Case History:

A 66-year- old male presented for screening colonoscopy. The patient had a past medical history of acute myeloid leukemia in remission (status post allogeneic bone marrow transplant), prostate adenocarcinoma (managed with active surveillance), hypertension, and diabetes mellitus. Screening colonoscopy revealed ten polyps, ranging from 3-4 mm to 35 mm scattered along the mucosa from the cecum to the rectum. The largest polyp in the descending colon (35 mm) was pedunculated and clinically felt to be a lipomatous lesion. Polypectomy was performed.

Histologic sections of the descending colon polyp along with immunohistochemical (IHC) stains are depicted below (Fig, 1-6).

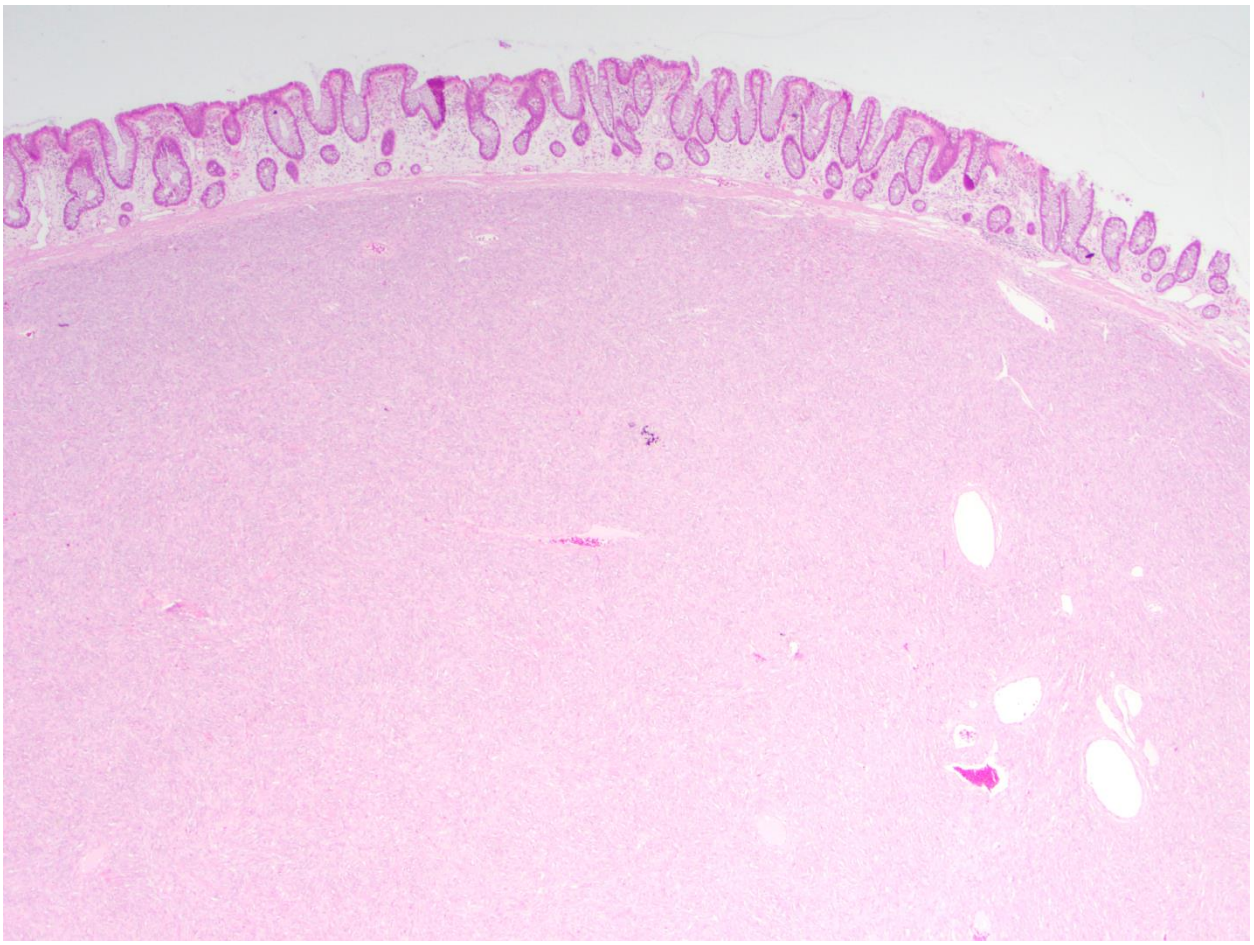

Figure 1. Hematoxylin and eosin (H&E), (20x).

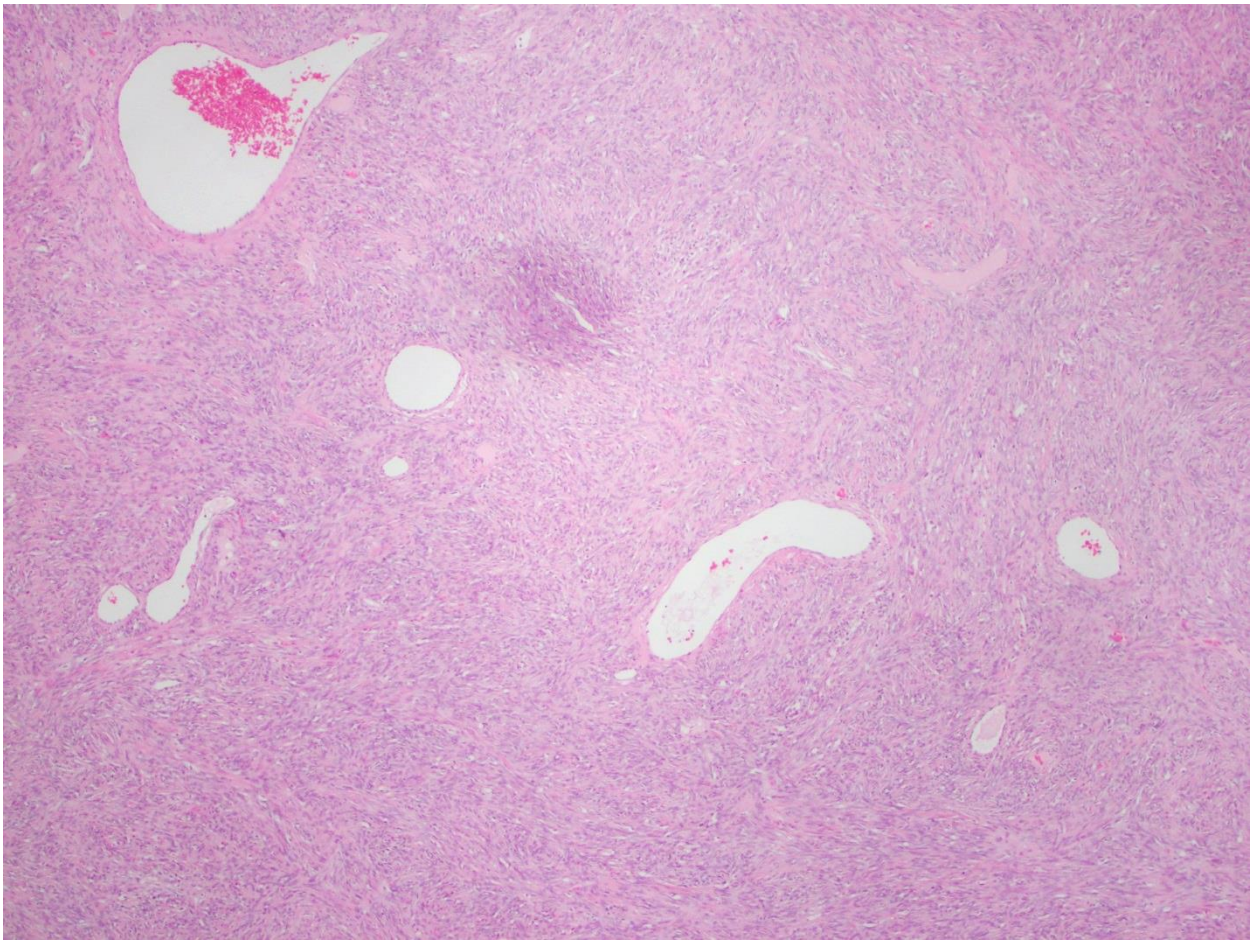

Figure 2. H&E, (40x).

