Sporadic aggressive fibromatosis of the colon and abdominal wall in an emergency setting

Francesca Cancellario d’Alena*, Carlo Di Marco**, Margherita Loponte***, Cesare Pirozzi°, Gazia Savino°°

*UOC Anatomia Patologica, AO “San Camillo” Hospital, Roma, Italy
**Dipartimento di Scienze Chirurgiche Policlinico “ Umberto I”, Sapienza Università di Roma, Roma, Italy
***Dipartimento di Scienze Chirurgiche sezione Durante, Policlinico “ Umberto I”, Sapienza Università di Roma, Roma, Italy
°UOC Chirurgia d’Urgenza, AO “San Camillo Forlanini”, Roma, Italy
°°Dipartimento di Scienze Chirurgiche Policlinico “ Umberto I”, Sapienza Università di Roma, Roma, Italy

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AIM: Our aim is to present an utterly unique case of sporadic aggressive fibromatosis (AF), infiltrating both the abdominal wall and the colon; and especially, to discuss the usefulness of porcine dermal meshes for the reconstruction of a large parietal gap in contaminated surgery and in an emergency setting.

CASE EXPERIENCE: We report the case of a 40 years old woman affected by sporadic AF, involving both the anterior abdominal wall and the colon, with an effective intestinal stricture. The surgery consisted in removing “en bloc” the portions of the colon and abdominal wall affected by fibromatosis, with no residual tumor (R0), that left an important parietal gap. A biological prosthesis of cross-linked acellular porcine dermal collagen (APDC) has been used for the contextual reconstruction of the abdominal wall, sutured inlay by a double line of non absorbable stitches. No complications have been observed. After one year follow up, there is no tumour recurrence and the abdominal wall has fully consolidated.

DISCUSSION: The peculiar problems arising about differential diagnosis, therapeutic indications and reconstructive surgical procedures are discussed, especially with regard to prosthetic implants in contaminated surgery and to cross-linked APDC prosthesis.

CONCLUSIONS: Complete surgical removal is the first line treatment in sporadic AF, whenever feasible. In the reported case, an R0 resection was obtained at the cost of a wide parietal gap. According to our experience, cross-linked APDC is effective for the prosthetic reconstruction of abdominal wall in contaminated surgery and in an emergency setting.

KEY WORDS: Abdominal wall, Aggressive fibromatosis, Desmoid tumor, Prosthetic devices, Reconstructive surgical procedures, desmoid tumor

Introduction

Aggressive fibromatosis (AF), also known as desmoid tumour (DT), is a rare neoplastic disease, characterized by fibrous/mioblastic proliferation, monoclonal in origin, not metastatic, but locally invasive and prone to recurrence 1,2. It originates most often from muscle-aponeurotic tissue, but may also arise from the connective tissue of mesentery. In this case, it has an intra-abdominal location (intra abdominal fibromatosis: IAF), and involves abdominal viscer 1,3, so a differential diagnosis with GIST may be essential 4,5. AF can be sporadic or FAP-related. Sporadic AF generally grows in striped muscles of the trunk and roots of the limbs, rarely inside the abdomen. It’s incidence is 2-4 new cases per million per year in the general population 6,7 - more frequently in young adults. In contrast, the incidence is much higher (800-1000 times) among people affected by famil-

Correspondence to: Cesare Pirozzi (e-mail: cespiro@live.it)

ial colorectal polyposis (FAP), owing to the APC gene mutation, usually found in these patients. In such cases, an intra-abdominal location (IAF) is more frequent. It is also well known the association of AF with several other conditions, such as trauma, surgery, pregnancy, Crohn's disease and prolonged treatment with estrogen, but it is not equally known the reason why. An emergency clinical onset (due to hemoperitoneum, abscess formation, rupture or other complications, such as hydronephrosis and intestinal stenosis) is rare but reported in the medical literature.

An intra-abdominal location in sporadic AF is rare in itself, lower than 5% of the cases; but must be regarded as exceptional when the abdominal wall is also involved. Actually, cases of desmoid tumor with synchronous infiltration of the abdominal wall and bowel are not recorded in the medical literature, as far as it results from the Medline and EMBASE search (query: “desmoid tumor” OR “aggressive fibromatosis”).

Complete surgical removal is usually the first line treatment in sporadic fibromatosis, although the incidence of recurrences is not negligible: up to 44.3% in a recent large collection of cases, lower in other records. Medical therapy, neoadjuvant or adjuvant, is rarely recommended in sporadic forms, not so in the FAP-related, in which recurrences are more frequent (up to 88% of cases) and different protocols are considered effective. In rare cases, when surgical removal is considered dangerous or mutilans, a hands-off approach can be adopted (wait and see): about one third of the reported cases have a slow progression, while the disease tends to remain stable in over half the cases, and in some cases may shrink.

In the case of large desmoid tumors of the abdominal wall, the direct closure after excision may be impossible and, consequently, a prosthetic mesh or more complex reconstructive procedures are needed. Seldom if ever, as in the reported case, the use of implants can pose some peculiar difficulties, because of the contemporary infiltration of the abdominal wall and the hollow viscera, with a consequent contamination of the surgical site.

We report the case of a 40-year-old woman affected by sporadic AF, interesting both the abdominal wall and the colon. The reported case, in addition, occurred as a surgical emergency, for the condition of effective colonic stricture. The aim of this article is not only to present an utterly unique case of sporadic IAF in an emergency setting, but especially to discuss solutions to a challenging surgical problem: how to reconstruct a large parietal gap in contaminated surgery.

**Case Report**

L.B., a 40 years old caucasian woman, nulliparous, was admitted to the emergency room complaining of abdominal pain, vomiting and constipation, poor appetite and weight loss over the last month. Despite the weight loss, her general conditions were not serious and her vital signs were within normal limits. On clinical examina-
tion, there was a palpable mass in the umbilical region, of hard-fibrous consistency, integral with the anterior abdominal wall. On CT scan (Figs. 1, 2) an abdominal neoplasm was detected, infiltrating both the transverse colon and the abdominal wall, up to subcutaneous tissue through the muscle-aponeurotic layers; within the mass, the colon was stenotic, while distended upstream. Laboratory tests were within the standard, with the exception of serum sodium level (Na 121 mMol/l). Intravenous hydration and peripheral parenteral nutrition were started shortly after admission, and a needle biopsy of the abdominal neoplasm was done. The histological features of the sample, examined on frozen sections with standard staining, were suggestive of aggressive fibromatosis. The symptomatic stenosis of the colon prevented us from making a pancolonoscopy, and the need for a timely surgical operation didn’t allow the wait for genetic screening of FAP. Since no case of polyposis was found in the patient’s family anamnesis, we considered the diagnosis of sporadic fibromatosis as the most likely, despite the visceral involvement, and decided on a surgical approach as first-line therapy. The patient was operated on after about 24 hours from admission. The surgery consisted in removing “en bloc” the portions of the abdominal wall and colon affected by fibromatosis (Fig. 3). The colon was deeply adhering to the wall in two distinct segments (namely transverse and sigma) and dilated upstream, so that a large portion of the colon had to be removed, from the transverse to the sigmoid including. Although quite extended, the colectomy has been carried out without formal lymphadenectomy, given that AF is not a metastasizing tumor; the cut edges were a few inches away from the tumor margins. Visceral synthesis was done by lateral-lateral anastomosis, as we usually do when the colon is dilated and not previously emptied, with the aim of a lower contamination of the surgical site, as well as because of the difference in size. The cut edges of the abdominal wall (they also several centimeters from the visible tumor margins) were checked for neoplastic infiltration by extemporaneous histological examination on frozen sections, that confirmed the resection being R0. The resection of the abdominal wall left a gap that didn’t allow a direct suture, making it necessary to interpose a prosthetic device. For this purpose, a prosthetic mesh (Permacol®) of acellular porcine dermis collagen (APDC) has been chosen. The mesh was first placed in-lay with a few detached Prolene stitches, passing full-thickness on the muscle-aponeurotic layers, about 3 cm from their edge, and then sewed to the wall by two parallel suture lines of the same material (Figs. 5, 6). An adequate overlap of the muscular wall to the mesh was thus obtained, in conditions of reduced mechanical tension. Subcutis and cutis were respectively sutured in Vicryl and Nylon, over two Jackson-Pratt drains. The “en bloc” resection did not leave macroscopic nor microscopic residual disease (R0).

