Giant Mesenteric Fibromatosis Associated with Non-Hodgkin Lymphoma. A Case Report and Literature Review

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Abstract

Background: Mesenteric fibromatosis are benign locally-aggressive mesenchymal neoplasms that lack the potential for metastasis. They are related to Gardner's Syndrome, previous trauma, abdominal surgery and prolonged intake of oestrogen. Differential diagnosis from similar tumours is crucial in order to establish the appropriate treatment and immunohistochemical features can only do the definitive diagnosis. Although medical therapies have a role for the treatment of mesenteric fibromatosis, surgical resection is the gold-standard procedure.

Methods: We are discussing about a 40-year-old male presented with a concomitant diagnosis of non-Hodking lymphoma and mesenteric fibromatosis not associated with any of the risk factors mentioned above. CT and PET scan are performed visualizing a vascularized and well-defined mesenteric center- abdominal solid mass with hypermetabolism contacting with the gastric body, the duodenum, the body and the tail of the pancreas, the transverse colon and the spleen. Ultrasound-guided trucut biopsy showed features suggestive of mesenteric fibromatosis.

Results: Elective laparotomy was done and a giant mass arising from mesentery was excised including partial gastrectomy and segmental resection of the transverse colon. Distal pancreatectomy, small bowel resection and successive splenectomy are performed due to a large hypertensive component. Postoperative period was uneventful. The histopathology of the surgical pieces is compatible with intra-abdominal desmoid fibromatosis.

Conclusion: As far as we know, it is the biggest case of mesenteric fibromatosis tumour excision reported in the literature. We also noticed that our case is the first reported case of concomitant presence of mesenteric fibromatosis and non-Hodgkin's lymphoma that has not relation to any described risk factor. More research is needed to establish what type of association this presentation may indicate.

Keywords: Mesenteric Fibromatosis; Desmoid Tumours; Non-Hodking Lymphoma

Introduction

Mesenteric fibromatosis is a rare intraabdominal desmoid tumour of the mesentery and constitutes a benign lesion characterized by proliferation of fibrous tissue with biological behaviour intermediate between benign fibrous lesions and fibrosarcomas [1]. It is locally invasive and tends to recur but never metastasize [2]. Its incidence is about 0.03% of all tumours [3], 0.73% among all abdominal...
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tumours and represents 8% of all desmoid neoplasm [2]. The aetiology is unknown with genetic, trauma, surgery and hormonal factors being implicated. These tumours may arise sporadically or in association with familiar adenomatous polyposis syndrome (FAP), especially Gardner’s syndrome [4]. Differential diagnosis from similar tumours has important clinical implications and can only be done by immunohistochemical evaluation [2,5]. The management of mesenteric fibromatosis is not well established. Chemotherapy and radiation have a role in the management of both primary and recurrent lesions, but surgical resection remains the cornerstone of treatment [2].

We report the first case of a man with a concomitant diagnosis of non-Hodgkin lymphoma and mesenteric fibromatosis in whom an elective laparotomy was performed in order to excise a giant mass involving pancreas, stomach and transverse colon. Finally, as the immunohistochemistry confirmed intra-abdominal desmoid fibromatosis, this report will review the literature concerning this extremely rare disease and discuss possible links between mesenteric fibromatosis and non-Hodgkin lymphoma.

Case Report

We present a case of a 40-year-old man with no medical or surgical history of interest in which leukocytosis with lymphocytosis with any apparent cause was detected in a routine blood test. The peripheral blood immunophenotype was compatible with chronic lymphoproliferative syndrome of B lymphocytes, immunophenotype CD5 + CD7+ CD19 + Kappa + FMC7 +. The patient didn’t report B symptoms but a palpable left inguinal adenopathy. The study was started by performing a CT scan, which reported a homogeneous splenomegaly of 18.5 cm and a lesion of 5.5 x 4.8 cm adjacent to the posterior wall of the stomach and the tail of the pancreas. PET scan evidenced splenic metabolically active disease and a pancreatic tail lesion. In addition, diffuse uptake in the gastric wall was observed (Figure 1).

Figure 1: First PET scan showing a lesion in the tail of the pancreas presenting pathological hypermetabolism (SUVmax 4.9). Spleen enlarged (SUVmax3.2). Diffuse uptake in the gastric wall (SUVmax5.2). A: axial; B sagittal sections.

After evaluation of the case in a haematological lymphomas and tumour committee, a splenic marginal lymphoma or a leukemized mantle lymphoma was suspected. Because it was an asymptomatic patient, it was decided to watch and wait and perform a bone marrow biopsy showing interstitial infiltration with focal paratrabecular reinforcement (10 - 12%) for low-grade non-Hodgkin’s lymphoma (positive for CD20 and bcl-2 and negative for bcl-6, CD25, CD10, CD5, CD23).

