Mesenteric Desmoid-type Fibromatosis with small and large bowel involvement: a case report

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Mesenteric Fibromatosis (MF), also called *intra-abdominal desmoid tumour* is a rare proliferative disease affecting the mesentery. It is part of the clinical-pathologic spectrum of deep fibromatosis, that are clonal fibroblastic/myofibroblastic tumor-like benign proliferations arised in the deep soft tissues, locally aggressive with potential to infiltrate or recur, but not metastasize. MF with intestinal involvement can be easily confused with other primary gastrointestinal tumours especially with that of the mesenchymal origin such as GISTs to varied potential malignant behavior. Misdiagnosis might result in inappropriate therapeutic decisions.
In June of current year (2013), a 29-year-old female was admitted to our hospital because of the intense abdominal pain lasting from a few days. Her past medical history was unremarkable.

The **physical examination** revealed a mass on palpation in the mid-abdomen that was easily movable. The physical examination was otherwise normal.

Laboratory findings were unremarkable and the level of CEA was within normal limits.
An *abdominal computer tomography* (CT) showed an 8x7x8 cm ovoid well-delineated hypodense mass located within the leaves of the ileal mesentery attached to the ileal and trasverse colonic bowel wall. The tumour enhanced poorly and homogenously with an intravenous contrast. There was no evidence of loco regional lymph-nodal involvement.

The patient was underwent to laparotomy for surgical excision en-block of the mesenteric mass together with a 58 cm segment of the ileal bowel and a 6 cm segment of transverse colonic bowel.
Results:

At gross examination, the specimen consisted of 8x7.6x8 cm ovoid well-circumscribed, nonencapsulated mesenteric mass, lined externally by peritoneal serosa and attached to the ileal and transverse colonic bowel wall. It was firm with an elastic consistency and cut with a gritty sensation; on cross section, the cut surface revealed a glistening white, coarsely fasciculated surface resembling scar tissues. There no necrotic areas.
Histologically, the lesion was composed of elongated, slender, spindle or stellate cells of uniform appearance arranged in sweeping bundles, set in a stroma with extensive keloid-like collagen deposition.
The cells lack hypercromasia or nuclear atypia and have small, pale-staining nuclei with minute nucleoli. No mitosis were found in 50 HPF.

These cells showed tentacular, melting insinuation into the muscolaris propria of ileal and colonic bowel wall without mucosal ulceration.
At immunohistochemical study, the cells stain positive for vimentin ...

... focally for smooth muscle actin and actin HHF35...
… but not for CD117, CD34, DOG1, desmin and S-100.
The first diagnostic hypothesis of a CD117-negative GIST was finally abandoned by the finding of diffuse Beta-catenin nuclear overexpression.

So, our diagnostic conclusion was “Intra-abdominal Mesenteric Desmoid-type Fibromatosis “.
Discussion:

MF is a intra-abdominal fibromatosis that develops in the mesentery or retroperitoneum.

According to Stout, fibromatosis has several distinguishing features: proliferation of well-differentiated fibroblast, infiltrative growth pattern, presence of variable amount of collagen between the proliferating cells, lack of cytological features of malignancy, scanty or absent mitotic activity and aggressive clinical behavior characterized by repeated local recurrences with no capacity of distant metastasis.
MF is relatively rare disease that may arise as an extracolonic lesion in patient with familial adenomatous polyposis such as FAP or especially Gardner’s syndrome, generally after previous abdominal surgery, a trauma or a hormonal stimulation, but also spontaneously. In these patients, MF is regarded as a proliferation of myofibroblasts that show APC gene mutations that lead to the overexpression of beta-catenin.

Nevertheless, MF has been reported also de novo in otherwise normal patients.
Conclusions:

MF with intestinal involvement can be easily confused with GISTs. A panel of gross, microscopic and immunoistochemical features allow, in the majority of cases, a correct identification. MF compared to GISTs is negative for CD34 and S-100 protein while the expression of vimentin, CD117, actin SMA, actin HHF35 and desmin do not seem to be remarkably different in the two neoplasm. In doubtful cases, it may be of diagnostic value to assess the nuclear beta-catenin that MF expresses as distinctive characteristic from other mesenchimal tumours.
References: