Giant mesenteric fibromatosis

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Abstract. – Mesenteric fibromatosis is a proliferative fibroblastic neoplasia of the small intestine mesentery which may occur as a unique or multiple formation. Mesenteric fibromatosis represents the 8% of all desmoid neoplasm.

Giant mesenteric fibromatosis is uncommon by itself (2-4 case/million/year). Since the rarity of this tumor and the difficulties in diagnostic and therapeutic ambit, we believe it justified to describe a case of giant mesenteric fibromatosis which came to our observation.

Key Words: Mesenteric fibromatosis, Desmoid tumors.

Background

Mesenteric fibromatosis is a proliferative fibroblastic tumor of the small intestine mesentery, most rarely of the gastrohepatic and gastro splenic ligaments or transverse colon, which may occur as a unique or multiple formation.

Mesenteric fibromatosis represents the 8% of all desmoid neoplasm being an intra-abdominal variety. Desmoid neoplasia have an incidence of 0.03% among all neoplasia and present abdominal and extra-abdominal varieties.

Its clinical behaviour can be compared to fibrosarcoma and fibroma. In fact poorly differentiated varieties show a higher probability of local recidivation and an infiltration of the closer tissues but do have not metastastic capabilities. For this reason it is considered a benign tumor.

Giant mesenteric fibromatosis is uncommon by itself (2-4 case/million/year) as its association with other pathologies is from 3% to 45% in the Gardner's Syndrome and 10% in association with familial colonic polyposis.

Case Report

A 33-year-old man referred the onset of a tumidity of the abdomen which became evident because of substantial weight loss following a slimming diet.

Abdomen echography discovered a large hypoechoic mass of 20 cm extended beside the liver from the right to the left hypochondrium.

The CT of the abdomen and of the pelvis revealed the presence of an expanding process extended from the anterior-left peripancreatic space anteriorly positioned along the body-tail region. This expanding process which passed the medial line extended itself along the right parietal-colic sulcus until the homolateral pelvic-cavity then connecting in a contraction with the iliopsoas muscle.

This lesion was 180 mm and 90 mm in diameter and it resulted hypodense.

The surrounding retroperitoneal structures were moderately displaced and compressed but there were no signs of infiltration.

Objective evidence of abdomen showed the presence of a tumidity of hard-fibrous consistency, smooth surface, cleanly incised margins, fixed which extents itself from the epigastrium to the hypogastrium and from the left to the right hip.

Surgical operation: a total medial laparatomy was performed. After the opening of the peritoneum a large ovoid lesion was observed. It was placed obliquely from the left hypochondrium to the right iliac region, of hard-fibrous consistency, apparently coated by a well-defined capsule, which had taken origin from the mesentery below. The lesion is firmly tied to the mesentery of two adjoining bowel loops and was richly vascularized by several mesenteric arteries and veins.
We proceeded with the separation of the lesion from the mesenterial root that, although very difficult, was performed with absolutely vascularization sparing of the foresaid loops. Section between ligations of the vascular peduncle and resection of the lesion from its site.

Anatomical pathology (gross study): the lesion measured 25 × 14 × 10 cm and weighted 2750 g (Figures 1 and 2).

Anatomical pathology (light microscopy): the lesion is mainly composed of the proliferation of fusiform or rowform cells with ovoid nucleus, monomorphic, placed in an abundant stroma richly vascularized.

Mitotic index was low. No necrosis nor intratumoral inflammatory infiltration.

The wide sampling of the neoplasys's margins, can not reveal the presence of a real capsule, the growth's pattern result to be of expansive type and only locally is possible to notice a tight adherence with the peri-intestinal adipose tissue.

The patient had a regular post-operatory period and was discharged after 7 days clinically healed.

Four months after the operation the patient had an episode of intestinal obstruction and for that was submitted to urgent surgery. During the operation was noticed an excessive stretching of the intestinal loops of the 2nd and 3rd mesenterial arch, with thickened and edematous walls. In order to avoid a large intestinal resection, and for the difference of calibre of the loops themselves, we decided to perform a latero-lateral enteroanastomosis.

The patient had a regular post-operatory period. In later follow-ups and three years after the operation the patient was in good health conditions with no symptoms of a relapse.

Discussion

Among the aetio-patogenetic factors, the genetic one is the most important.

Infact the association of fibromatosis and Gardner’s syndrome, autosomal dominant disease characterized by familial colonic polyposis, osteomas, epidermoid cysts, is a prognostic aggravation of the Gardner’s syndrome itself: as well as a frequent malignant transformation of the colonic polyposis, fibromatosis relapses frequently a radical excision is the only possibility on many occasions.

Mesenteric fibromatosis, as we said before, is even associated with familial colonic polyposis caused by a mutation of the gene apc (adenomatous poliposys coli).

A previous laparotomy can be found in a fifty percent of patients with fibromatosis with a latency period of five years maximum.

The hormonal factor considered responsible of the growth of abdominal varieties of desmoid tumors is in relation to a larger oestrogenic activity during the pregnancy and therefore the treatment with tamoxifen finds a correct indication in these cases.
The symptoms of this tumor are often absent, until its enlarged dimensions cause intestinal occlusion, mesenteric ischaemia and ureteral compression. Surgical treatment is often due to the large dimensions of the mass, the local infiltration and the possibilities of a relapse. In a 50% of patients is necessary to perform a wider demolition. In the case we described the mass was completely taken out in its entirety. The occlusive complication occurred four months after the operation was probably due to a chronic suffering of the intestinal loops as a result of an excessive skeletization of the vascular pedicles caused by the tight adherences with the neoplasia.

References


