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Jejunal Desmoid Tumor after Roux-en-Y Gastric Bypass Mimicking GIST

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Background	Intraabdominal desmoid tumors are benign fibrous masses, accounting for approximately 0.03% of all neoplasms. In the setting of bariatric surgery, desmoid tumors are exceptionally uncommon, with just two prior reports. We present the first case of a jejunal desmoid tumor diagnosed two years after Roux-en-Y gastric bypass (RYGB).
Summary	A 41-year-old woman with a history of Roux-en-Y gastric bypass (RYGB) two years prior for morbid obesity presented with diffuse abdominal pain, nausea, and malaise. Computed tomography (CT) scan revealed a 6 cm mass in the mesentery near the jejunojejunostomy and gastrojejunostomy. Given these findings, magnetic resonance imaging (MRI) was ordered, demonstrating a 6.8 × 5.6 × 6.8 cm cystic-solid mass arising from the proximal jejunal wall near the anastomosis, suggestive of a gastrointestinal stromal tumor (GIST). Exploratory laparotomy revealed a large, solid tumor originating from the redundant jejunal segment of the biliopancreatic limb post-RYGB. Resection of the redundant jejunum with the tumor was performed, preserving the jejunojejunostomy. Histopathology and immunohistochemistry confirmed the mass as a desmoid tumor.
Conclusion	Desmoid tumors arising from the jejunum are exceptionally rare, with their diagnosis relying solely on case reports scattered throughout the literature. This case presentation contributes to this limited knowledge base by documenting the first reported instance of a jejunal DT developing specifically after RYGB surgery.
Key Words	desmoid tumor; Roux-en-Y gastric bypass; bariatric surgery

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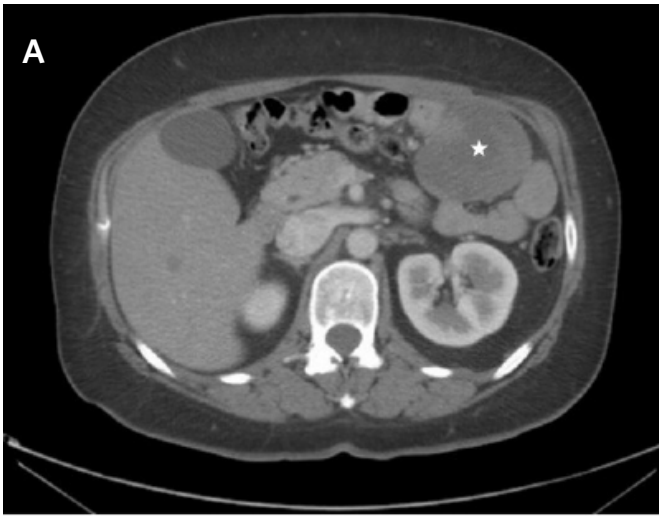
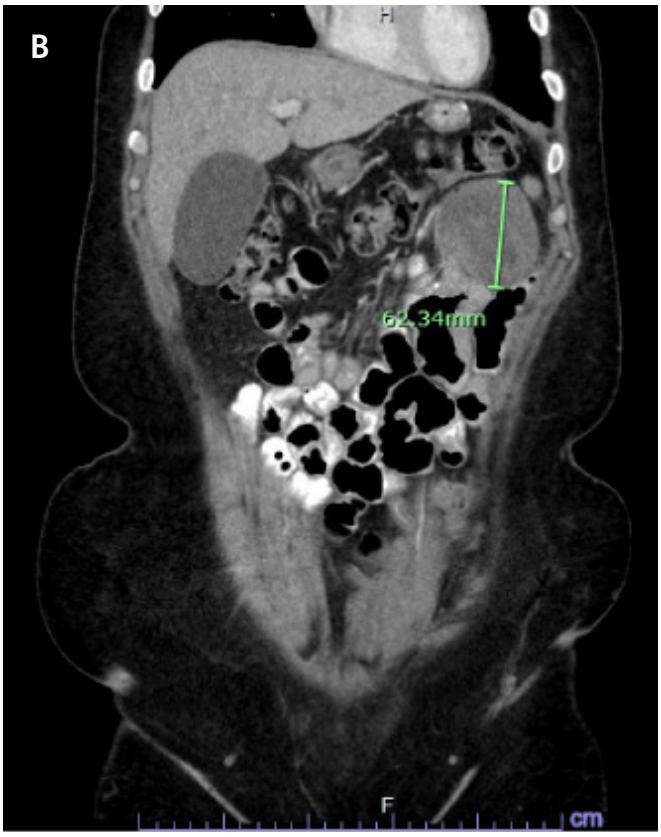
Case Description