Histopathological examination confirmed that it was a desmoid tumor, size of 8x5x3 cm; it had extensive intrabdominal invasion and infiltrated the rectus abdominis, the subcutaneous adipose tissue, the parietal peritoneum and finally the colon. The distance between tumour edges and the margins of resection was not less...
than one centimeter, even in the closest point. No adenomatous polyps were found in the resected colon. The histological slides were stained with hematoxylin and eosin and standard diagnostic immunohistochemistry, both performed on formalin-fixed, paraffin-embedded tissue sections. Two representative samples were stained by antibody specific for smooth muscle actin (1A4; cell marque), CD34 (PGM-1; DBS; dilution 1:50), muscle-specific actin (HHF35; cell marque), CD34 (QBEnd/10; Ventana), CD117 (c-kit YR145; cell marque) and desmin (D33; cell marque). The pathologic tissue consisted of myofibroblastic proliferation with infiltrative pattern of growth, composed of wavy spindle cells arranged in irregular bundles with admixed mild collagen deposition. These cells showed plump nuclei, scanty mitotic activity and were lacking of nuclear atypia. Such myofibroblastic proliferation was associated with an intense chronic inflammation composed of lymphocytes, plasma cells and foamy histiocytes (CD68 positive cells). The neoplastic cells revealed positive cytoplasmic staining for smooth muscle actin, whereas CD34, CD117 and desmin were negative (Figs. 7-9).

The postoperative course was uneventful, regarding both general and local complications. In particular, the wound healed by first intention and there was no surgical site infection. The patient was discharged on the 10th postoperative day. A pancolonoscopy, an esophageal-gastric-duodenal endoscopy and genetic tests were carried out, with no evidence of FAP. The follow-up is now over one year and, after clinical examination and CT abdominal scan, there is no tumor relapse; the abdominal wall is sound and there isn’t any incisional hernia.

Discussion

This case is very rare and possibly unique, since DT of both the abdominal wall and colon are not reported in the medical literature, as far as it results from the Medline and EMBASE search. It raised several problems about differential diagnosis, therapeutic indications and reconstructive surgical procedures, especially with regard...
to prosthetic implants in contaminated surgery. Although the treatment of AF must be individually tailored 16,7, surgical resection is generally considered the first-line therapy for non FAP-related fibromatosis. There is agreement that surgery for sporadic fibromatosis has low mortality and morbidity, and a low incidence of recurrences 7,16,17, A recent survey 7 on 426 patients reported no mortality, and a recurrence rate of 44.3% after surgical treatment. Univariate analysis identified age less than 37 years, a tumor size of more than 7 cm, extra-abdominal localization and a residual macroscopic tumor, as adverse prognostic factors. Patients with one or none of such factors should have the best prognosis. In the case we report, there was only one adverse prognostic factor (tumor size of 8 cm as maximum diameter), so a low risk of recurrence is predictable, and the surgical approach can be considered the most appropriate, regardless of the circumstances of colonic stricture. Since our surgery department is mainly dedicated to emergencies, several times we had to deal with abdominal wall reconstruction in the course of contaminated or infected surgery, namely after long lasting laparostomy, strangulated incisional hernia with bowel rupture, or removal of an infected synthetic mesh. In such cases, we adopted a cross-linked acellular porcine dermis collagen (APDC) mesh (Permacol®). This type of biological mesh has been principally selected in relation to the following issues: susceptibility to infection, suitability for in-lay position, biomechanical characteristics and half-life. The early colonization by cells and vessels from the host tissues, which occurs in the biological prosthesis, seems to provide a lower susceptibility to infection than synthetic implants 30,31,34,35. We may say that, in the presence of germs, biological meshes behave like the body’s own tissues rather than like a foreign body. Conversely, synthetic non-absorbable meshes increase the incidence of surgical site infection, probably leading to the implant removal 33. Synthetic absorbable implants are less prone to be removed for infection, but generally produce an unsatisfactory repair, which can be considered as a “programmed incisional hernia”, because of their short half-life 34,35. Biological prosthesis are suitable for the in-lay position, also on account of their microporous structure 31. There is good evidence that they are well tolerated in contact with intraperitoneal viscera and probably represent the best choice for in-lay position, in comparison to synthetic microporous meshes 36. With regard to bio-mechanical characteristics, APDC meshes have tensile strength, elasticity and duration sufficient to result in an effective repair 37,38. The cross-linking of collagen fibrils makes them more resistant to the collagenase enzyme and ensures a longer half-life to the fabric, appropriate to the time required for the optimal consolidation of the abdominal wall 30,31,35. Moreover, the presence of elastin fibers gives elasticity to the mesh 36,38.

When the parietal gap doesn’t allow a direct closure, like the present case, it is not possible to achieve a proper tension-free reconstruction, as generally required in reconstructive surgery of the abdominal wall 40. According to our previous experience, we believe that some technical precautions may be useful to reduce the risk of incisional hernia. The first one is that the cut edges of the abdominal wall overlap the mesh for several centimeters, without tension. The second, is to perform two parallel suture lines, so as to better distribute the mechanical stress. The third, is to use non-absorbable sutures, since all absorbable ones have shorter half-life than APDC. The follow-up is now over one year and, till now, there aren’t abdominal wall defects nor tumor recurrence. It is possible to exclude any infection of the surgical site, and to assert that the reconstructed wall is sound. Obviously, a longer period is required for a final assessment on the issues of tumor recurrence and incisional hernia.