After 6 months of observation, the patient was presented in a committee for re-staging due to new symptomatology: night sweats of 1 month of evolution, dysthermic sensation and asthenia.

A new CT scan was then performed visualizing a voluminous, heterogeneous, vascularized and well-defined mesenteric center-abdominal solid mass with hypodense areas, measuring 25 x 12 x 26 cm in diameter. It presented air content in relation to fistulization and it was displacing the bowels and the mesenteric vessels to the right. This mass contacted extensively with the gastric body, the duodenum, the body and the tail of the pancreas, the transverse colon and the spleen, causing focal stenosis of the splenic vein (Figure 2). The subsequent PET scan showed hypermetabolism in a large abdominal mass (SUV max 4.3), splenomegaly (18 cm anterior-posterior) with a slight increase in diffuse splenic uptake (SUV max 2.4) (Figure 3).

Therefore, it was decided to perform ecoguided biopsy-trucut of the abdominal mass showing moderately-cell fusiform mesenchymal tumour with myxoid areas, elongated cells of fibroblastic appearance, without atypia, positive for AML, AME, focal calponin, intense H-caldesmon and beta catenin intranuclear positive, Ki 67 < 10%; all compatible with intraabdominal/mesenteric desmoid fibromatosis.

Elective surgery was planned and the patient was admitted in our hospital 24 hours before the intervention. A suprainfraumbilical laparotomy was performed, finding splenomegaly and the large mass described. We proceeded to the externalization of the mass: firstly, a partial gastrectomy of the major curvature of the stomach was performed. The mass of the splenic angle was liberated and the spleen could be respected initially. The section of transverse colon that was intimately attached to the lower portion of the mass was performed. Once the colon section was finished, the block exeresis was completed. Distal pancreatectomy and successive splenectomy were performed due to a large hypertensive component. We proceeded to reconstruction of transit through mechanical latero-lateral colo-colonic anastomosis and gastro-gastric anastomosis to correct the defect of the major gastric curvature. The surgical time was about 130 minutes.

Figure 2: Contrast enhanced computed tomography scan showing a voluminous, heterogeneous, vascularized and well-defined mesenteric center-abdominal solid mass with hypodense areas, measuring 25 x 12 x 26 cm (T x AP x L). It presents air content in relation to fistulization. A: axial; B sagittal sections.

After 8 days of admission, the patient evolved favourably and without any postoperative complication; CLAVIEN-DINDO: 0.

The histopathology of the surgical pieces revealed:

- **Sample A**: Spleen of 1490 g measuring 20 x 18 x 7 cm. Under the microscope, white pulp invasion was observed due to a lymphoid neoplasm with retrograde invasion of germinal centres. Immunophenotype: CD20, Bcl2, CD79 IgD, IgM and Ki 67 low.
- **Sample B**: Segment of intestine of 6.5 cm. No particularities.
- **Sample C**: Large well-delimited pseudocapsulated mass of 35 x 30 x 10 cm that weighed approximately 5565 g. It contained omentum, stomach (10 cm) and transverse colon (8 cm) (Figure 4). The tumour was white-pink colour with elastic consistency after fixation without foci of necrosis (Figure 5). Under the microscope, moderately-cell fusiform mesenchymal tumour with elongated cells of fibroblastic appearance without atypia was observed (Figure 6).

Immunohistochemistry showed positivity for muscle-specific actin (Figure 7), caldesmon (Figure 8) and intense nuclear positivity for beta-catenin (which guided the diagnosis to desmoid fibromatosis) (Figure 9). Low proliferation index (Ki67) (< 10%) (Figure 10). Free margins, although close.

Currently, the patient goes monthly for medical check-ups to External Consultations of General Surgery, Oncology and Haematology. The CT scan performed two months after the surgery did not visualize pathological alterations (Figure 11). Given that the patient is asymptomatic and with correct vaccination after splenectomy, we opt for monthly watch and wait without adding any other adjuvant therapy.
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**Figure 4:** Well circumscribed mass that includes an intestinal segment (without lesions in the mucosa).

**Figure 5:** Mass that has a pseudoencapsulated appearance with a whitish-pink color, without macroscopic areas of necrosis.

**Figure 6:** Hematoxylin-eosin stain showing proliferation of fusiform cells with fibroblastic appearance and elongated morphology without atypia.