Hirschsprung's disease (HD) is a uncommon development dA 41-year-old woman presents with a triad of mild diffuse abdominal pain, nausea, and malaise. Abdominal CT scan revealed a 6 cm cystic-solid heterogenous complex mass in the left upper quadrant (Figure 1). Her past medical history is significant for morbid obesity, currently controlled hypertension, and prediabetes status following Roux-en-Y gastric bypass (RYGB) two years prior. Surgical history includes left salpingectomy for ectopic pregnancy in 2006 and total laparoscopic hysterectomy for cervical intraepithelial neoplasia grade 2 (CIN-2), dysfunctional uterine bleeding, and severe dysmenorrhea in 2011. Family history is notable for breast cancer in her sister and trisomy 4 in her eldest son. Physical exam revealed only mild left upper quadrant abdominal tenderness.

Figure 1. CT Scan of Well-defined Heterogeneous Mass in Left Upper Quadrant. Published with Permission

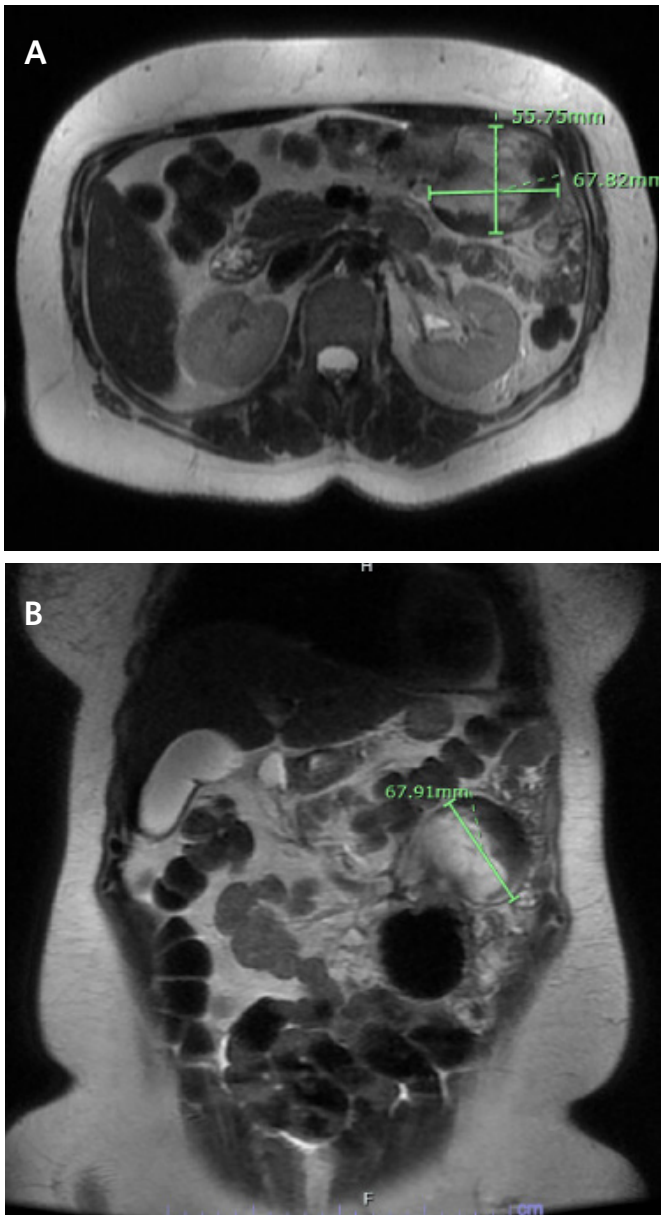




A) Horizontal view (★tumor); B) coronal view

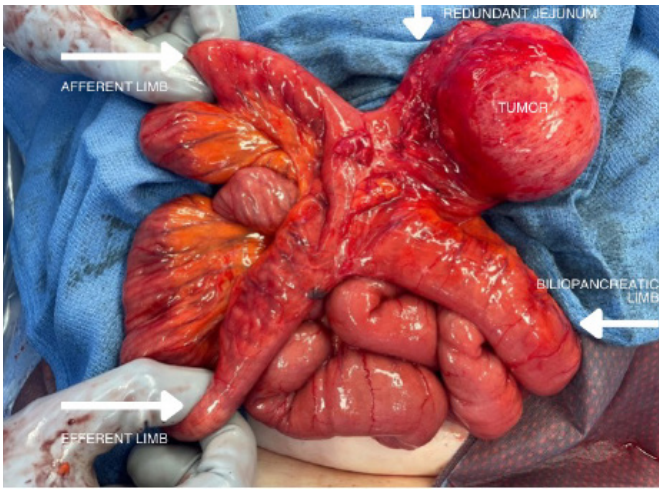
