Rare presentation of Mesenteric Desmoids as Internal Herniation- Case report

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Abstract

Desmoids tumors are cytologically bland fibrous neoplasms originating from the musculoaponeurotic structures throughout the body. Desmoid tumors often appear as infiltrative, usually well-differentiated, firm overgrowths of fibrous tissue, and they are locally aggressive. The synonym aggressive fibromatosis describes the marked cellularity and aggressive local behavior. This course and the tendency for recurrence make the treatment of these relatively rare fibrous tumors challenging. We present a case of mesenteric desmoid which presentated as internal herniation into the transverse mesocolon. Internal hernia itself is a rare entity and big desmoid tumors having such peculiar presentation are difficult to diagnose preoperatively and hence can pose a diagnostic challenge for the surgeon and the radiologist.

Key wards: Internal herniation, internal hernia, mesenteric desmoids

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Case report

A 36 year old female patient came to Sawai Man Singh (SMS) hospital, jaipur OPD with chief complaints of lump lower abdomen since 1 year and pain in the epigatsrium region for last 2 months. Lump in lower abdomen initially small in size, gradually progressed to present size.. She underwent total abdominal hysterectomy via Pfannenstiel incision 2 months before admission for complaint of mennorrhagia; no histopathology report of specimen was available with the patient.

On examination the lump was non tender of size 12 x 12 cm, firm in consistency, smooth surface, globular in shape, well defined margins, freely mobile in all the directions and overlying skin could be freely moved. FNAC done was inconclusive. CT scan of abdomen revealed well defined smooth margined homogenous hypo dense mass lesion of

size12.5x7x13.5cm in right half of lumbar region. No definite internal contrast enhancement seen. Mass is displacing surrounding vessels and bowel loop fat plane in surrounding structures is clear.

On exploration we found a large mesenteric mass of size 12 x 15 cm arising from mesentery of proximal jejunum. The mass was herniating through transverse mesocolon and lesser sac along with small bowel loops along the lesser curvature of stomach (Figure 1 and 2). Finally excision of mesenteric mass with duodeno-jejunal resection anastomosis was done. Patient was discharged uneventfully on day 5. Biopsy revealed that morphology is consistent with diagnosis of fibromatosis (Desmoid) (Figure 3) of the mesentry. The tumor cells are negative for DOG1, Gastrointestinal Stromal Tumor (GIST), and S100. Sections given from intestine are unremarkable.

