdomen of unclear origin (Figure 1A). Follow up CT demonstrated a bi-lobed cystic tumor that measured 9 cm × 5.1 cm with a thick enhancing wall and localized...
lymphadenopathy (Figure 1B). Interestingly, at this point in his work-up the patient’s symptoms had resolved entirely and evaluation was continued because of the initial radiographic abnormality. The patient then underwent a colonoscopy which revealed a semi-circumferential non-bleeding 6 cm mass in the proximal ascending colon that was biopsied (Figure 1C). Pathology revealed focal low-grade dysplasia and colonic fragments with reactive changes, but no invasive carcinoma. The patient underwent an additional CT torso for staging of a presumed malignancy, which demonstrated no new thoracic findings but worsening lymphadenopathy near the mass.

Based on concern for malignancy given his imaging, the patient underwent a laparoscopic-assisted right colectomy. The gross pathology revealed a tumor that consisted of two conjoined masses coming from the wall of the colon. The first was an 8.6 cm firm, white intramural cystic mass that protruded through the mesentery. The second mass, a hemorrhagic cystic, measured 6.4 cm and similarly protruded through the mesentery (Figure 2A). We theorize this self-limited bleed into the wall of the colon is what caused the initial symptoms that lead to identifying the mass.

Histology demonstrated a plexiform, multinodular spindle cell lesion (Figure 2B) infiltrating the wall of the colon, with areas showing extension through and ulceration of the colonic mucosa. The nodules were composed of tumoral spindle cells within a fibromyxoid background that demonstrated a prominent vascular component. Cellularity was variable, with some regions appearing hypercellular (Figure 2C) and others appearing hypocellular (Figure 2D), in keeping with most reported examples of this neoplasm. The spindle cells themselves were bland and uniform, with rare enlarged nuclei and inconspicuous nucleoli. Mitotic activity and areas of necrosis were absent. The tumor stained diffusely positive for smooth muscle actin, with focal desmin positivity. Immunohistochemical stains for c-kit/CD117, DOG1, cytokeratin MNF116, and CD34 were all positive.
fibromyxomas have been shown to have a MALAT1-GLI1 translocation that results in overexpression of GLI1 protein, which is a component of the Sonic Hedgehog signaling pathway and has control of oncogenic genes [18]. While this patient’s tumor was negative for this translocation, there was nuclear amplification of uncertain significance of MALAT1.

Conclusion

Overall, the diagnosis of plexiform fibromyxoma in this patient’s colon was an incidental diagnosis found on imaging and confirmed with pathology in this patient who initially had a suspected colonic malignancy. This rare, benign mesenchymal neoplasm typically arises in the stomach; we report the first case arising in the colon, demonstrating that it should be considered in differential diagnoses in this location.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

References