MRI further characterized the abdominal mass, revealing a 6.8 cm lesion arising from the proximal jejunal wall near the left upper quadrant gastrojejunostomy anastomosis. The mass exhibited ill-defined central enhancement with peripheral cystic degeneration, suggestive of a small bowel neoplasm (Figure 2). Based on these imaging findings, a provisional diagnosis of gastrointestinal stromal tumor (GIST) was favored. Percutaneous or endoscopic biopsy of the mass was deferred due to concern for peritoneal seeding.

Figure 2. MRI of Abdomen and Pelvis. Published with Permission



A left upper abdominal mass measuring 6.8 × 5.6 × 6.8 cm is observed to originate from a proximal jejunal loop in close proximity to the gastrojejunostomy anastomosis. The lesion displays ill-defined central enhancement near its attachment to the small bowel, surrounded by T2 hyperintense cystic contents and a peripheral T2 hypointense rim. Panels A and B depict horizontal and coronal views, respectively.

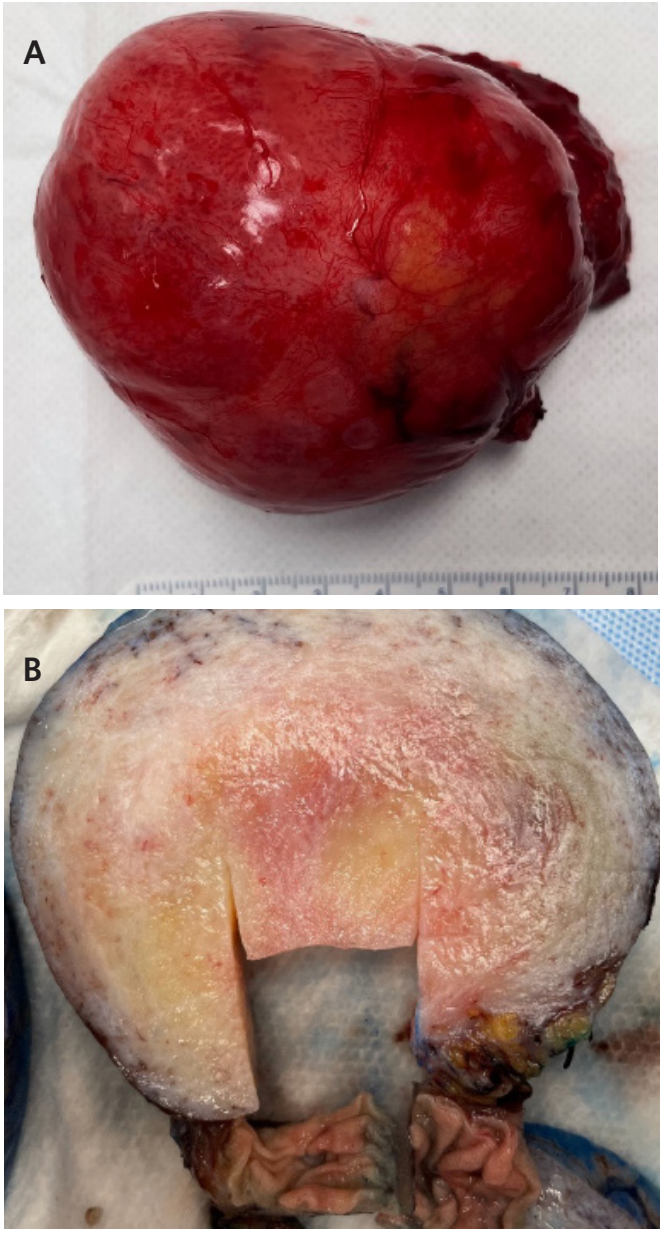
Figure 3. Intraoperative Findings. Published with Permission