Conclusions

Complete surgical removal is considered to be the first line treatment in sporadic AF, whenever feasible: in the reported case, an R0 resection was obtained at the cost of a wide, full thickness parietal gap, whose surgical repair was challenging, mainly because of bacterial contamination. In such a contest, cross-linked APDC devices have proven effective for the definitive prosthetic reconstruction, and suitable for the inlay position. In particular, they enable to repair the abdominal wall at the same time of an emergency contaminated procedure, with a low risk of infection or incisional hernias. We believe it advisable to use some technical precaution, namely a double suture line of non-absorbable stitches with an adequate overlap of the abdominal wall to the mesh. In the reported case, no complications have been observed. After one year follow up, there is no tumour recurrence and the abdominal wall has fully consolidated. The surgical procedure we present is, in our opinion, simple and effective.

Riassunto

INTRODUZIONE: La fibromatosi aggressiva (AF) o tumore desmoide (DT) è una malattia neoplastica non metastatizzante, ma localmente invasiva e tendente alla recidiva. Può essere sporadica o correlata alla poliposi adenomatosa familiare (FAP). Le forme sporadiche sono più rare e, nel loro ambito, la localizzazione addominale non supera il 5% di tali casi; tuttavia, non risultano in letteratura casi di AF, contemporaneamente infiltrando la parete ed i visceri addominali. OBIETTIVO: La finalità di questo articolo è duplice: sia presentare un caso del tutto unico di AF sporadica, infil-
trante sia la parete addominale che il colon, operato in urgenza per occlusione intestinale; si discutere la tecnica chirurgica ed il materiale protesico adottato nella ricostruzione di un’ampia breccia pialet tale in condizioni di contaminazione del sito chirurgico ed in regime d’urgenza.

**Caso clinico:** Riportiamo il caso clinico di una donna di 40 anni, affetta da AF sporadica, infiltrante sia la parete addominale anteriore, sia il colon, giunta in Pronto Soccorso in condizioni di occlusione intestinale. L’intervento chirurgico è consistito nella rimozione “en bloc” delle porzioni di colon e di parete addominale interessate dal processo neoplastico (con esito R0). La resezione ha lasciato un’ampia breccia pialetale, non passibile di sutura dreta. Per la sua contestuale ricostruzione, è stata usata una protesi biologica in collagene di derma suino (APDC) di tipo “cross-linked” (Permacol®), suturata alla parete in posizione “inlay”, con una duplice linea di sutura in materiale non assorbibile. In oltre un anno di follow-up, non si sono verificate complicanze, recidive neoplastiche o ernie incisionali.

**Discussione:** Sono discusse le problematiche inerenti la diagnosi differenziale in condizioni d’urgenza tra fibromatosi sporadica e FAP-correlata, le indicazioni terapeutiche e le procedure chirurgiche, con particolare riguardo alla ricostruzione protesica della parete in chirurgia d’urgenza per occlusione intestinale; sia discutere la tecnica che la parete addominale le procedure chirurgiche, con particolare riguardo alla ricostruzione protesica della parete in chirurgia contaminata ed all’impiego di mesh in APDC di tipo “cross-linked”.

**Conclusioni:** La resezione chirurgica R0 è da considerare la terapia di scelta nelle AF sporadiche. Nel nostro caso, si è ottenuta una resezione R0, a costo di un’ampia breccia pialetale, la cui riparazione protesica appariva problematica a causa della contaminazione batterica del sito chirurgico. In tale condizione, la resezione chirurgica R0 è dimostrata efficace per una ricostruzione protesica definitiva, oltre che idonea alla posizione inlay, consentendo la riparazione della parete contestualmente ad un intervento di tipo contaminato ed in urgenza. Alcuni accorgimenti tecnicì (come suturare la protesi con una duplice linea parallela di materiale non assorbibile e con un’adeguata sovrapposizione dello strato muscolo-aponeurotico alla protesi) ci sembrano consigliabili. Nel presente caso, non si sono osservate complicanze, recidive neoplastiche, né laparocelie in oltre un anno di follow-up. La procedura chirurgica ricostruttiva adottata è, a nostro avviso, semplice ed efficace.

**References**


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