YO29: ASYMPTOMATIC MESENTERIC DESMOID FIBROMATOSIS: A DIAGNOSTIC AND THERAPEUTIC CHALLENGE

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BACKGROUND

Desmoid fibromatoses are rare, benign neoplasms of myofibroblast origin that lack metastatic potential but can be locally invasive. Intra-abdominal desmoids may present as asymptomatic masses or with non-specific symptoms that may signify intestinal or urinary obstruction, vascular or neural involvement. Its clinical course is also unpredictable, with approximately 20% capable of spontaneous stabilization or regression. Management is often challenging and based on a multitude of factors such as symptomatology, tumor location, risk of recurrence and the possibility of treatment-induced morbidity. An upfront multidisciplinary team approach is recommended.

CASE PRESENTATION

A healthy 48 year old Filipino woman presents with a 2cm x 3cm incidental jejunal mesenteric mass intraoperatively during the excision of her choledochal cyst. The mass was not resected at the time due to its highly vascular location which, consequentially, may cause an unplanned and extensive bowel loss. Hence, only a core biopsy of the mass was done. Histopathology of the core-needle biopsy revealed desmoid fibromatosis. To assess for familial adenomatous polyposis (FAP), a colonoscopy was also done, yielding normal findings.

An initial watchful waiting approach was instituted since the patient was asymptomatic and resection would cause significant bowel loss. After three months, her abdominal CT scan showed that the mass had progressed, to 4.7 x 4.5 x 4.7cm. Anti-hormonal therapy with high-dose Tamoxifen (120mg daily) was initiated due to its limited toxicity and low cost.

The patient is currently asymptomatic, on sixth month of treatment, with good compliance. She will be undergoing a transvaginal ultrasound to assess for endometrial thickness. The tumor has remained stable at 4.5 x 4.6 x 4.7cm, on repeat abdominal imaging

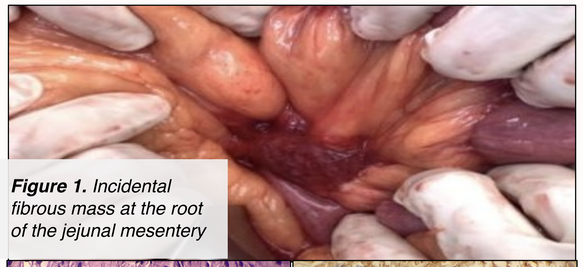

Figure 1. *Incidental fibrous mass at the root of the jejunal mesentery*

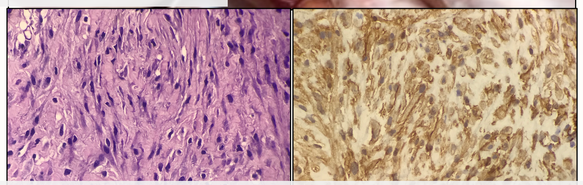

Figure 2. A: *Low-grade spindle cell neoplasm with long fascicles of elongated and slender spindle cells in collagenous stroma;* ***B:*** *β-Catenin is positive with nuclear and cytoplasmic staining*

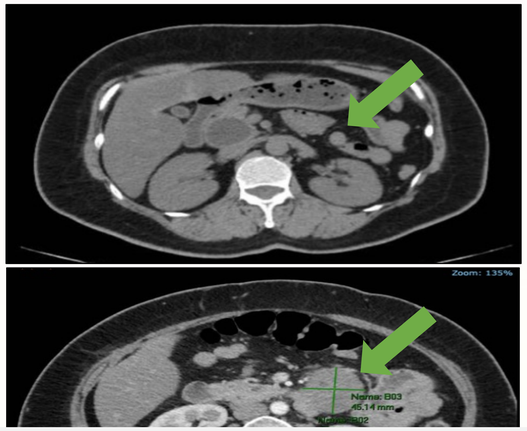

Figure 3. *A: On dynamic pancreas scan, a 1.5 x 1.5cm mass can be visualized near the jejunal segment, consistent with the desmoid tumor's intraoperative location. B: On an IV contrast-enhanced abdominal CT scan, there is progression of the mass to 4.7 x 4.5 x 4.7cm after 3 months*

DISCUSSION

In recent years, significant steps by international societies have been made to standardize treatment strategies among clinicians. Watchful waiting is the first line approach in all types of desmoid tumors. A decision towards more active treatment should be postponed until the occurrence of subsequent progression or an increase of symptom burden. Our patient had two documented progressions. Furthermore, her tumor originates at the root of the jejunal mesentery, a critical conduit for neurovascular structures supplying the small intestine. Further increases in the tumor's size may cause worrisome complications, such as bowel obstruction, ischemia, bleeding or perforation. For those failing initial observation, medical therapy, instead of surgical excision, has become the second-line option. Most studies and efficacious regimens include **sorafenib, pazopanib, vinorelbine and methotrexate with vinblastine**. Less established options include **imatinib**, **anti-estrogen therapy and NSAIDs**. It is possible to stop active therapy and resume watchful waiting in patients with stable disease for two years.

Learning Points

- **The rarity and unpredictability of desmoid fibromatoses warrants a multidisciplinary team approach to achieve appropriate management.**
- **Watchful waiting is the first line strategy for all types of desmoid fibromatoses.**
- **For unresectable, intra-abdominal desmoid tumors failing observation, systemic medical therapy is second-line.**

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