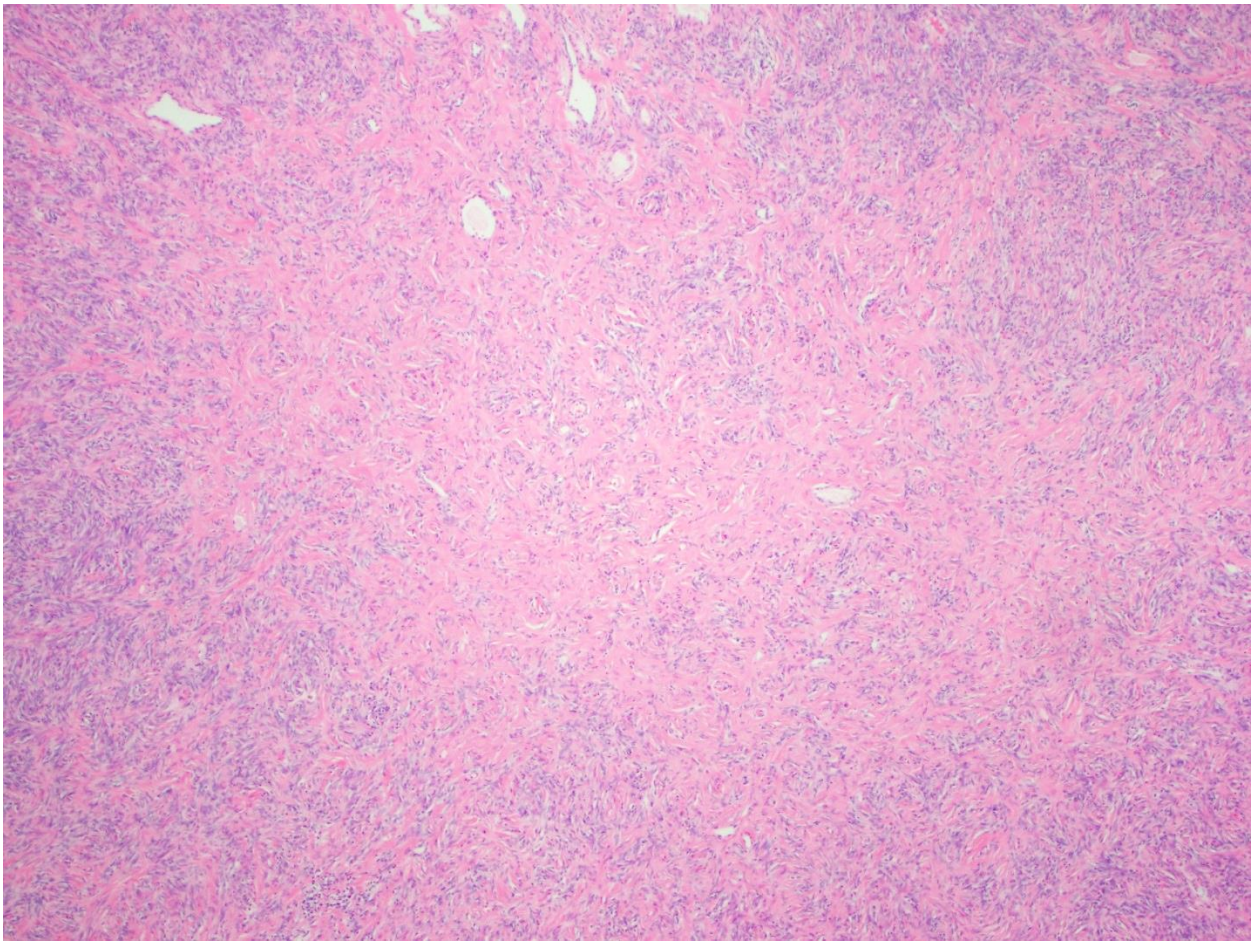

Figure 3. H&E, (40x).