**Figure 7**

Figures 7-10: Immunohistochemistry shows positivity for muscle-specific actin (Figure 7), caldesmon (Figure 8), and intense nuclear positivity for beta-catenin (which guides the diagnosis to desmoid fibromatosis) (Figure 9). Low proliferation index (Ki67) (< 10%) (Figure 10).
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Discussion

Mesenteric fibromatosis, also known as deep fibromatosis, aggressive fibromatosis or intraabdominal desmoid tumours are benign locally-aggressive mesenchymal neoplasms that lack the potential for metastasis [6]. John Macfarlane first described it more than 150 years ago. Currently, these rare tumours represent less than 3% of all soft tissue sarcomas and about 0.03% of all malignancies [4]. Desmoid tumours appear to be of aponeurotic origin and have a benign histologic appearance. It results from an aggressive proliferation of the fibroblast-myofibroblast. The real aetiology of mesenteric fibromatosis is unknown. Most reported cases have been in association with Gardner’s Syndrome, previous trauma and prolonged intake of oestrogen, but mesenteric fibromatosis can occur as a primary condition in the absence of any predisposing factor [8].

In the literature, some curious cases of mesenteric fibromatosis that have no relation with the previously described risk factors have been reported. Secondary desmoids have also been observed within radiation fields after treatment for several different types of cancer, including Hodgkin disease [9,10], neuroblastoma [3] and brain tumours [11]. However, it is the first reported case of concomitant presence of mesenteric fibromatosis and non-Hodgkin’s lymphoma that has not received any type of therapy (neither chemotherapy nor radiotherapy) before its diagnosis.

Measures of mesenteric fibromatosis tumours are variable. Lesions up to 24 cm have been reported and even a mesenteric fibromatosis tumour about 30 × 21 × 12 cm was excised by Mohammed Khalid Mirza Gari’s team [8]. The tumour described in this paper is one of the biggest cases of mesenteric fibromatosis tumour reported in the literature, measuring 35 x 30 x 10 cm.

The clinical presentation is variable since it ranges from the absence of symptoms to abdominal pain, constipation, vomiting and weight loss. In late phases, intestinal ischemia as well as organ compression symptoms such as small bowel obstruction and hydronephrosis can be present [13]. This happens because the tumour can grow inside the abdominal cavity. Disease progression of mesenteric
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fibromatosis is also unpredictable: it can develop from rapid growth to spontaneous tumour regression [14]. Growth of these tumours is slowest in young girls and reaches a peak at menopause, suggesting that oestrogen may act as a growth factor. Nevertheless, in our case, we've observed a very fast growth that cannot be explained by oestrogen mechanisms.

Differential diagnosis includes GIST, solitary fibrous tumour and inflammatory myofibroblastic tumours and immunohistochemical features establish definitive diagnosis.

Nevertheless, CT scan is mandatory in order to determine the site, size, extent of the lesion and degree of local invasion. Mesenteric fibromatosis are seen as hypodense well circumscribed homogenous masses [4]. Differentiating between GIST and mesenteric fibromatosis is important because while mesenteric fibromatosis tumours are usually benign, GISTs have malignant potential. Histology and immunohistochemistry is helpful in developing a treatment strategy since these diseases differ greatly in their initial approaches to management [5]. As in our case, immunohistochemistry shows diffuse nuclear positivity for Beta-catenin in mesenteric fibromatosis but not in GIST, solitary fibrous tumour and myofibroblastic tumours [2,4]. In addition, CD117 immunostain is regarded as one of the key criteria for the diagnosis of GIST and CD34 is often present in those tumours, but they are absent in mesenteric fibromatosis [15].

Although large randomized studies are not present due to the rarity of these tumours, medical and surgical therapies have been trialed for the treatment of mesenteric fibromatosis. As initially in our case, watchful waiting has been advocated for asymptomatic patients as it has been shown to be equivalent to medical therapy in progression-free survival [5]. When symptoms are present, wide field surgical excision is the first line treatment because primary resection with negative margins is the most effective treatment. As noted in our case, the majority of these lesions require resection of the attached segment of the bowel [8]. However, local recurrence following resection of mesenteric fibromatosis is common. When complete resection may not be possible because of its size or intimate involvement with vital structures, other nonsurgical treatments including radiation and systemic drug therapy, such as tamoxifen, doxorubicin, dactinomycin, vincristine and cyclophosphamide and anti-inflammatory drugs (sulindac, indomethacin) can be useful as well as in cases of recurrence [5,6,8]. What is clear is that despite the treatment modality, the recurrence rate for mesenteric fibromatosis is 30% to 40% [5].

Conclusion

We noticed that our case is the only reported case where there is a synchronic non- Hodgkin's lymphoma and a mesenteric fibromatosis tumour. In our case, mesenteric fibromatosis was unusual since it was not associated with Gardner's syndrome, previous abdominal surgery, or familial adenomatous polyposis.

Since this is the first case report on a patient with these two neoplasms, it’s hard to denote what type of association this presentation may indicate. It may be a coincidental association; it may also be a non-random association that is yet to be revealed statistically.

Nevertheless, this report will contribute to and guide further investigation on the pathogenesis, pathophysiology and associative features of mesenteric fibromatosis in order to better detect and treat disease.

Conflict of Interest Statement

There are no financial disclosures and no conflict of interest.

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