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A case report

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Intra-abdominal desmoid tumors. A case report

INTRODUCTION: *Desmoid tumors are defined as a rare fibroblastic proliferative disease and may occur in any musculoaponeurotic or fascial tissue .*
CASE REPORT: *A 54-year-old man with FAP referred to our institution presenting fever and abdominal pain. Four years before a prophylactic colectomy with ileal-rectal anastomosis was done. Three years later a wide intraabdominal mass with desmoid characteristics was detected. In spite of therapy with tamoxifene the RM control showed an increase of mass dimension. Few days later the patient arrived in our istitute with fever and abdominal pain. Medical therapy didn't improve clinical symptoms and the patient underwent surgery.*
DISCUSSION: *Desmoid tumors are benign tumors, but not a benign disease. Most of these tumors occur sporadically, but about 5% arise in association with familial adenomatous polyposis. Intra-abdominal desmoid tumors remain asymptomatic until their growth and infiltration causes visceral compression. They can cause serious complications and treatment is often unsuccessful. Aggressive fibromatosis treatment should be evaluated and managed by a multidisciplinary team with expertise and experience in sarcoma, prior to initiation of therapy. In FAP, surgery has been recommended for abdominal wall tumors but there are only few indications for intra-abdominal disease like pain, bowel obstruction and desmoids perforation.*

KEY WORDS: Desmoid tumor, FAP, Intra-abdominal fibromatosis.fistulation to bowel, small

Introduction

Desmoid tumors are defined as a rare fibroblastic proliferative disease and may occur in any musculoaponeurotic or fascial tissue. Although they are considered benign, they could be frequently locally aggressive with a high rate of recurrence but with no tendency to metastasize [1]. Most of these tumors occur sporadically, but about 5% arise in association with familial adenomatous polyposis (FAP)[2] .Aggressive fibromatosis treatment should be evaluated and managed by a multidisciplinary team [3] because it is still under debate. The authors report a case of a 54-year-old male with a large desmoid lesion involving the jejunal and ileal mesentery who arrived with fever and abdominal pain.

Case report

A 54-year-old man with FAP referred to our institution presenting fever and abdominal pain. FAP was diagnosed when the patient was 50 years old and his father was

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affected too. Clinical examination presented supernumerary teeth, frontal, cranial and mandibular osteomas. After two months from the diagnosis a prophylactic colectomy with ileal-rectal anastomosis was done and he was later managed with close colonoscopic surveillance. After two years a radical resection of a desmoid tumor of the right rectus abdominis muscle was performed down to the peritoneum. One year later a gastroduodenoscopy showed multiple polyps in the duodenum which were removed and a RM reported a wide intraabdominal mass with desmoid characteristics. RM indicated double irregular fibrotic lesions ,of about 68x51mm and 35x31 mm, in the jejunal mesentery and other two smaller in the ileal mesentery. The patients started therapy with tamoxifene. Three months later the RM control showed an increase of mass dimension. Few days later the patient arrived in our istitute with fever and abdominal pain. A TC scan (Fig. 1) showed a mass of 14x9 cm with colliquation areas in the jejunal mesentery and nearly another mass of 5x6 cm. Additional smaller localizations were described in ileal mesentery and in perisigmoid tissue. Medical therapy didn't improve clinical symptoms and the patient underwent surgery. By median laparotomy and with difficulties in abdominal access, a wide mass was found (longer diameter about 14 cm). The mass infiltrated jejunal loops hindering intestinal transit. Final decision was for a partial jejunal resection and intestinal continuity was estabilished with a double layer suture. Drainage of the colliquation areas was done. Unfortunately the complete mass excision proved impossible due to infiltration in mesenteric vessels. Surgery was performed just for decompression due to the tumor mass. There were no intra or perioperative complications. The patients was admitted to soft diet on postoperative day 4 and discharge on 10 p.o. day. Pathology report confirmed a desmoid tumor. After demission the patient started chemiotherapy with cytotoxic agents.

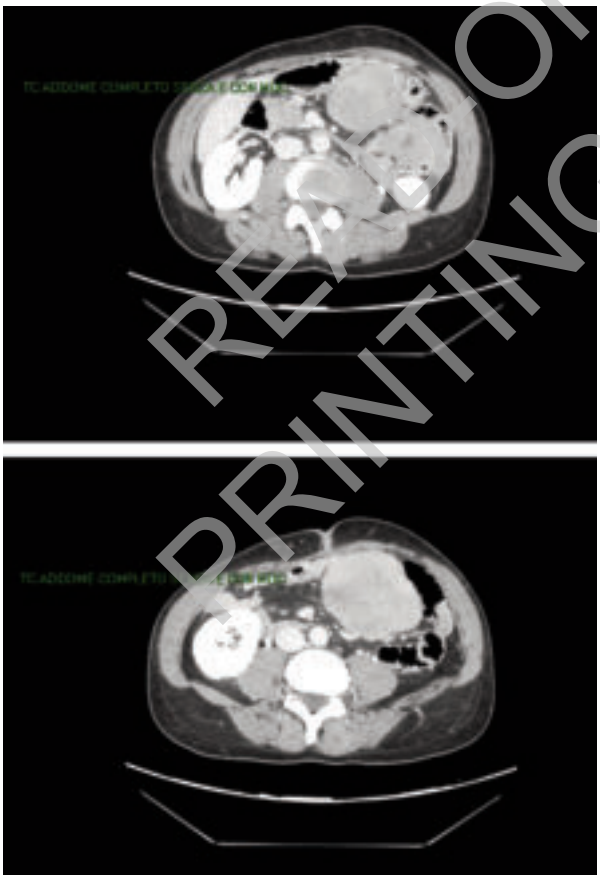

Fig. 1: Preoperative TC scan.