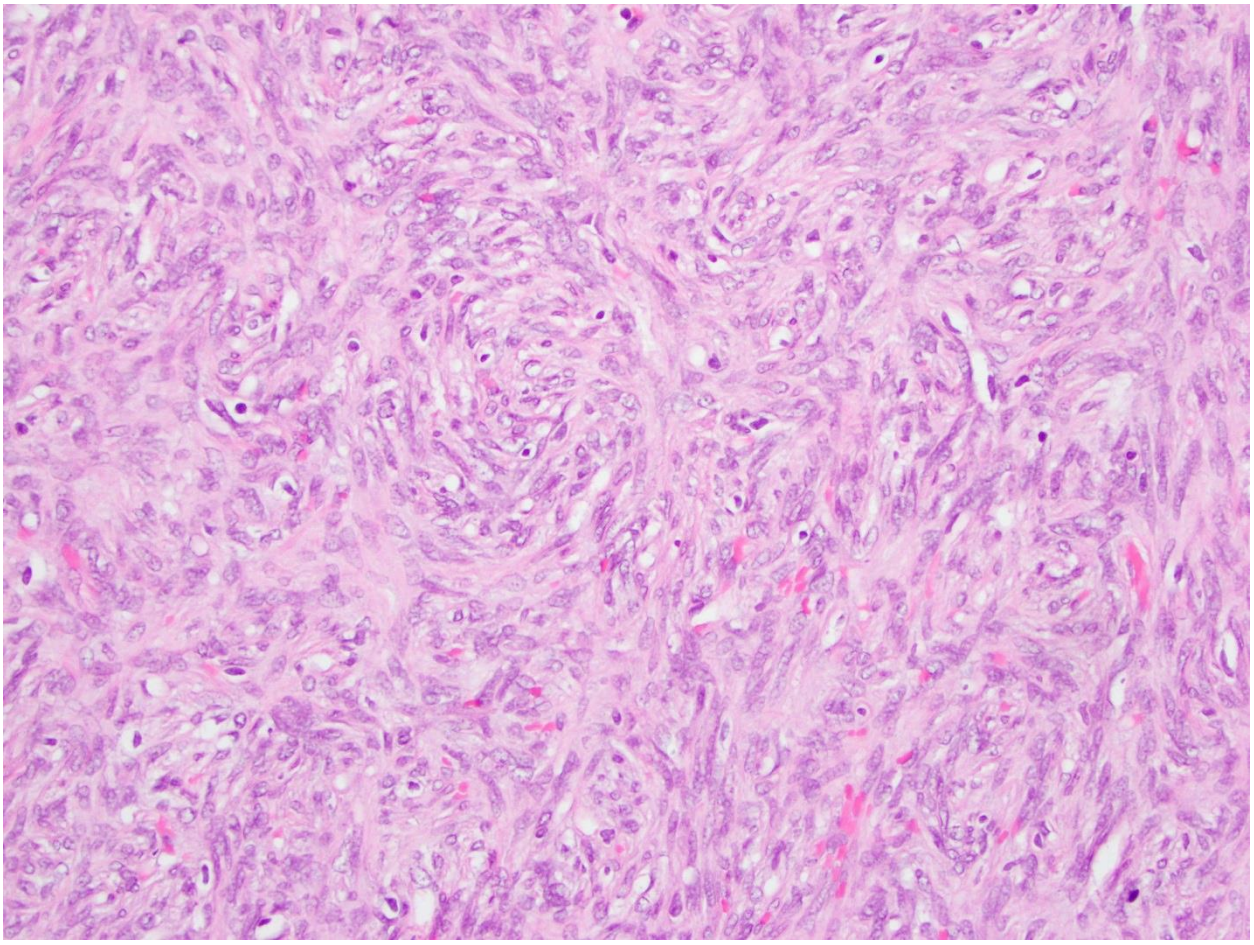

Figure 4. H&E, (200x).