Mass originating from the redundant jejunum and invading the mesojejunum, measuring 8.5 ? 7.0 ? 7.0 cm.

Exploratory laparotomy revealed a large, well-vascularized smooth surfaced mass arising from the redundant jejunal wall. The jejunojejunostomy anastomosis created after Roux-en-Y gastric bypass (RYGB) displayed clear margins (Figure 3 and Figure 4). The mass and surrounding redundant jejunum were resected en bloc, achieving adequate gross margins proximally and distally while preserving the original jejunojejunostomy. Postoperatively, the patient experienced mild nausea, vomiting, and persistent spinal headaches requiring intervention. She was ultimately discharged on postoperative day 6.

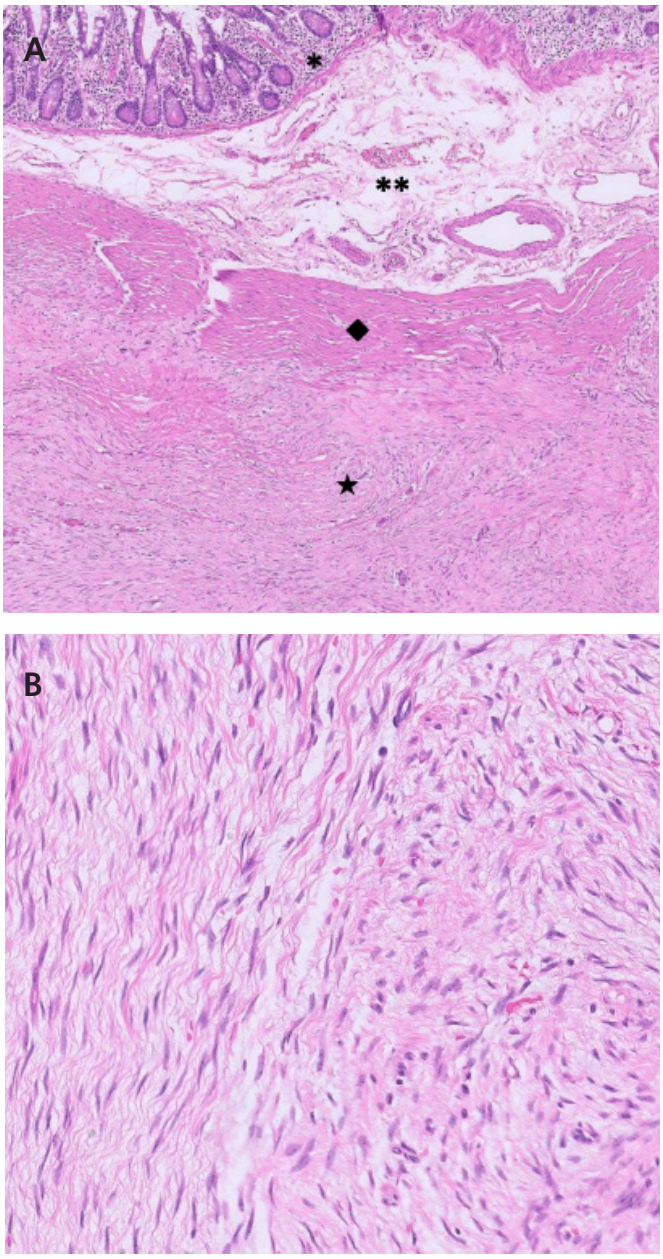
Figure 4. Excised Tumor. Published with Permission



A) Tumor post-resection; B) Cross-section displaying tan-white to pink-yellow coloration, striated/whorled texture, and rubbery consistency

Histopathological examination revealed a subserosal fibromatosis extending into the small bowel wall without mucosal invasion (Figure 5A: H&E, x50). Higher magnification (Figure 5B: H&E, x400) showed spindle cell proliferation with somewhat ill-defined borders, comprised of uniform cells with plump ovoid nuclei and ample eosinophilic cytoplasm. The stroma was collagenous, focally edematous, and contained thin-walled small vessels scattered throughout the lesion. Additionally, some areas showed hyalinized, keloid-like changes. Resection margins were negative.

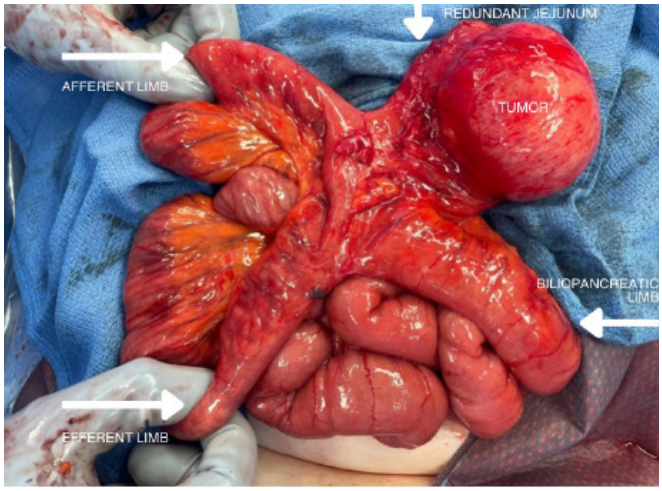
Figure 5. Hematoxylin and Eosin (H&E) Stains (50x and 400x, respectively). Published with Permission



*A) Predominantly subserosal involvement characterized by an indistinct spindle cell proliferation, with localized extension into the muscularis propria (*mucosa; **submucosa; ◆muscularis propria; ★tumor). B Proliferation of spindle-shaped cells featuring elongated, slender, and uniformly shaped nuclei with pale cytoplasm set within a collagenous stroma.*

Immunohistochemistry (IHC) demonstrated positive beta-catenin staining (Figure 6), focally positive smooth muscle actin (SMA), and nuclear positivity of cyclin D1 in spindle cells. Properly controlled immunoperoxidase stains for CD117, DOG-1, desmin, S100, CD34, and calretinin were negative. These findings are diagnostic of a desmoid tumor.

Figure 6. Immunohistochemical Stain, 400x. Published with Permission



Demonstrating positive nuclear staining for beta-catenin in tumor cells

Discussion

The global obesity epidemic has reached alarming proportions, exceeding 33% prevalence in recent decades. This surge is accompanied by concerning mortality rates, with estimates suggesting obesity contributed to 3.4 million deaths in 2010.[1]

Bariatric surgery stands as the most effective treatment for obesity and its comorbidities, with strong short- and long-term data supporting its use. It is one of the most commonly performed gastrointestinal procedures worldwide.[2] While some studies report conflicting results, the majority demonstrate a reduced risk of cancer, particularly obesity-related cancers, in patients who undergo bariatric surgery.[3,4] The development of gastric cancer post-surgery is rare, with limited evidence suggesting a direct link between bariatric surgery and gastrointestinal tumor formation.[5-7]

Intraabdominal desmoid tumors (also known as desmoid-type fibromatosis or aggressive fibromatosis) are locally aggressive neoplasms arising primarily in the mesentery or retroperitoneum. These tumors infiltrate adjacent tissues but rarely metastasize.[8,9] Patients with familial adenomatous polyposis (FAP) have a significantly increased risk, with an incidence of 10-20%.[10-13] While extremity desmoids can occur, half of desmoid tumors are intra-abdominal, with the remaining half affecting the abdominal wall.[10] Notably, the risk of IADT development in FAP patients is 852 times higher compared to the general population.[14]