Figure 1 Large fibroid twisted around the mesentery

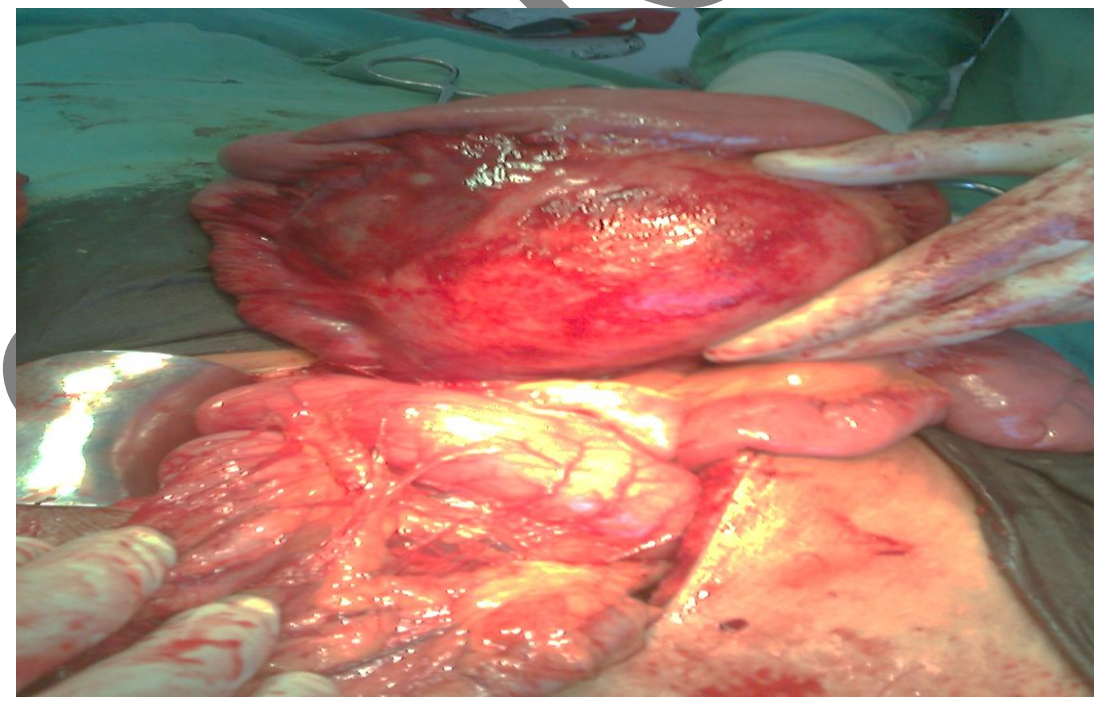


Figure 2: Fibroid herniating through transverse mesocolon

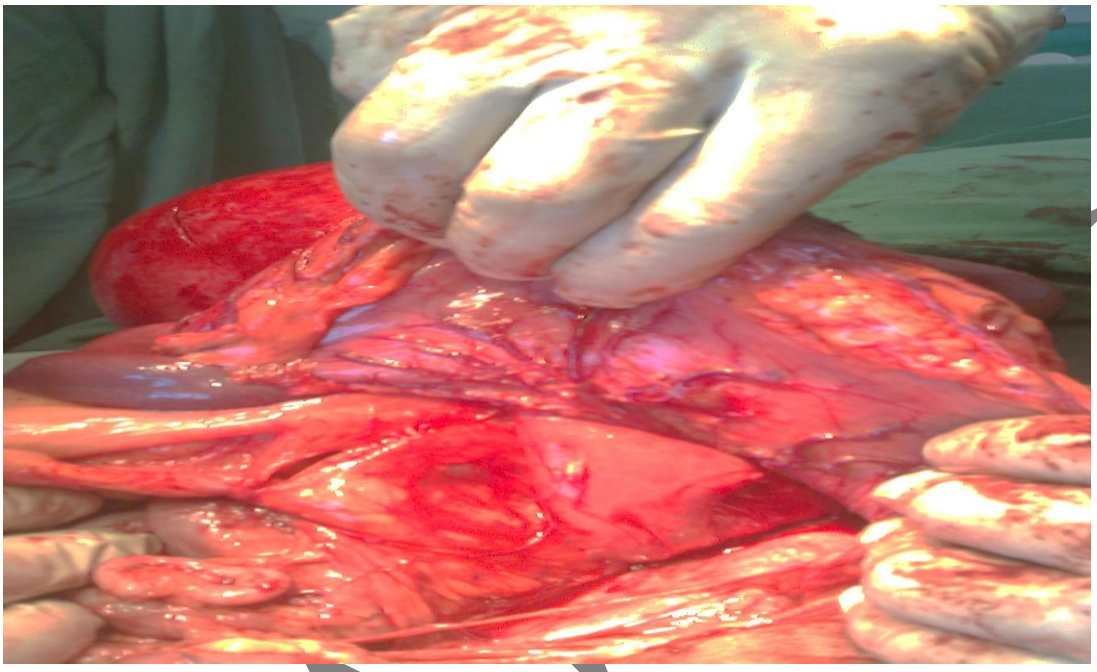
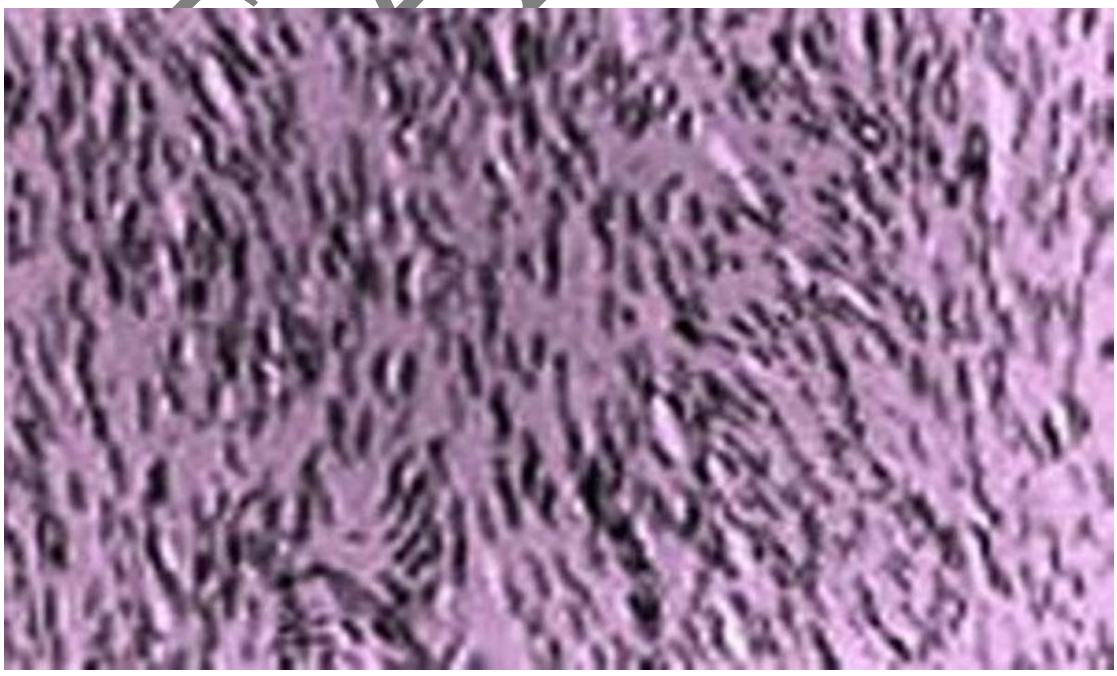