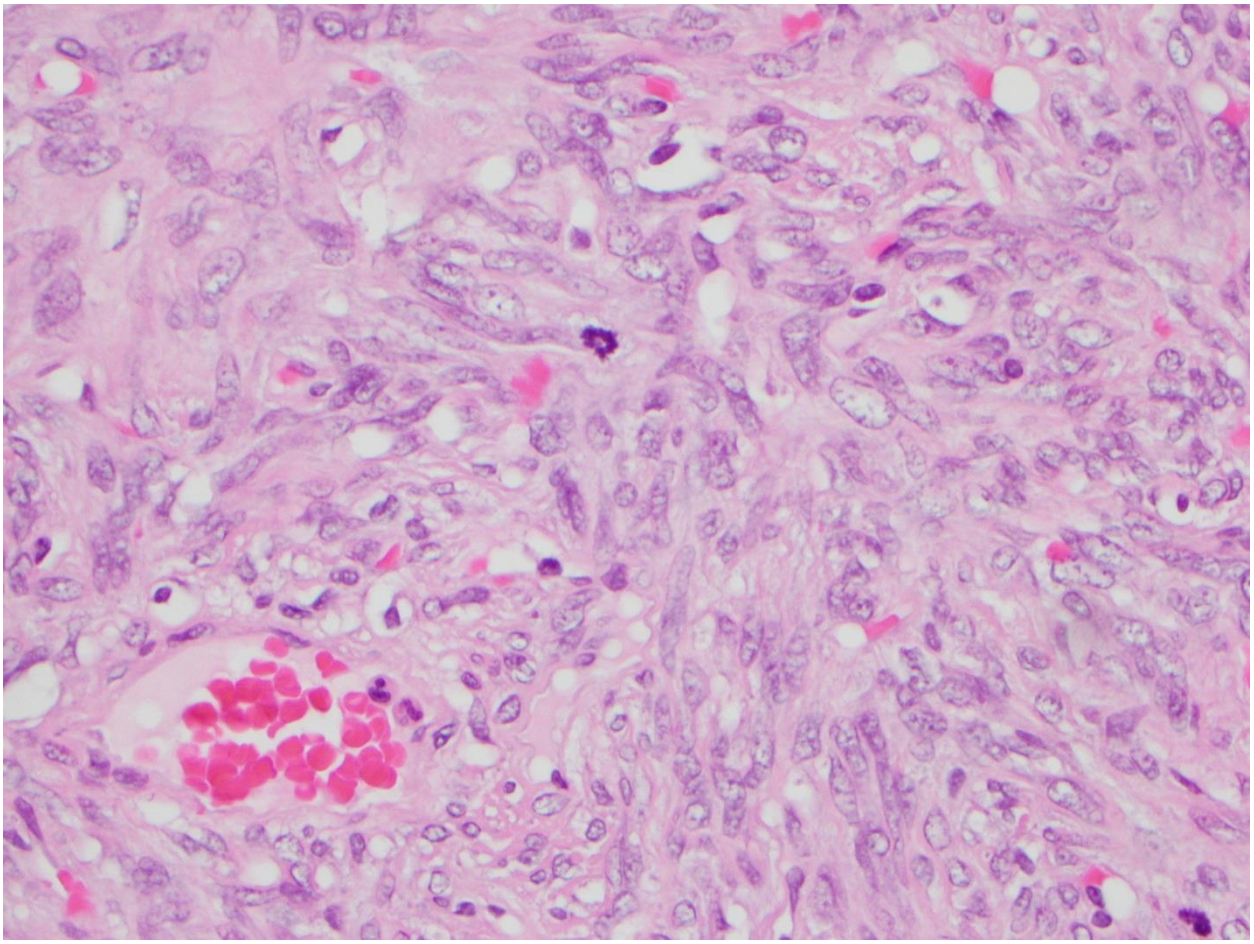

Figure 5. H&E, (400x).