Intraabdominal desmoid tumors originating from the small bowel are rare, with only a handful of cases reported in the literature.[15-17] Similarly, gastric bypass surgery appears to be an uncommon risk factor, documented in just two prior cases (Table 1).[9,15] The terminology used to describe these tumors has evolved over time. Initially, McFarlane defined it as fibromatosis in 1832.[18,19] Later, in 1838, Mueller proposed replacing "aggressive fibromatosis" with "desmoid tumor." Currently, the World Health Organization categorizes deep fibromatosis, aggressive fibromatosis, and desmoid tumor under the unifying term "desmoid-type fibromatosis."

Table 1. Prior Cases of Desmoid Tumors Post-Gastric Bypass in Literature

Reference No.	Gender	Years after Sx	Site	Size (mm)	Treatment	Preoperative diagnosis	Recurrence
Our case	F	2	Jejunum	85	Resection	GIST	Disease-free
9	F	1.5	Mesentery	180	Resection	GIST	Disease-free
15	M	3	Jejunum	158	Resection	GIST	Disease-free

Primary desmoid tumors are rare and represent a small fraction, comprising only 0.03% of all neoplasms and less than 3% of soft tissue neoplasms.[20] The estimated yearly incidence in the general population ranges from 2 to 4 cases per million individuals.[15] These tumors predominantly affect individuals aged 15 to 60 years but can occur across other age groups, including women post-childbirth. The female-to-male gender ratio is 2:1, a proportion that remains consistent in children. Notably, there is no significant racial or ethnic predilection.[21]

The etiology of desmoid tumors remains unclear. However, the presence of clonal chromosomal changes in a significant number of cases suggests that these tumors are neoplastic in nature.[22,23] Further, emerging evidence implicates dysregulated wound healing in the pathogenesis of these and other fibroblastic lesions.

The long-term oncological risk in the gastric remnant and the afferent/efferent jejunal limbs following RYGB remains unclear. While in our case, the redundant jejunal segment is excluded from the alimentary channel, potentially mitigating exposure to exogenous carcinogens. However, a theoretical concern centers around chronic exposure to stagnant bile as experimental studies have established a link between bile acids and gastrointestinal carcinogenesis.

Gastric restrictive surgery may theoretically contribute to tumor growth through several mechanisms. Prolonged food exposure within the gastrointestinal tract could increase contact with potential exogenous carcinogens. Furthermore, chronic irritation and inflammation in the redundant jejunal pouch due to increased pressure, along with potential inflammation from surgical materials, are hypothesized as potential contributors to carcinogenesis.[24] However, these hypotheses are speculative due to limited case reports. Most cases were diagnosed at an advanced stage, with patients experiencing nonspecific UGI symptoms such as epigastric pain, nausea, and vomiting for extended periods before diagnosis.

Following bariatric surgery, there's heightened concern for delayed diagnosis of GI malignancies. Postoperative nausea and vomiting (PONV) are frequently dismissed as dietary indiscretion (hard foods, overeating) or related to the surgical procedure (gastroenterostomy). However, intramural tumor growth within the GI tract can be substantial before causing overt symptoms. Furthermore, the creation of long Roux-en-Y limbs or biliopancreatic diversions significantly complicates endoscopic visualization of the GI tract.

Desmoid tumors exhibit a distinct anatomical distribution: roughly 50% occur in the retroperitoneum, 9% in the mesentery, and 40% originate from the small bowel wall.[25] Sporadic cases are often linked to abdominal surgery, pregnancy, hormone therapy, or Gardner's syndrome. In contrast, intraabdominal desmoid tumors are rare and typically associated with FAP.[15] To our knowledge, only two documented cases of desmoid tumors following gastric bypass surgery exist, with no prior reports of this tumor arising specifically from the jejunum after RYGB.

The symptoms of desmoid tumors are nonspecific and depend on the size and location, ranging from nonspecific pain and discomfort to obstructive and mass effect symptoms.[9,26] Our patient primarily reported abdominal discomfort with recent nausea and malaise, which are nonspecific findings.