Figure 3 Histopathology



Discussion

Desmoid tumor, also known as aggressive fibromatosis or musculo-aponeurotic fibromatosis is a monoclonal, fibroblastic proliferation arising in musculoaponeurotic structures. Mueller in 1838 coined the term desmoid tumor (derived from the Greek *desmos* that means tendon-like). First description was given by McFarlane who reported it in the abdominal wall of a young woman after delivery in 1832.Histologically; these tumors consist of spindle-shaped cells in a collagenous matrix without the pleomorphic, atypical, or hyper chromatic nuclei of malignancy. Desmoid tumors have been recently [1] subdivided according to their location into extra-abdominal, abdominal and intra-abdominal. The intra-abdominal are further subdivided into mesenteric fibromatosis and pelvic fibromatosis. The frequency of desmoid tumors in the general population is 2.4 to 4.3 cases/million; this risk increases 1000-fold in patients with FAP.[2,3] The vast majority of desmoid tumors are sporadic, typically in young women during pregnancy or within a year of childbirth. Mesenteric desmoids account for less than 10% of sporadic desmoid tumors although they are a particularly common tumor in patients with FAP.[3] The association between desmoid tumor and FAP is particularly strong in the subset of patients with Gardner's syndrome. Intra-abdominal desmoids are more lethal than those that occur at other anatomic sites because of the possibility of bowel obstruction or ischemia. Although mesenteric desmoid tumors tend to be aggressive, there is considerable variability in their growth rate during the course of the disease. In fact, the biology of intra-abdominal desmoid tumors may be characterized by initial rapid growth followed by stability, or even regression.[4]Mesenteric desmoid tumors, by virtue of their relationship to vital structures and ability to infiltrate adjacent organs, may cause significant local complications requiring operative management, including intestinal obstruction, ischemia and perforation, hydronephrosis, and even aortic rupture. Despite these complications, the overall 10-year survival rate for patients with intra-abdominal desmoid tumors is 60% to 70 %.[3,5]. Practice parameters from the Standards Task Force of the American

Society of Colon and Rectal Surgeons, suggest that surgery should be reserved for small tumors with a well-defined and clearly resectable margin.[6] Reported recurrence rates for intra-abdominal desmoids are higher than for other sites and range from 57% to 86%, although surgery can be curative in select patients.[5]Given the high likelihood of recurrence and prolonged survival, even in the setting of advanced disease, some have suggested that a trial of watchful waiting, along with minimally toxic agents such as sulindac and antiestrogen therapy, may be the best strategy, particularly in patients with minimal symptoms. In this nascent era of target-specific biologic therapy, clinical response to imatinib by patients with heavily treated desmoid tumor has been reported. The observation that patients with desmoid tumors have partial tumor response and arrest of disease progression while on oral imatinib offers an alternative to surgical resection of desmoid tumors arising in the mesentery.[7]

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