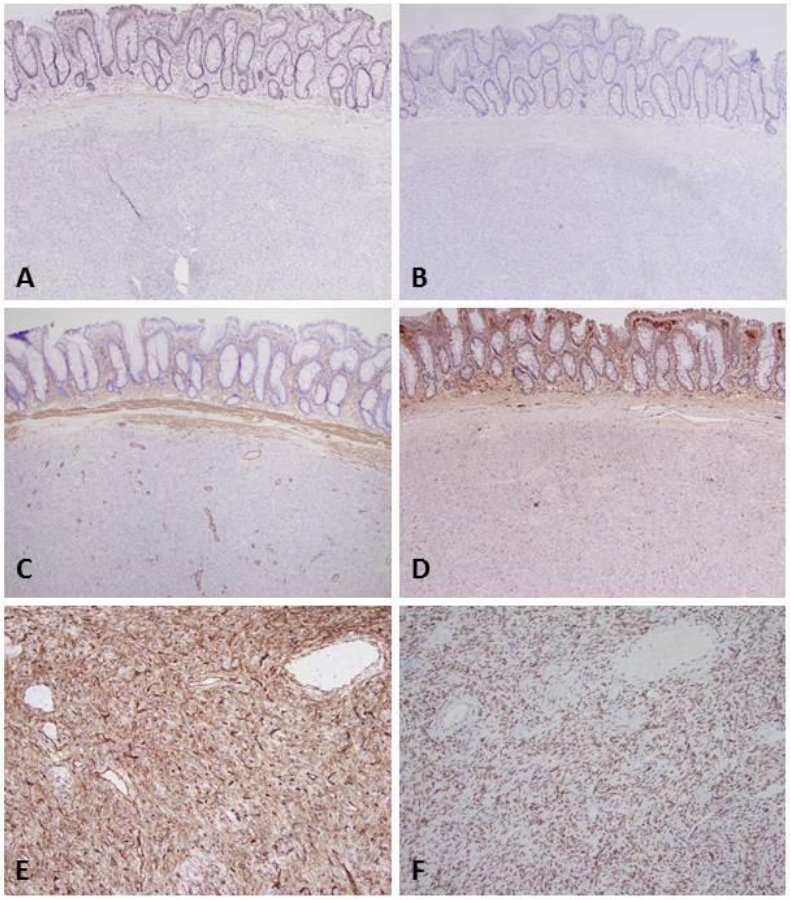

Figure 6. IHC stains for c-Kit(A), DOG1(B), SMA(C), S100(D), CD34(E), and STAT6(F).

What is the most likely diagnosis?

A- Schwannoma.
B- Leiomyoma.
C- Solitary fibrous tumor (SFT).
D- Gastrointestinal stromal tumor (GIST).
E- Inflammatory fibroid polyp (IFP).

Correct Diagnosis: (C) Solitary fibrous tumor

Case discussion

Histologic sections of the largest resected polyp demonstrate a well-demarcated submucosal spindle cell tumor (Fig. 1) with scattered, ectatic, irregularly shaped blood vessels (Fig. 2), and areas which are more sclerotic (Fig. 3). On higher power, the spindle cells are arranged haphazardly with no particular pattern and are cytologically bland without any striking atypia (Fig. 4). Infrequent mitotic figures are seen (Fig. 5). By immunohistochemistry (IHC), the neoplastic cells are negative for c-Kit, DOG1, SMA, and S100; while they are positive for CD34 and STAT6 (Fig. 6 A-F). The combined morphologic and immunophenotypic findings are consistent with a solitary fibrous tumor (SFT). The remainder of the resected polyps were tubular adenomas and hyperplastic polyps.

SFT is a rare benign mesenchymal tumor, which affects middle-aged adults of both sexes equally with a wide anatomic distribution. Clinically it presents as a slowly growing painless mass. Most tumors are well-delineated, exophytic, firm, and lobulated with gray-white cut surfaces and occasional hemorrhage [1]. The tumor most commonly presents in the pleura/lung as a pleural SFT, or less commonly as an extra-pleural SFT in the retroperitoneum, abdominal cavity, trunk, head and neck, and extremities [2]. It is rarely reported in the luminal gastrointestinal (GI) tract. It has been described arising from the subserosa/serosa of the colon and stomach [3, 4]. To the best of our knowledge this is only the second case reported as a polyp arising from the submucosal layer of the colon [5], and the third case overall of SFT arising within the colonic wall [6].