Cross-sectional imaging with CT or MRI is crucial to define the tumor's relationship with adjacent structures. This information guides surgical feasibility and treatment decisions. However, radiographic differentiation between desmoid tumors and malignant soft tissue tumors can be challenging. Additionally, staging radiographic studies for distant spread are unnecessary, as desmoids typically lack regional or distant metastasis.[27]

The diagnosis is typically established through histopathology because the tumor may mimic other tumors, both grossly and radiologically. This case exemplifies this challenge, as the patient's presentation mimicked a gastrointestinal stromal tumor (GIST). Immunohistochemistry played a crucial role in definitive diagnosis. The absence of staining for Desmin, CD34, and S-100 excluded fibrosarcoma, leiomyoma, leiomyosarcoma, and malignant peripheral nerve sheath tumors. Additionally, the negative staining for CD117 and DOG-1 effectively ruled out GIST. Conversely, positive beta-catenin staining confirmed the diagnosis of desmoid tumor.

R0 resection with negative margins remains the gold standard for resectable desmoid tumors, offering a 25% to 50% reduction in recurrence rates.[28] However, experts advocate a cautious approach, especially for static tumors, due to associated surgical morbidity and relatively high recurrence rates. For unresectable intraabdominal desmoid tumors, particularly those involving the mesentery or encasing vessels and/or organs, especially in Gardner syndrome patients, a multimodality treatment strategy is often favored.[29,30] This approach typically involves initial medical therapy, with options including noncytotoxic agents such

as NSAIDs or celecoxib, tamoxifen, radiotherapy, targeted therapy with imatinib, or cytotoxic chemotherapy with agents like vinblastine, methotrexate, doxorubicin, and dacarbazine.

Sporadic desmoids are most common, with 5% to 15% associated with FAP,[10,31,32] caused by mutations in the adenomatous polyposis coli (APC) gene (chromosome 5q21-q22). Due to the predominance of abdominal desmoids in FAP),[10] some experts suggest screening for FAP (colonoscopy or genetic testing) in patients with intra-abdominal or truncal desmoid tumors.[32] However, the yield for detecting FAP in such patients without a prior history is relatively low (4% to 5% in two separate series[32,33]). Therefore, we propose a selective approach, not recommending universal FAP screening for all patients with intra-abdominal or truncal desmoids. Consequently, a screening colonoscopy was not advised for this patient.

Currently, there are no established evidence-based protocols for post-treatment surveillance. The National Comprehensive Cancer Network (NCCN) recommends history, physical examination, and appropriate imaging (q3-6 months for 2 to 3 years) followed by annual evaluations.[34] In our case, we proposed a more intensive regimen for this patient: follow-up with an oncologist, including clinical examinations and alternating CT and MRI scans every six months for the first two years. This would transition to annual assessments until year six and then biennial evaluations after that.

Sporadic desmoid tumors frequently harbor somatic APC mutations and activating mutations in CTNNB1, both leading to β-catenin accumulation.[35-38] The prevalence of CTNNB1 mutations exhibits some variability in sporadic desmoid tumors (39% to 87%), with larger and more recent studies estimating it around 85%.[39] Genetic screening may be particularly valuable for patients with multiple desmoids, a characteristic more frequent in the FAP group.

A targeted approach focusing on an "enriched population" has been proposed, directing screening towards individuals with desmoid tumors and specific risk factors such as: age below 40 years (11% yield), intra-abdominal or retroperitoneal location (5.4% yield), multifocal disease (29% yield), and family history (8% yield).[33] Given her medical and family histories, we recommended a genetic counseling session.

Conclusion

Desmoid tumors arising from the jejunum are exceptionally rare, with only a scant number of case reports documented in the literature. This case presentation adds to this limited body of knowledge by describing the first reported instance of a jejunal desmoid tumor developing after an RYGB surgery. While the association between desmoid tumors and surgical procedures is documented, the relationship between RYGB and jejunal desmoid tumors specifically is entirely novel.

Lessons Learned

Diagnosing a desmoid tumor is typically established based on histopathology because the tumor may mimic other tumors, like GIST, both grossly and radiologically. A multidisciplinary approach to individualized patient care is proposed for optimal therapeutic management.

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