Microscopically these neoplasms characteristically demonstrate a haphazard “patternless” architecture (rarely fascicular or whirling growth) with alternating hypercellular and hypocellular areas, variable stromal collagen, and thin-walled dilated branching vessels (hemangiopericytoma like “staghorn” vessels). Cytologically, the spindle cells typically lack significant atypia with distinct ovoid to short spindled nuclei, and indistinct cytoplasm [7].

By IHC, the tumor cells are positive for CD34 in 95% of cases. CD34 is not a specific marker of SFT, as many other tumor types are positive for CD34 including gastrointestinal stromal tumor (GIST) and inflammatory fibroid polyp (IFP) which are in the differential diagnosis. The most specific immunohistochemical marker is STAT6 which shows diffuse nuclear positivity [8].

Although the majority of SFTs behave in a benign fashion, a subset (10%) have an aggressive clinical course. Demicco *et al.* validated their risk stratification scheme to help predict which SFTs may behave aggressively. Using the variables including age, tumor size, mitotic count, and tumor necrosis, allows tumor stratification into low, intermediate, or high risk groups with a 5 year risk of metastatic disease of 0%, 10%, and 73% respectively [9]. High risk SFT behaves as an aggressive sarcoma, and cellularity by itself does not predict prognosis [9]. Taking the above risk stratification scheme into consideration, our case corresponds to a low risk category with a total score of 3 (age >55, tumor size < 5cm, mitotic count > 4 per 10 HPF, and tumor necrosis absent).

Answer A. Schwannomas of the colon are rare and the large bowel represents the least common location for GI tract schwannomas [10]. They are benign spindle cell neoplasms that originate from Schwann cells and present as a well-circumscribed but not always encapsulated mural lesion. [10]. When arising in the GI tract, schwannomas typically do not show the classic histologic features of their soft tissue counterparts, such as palisading and hyalinized vessels, however, a characteristic peripheral cuff of lymphoid aggregates is typically seen [11]. Nuclear atypia might be seen in old lesions (ancient schwannoma). Mitotic figures are usually rare to absent. IHC stains are positive for S100 and GFAP; while negative for CD34 [12].

Answer B. Leiomyoma of the colon is a benign smooth muscle tumor, generally discovered incidentally during colonoscopy. These tumors grossly present as a well-circumscribed nodules arising from the muscularis mucosae, and are composed of bundles of uniform smooth muscle spindle cells with eosinophilic cytoplasm, blunt-ended nuclei, and fine chromatin with variable cytoplasmic vacuoles. IHC stains are positive for smooth muscle actin, desmin, h-caldesmon, while they are negative for CD34, CD117, and S100 [13].

Answer D. GISTs are the most common mesenchymal neoplasm of the GI tract, arising from the interstitial cells of Cajal. The most common location for GISTs is the stomach followed by the small intestine; colonic GISTs represent only 1-2% of cases [14]. GISTs present as a well-circumscribed mural lesion that has a fleshy cut surface. Microscopically tumors show spindled, epithelioid, or mixed cell morphology and are arranged in sheets and fascicles. The nuclei are relatively monomorphic, with fine chromatin, inconspicuous nucleoli, and collagenous-to-myxoid stroma. No prominent vasculature is seen. Immunohistochemical stains for CD117/c-Kit are positive in 95% of cases; DOG-1 is more specific and sensitive and is positive in 98% of cases [15].

Answer E. IFPs are most commonly seen in the stomach, arising from the submucosal layer and are rarely seen in the colon [16]. These benign neoplasms are variably cellular, comprised of bland spindle cells with interspersed inflammatory cell infiltrate consisting most notably of eosinophils. Scattered thin-walled blood vessels typically demonstrate characteristic whorled/“onion skin” fibrosis. By IHC, these neoplasms are positive for CD34 and show variable expression of smooth muscle actin; they are negative for CKIT, DOG1, and S100 [16].

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