Discussion

Desmoid tumors, or aggressive fibromatosis, are a subtypes of soft tissue sarcomas. They are benign tumors, but not a benign disease [1]. Desmoid tumor arises from myofibroblast, lacks a true capsule, and usually infiltrates into adjacent muscle bundles. The telomerase length and activity is normal, nuclei are small and regular, and mitoses are infrequent - all of which support its histologically benign nature [4].

Although desmoids have similar bland microscopic appearance, their clinical behavior varies with wide local infiltration and with a marked propensity for recurrence after conservative resection. Their agressivity should never be underestimated. Desmoid tumors are reported to be the second most common cause of death in patients with FAP, after colonrectal carcinoma [5].

They can be classified as extra-abdominal and abdominal disease. This second type can be further sub-classified in superficial (or abdominal wall) disease, the commonest location, and intra-abdominal disease [6]. The intra-abdominal fibromatosisis can be divided in mesenteric, pelvic and retroperitoneal according to the site of origin. Small bowel mesentery is the most common site of origin of mesenteric fibromatosis [6,7]. Desmoid tumors localized in the abdomen have a worse prognosis, as they may cause intestinal obstruction, ureter obstruction with hydronefrosis and may also invade major vessels [8].

Most of these tumors occur sporadically, but about 5% arise in association with familial adenomatous polyposis[2]. In FAP patients, the development of desmoids tumors is associated with the position of the APC (Adenomatous Polyposis Coli) germline mutations (mutations 3' of codon 1440), previous abdominal surgery, being female and a family history of desmoids tumors [6,9]. This genetic disease could be associated with other extracolonic manifestations like osteomas, gastric and duodenal polyps. Desmoid tumors would be responsible for increasing morbidity and mortality rates in patients affected by FAP. In fact intra-abdominal desmoid tumors remain asymptomatic until their growth and infiltration causes visceral compression. Intra-abdominal desmoids can cause serious complications and treatment is often unsuccessful [10].

Aggressive fibromatosis treatment should be evaluated and managed by a multidisciplinary team with expertise and experience in sarcoma, prior to initiation of therapy [3]. A variety of systemic agents - including anti-estrogens (like tamoxifen), non-steroidal anti-inflammatory agents (such as sundilac), interferon-alpha and cytotoxic chemotherapeutic agents - have been usually considered in pharmacological treatment of abdominal desmoids tumors [3,6]. However, the numbers of patients treated with these agents are small and there are no randomized, controlled trials [6] because of difficulty of performing them in such a rare disease.

Generally radiation therapy is only recommended for desmoids tumors that are in the extremity, not for the retroperitoneal/intrabdominal ones [3].

The first approach in asymptomatic tumors could be observation and other treatment pathways could be considered in case of a progression of the disease [3]. In FAP, surgery has been recommended for abdominal wall tumors but there are only few indications for intra-abdominal disease like pain, fistulation to bowel, small bowel obstruction and desmoids perforation [2]. This because the risk of recurrence is quite high and a complete resection may be difficult or even impossible [2,10].

In the reported case surgery was a necessary decision in the emergency situation because of abdominal pains and it proved to be a simple palliative.

Riassunto

INTRODUZIONE: I tumori desmoidi sono rare malattie fibroblastiche proliferative e nonostante siano considerati come neoplasie benigne, sono localmente aggressivi con facile tendenza alla recidiva. Circa un 5% di questi tumori si presenta in associazione alla FAP (poliposi adenomatosa familiare). Gli autori riportano il caso di un uomo di 54 anni affetto da FAP arrivato con febbre e dolore addominale causati da un'importante lesione desmoide infiltrante il mesentere digiuno-ileale.

CASO CLINICO: Un uomo di 54 anni con diagnosi di FAP giungeva al nostro istituto per febbre e dolore addominale. Il paziente era stato precedentemente sottoposto, in altro istituto, a colectomia profilattica con ileo-retto anastomosi e a resezione di tumore desmoide interessante il muscolo retto destro dell'addome. Pochi mesi prima del suo arrivo nel nostro reparto era stata posta diagnosi radiologica di tumore desmoide intraddominale. Veniva iniziata terapia con tamoxifene, ma pochi giorni dopo l'ultimo controllo, che mostrava un incremento di dimensione della massa, il paziente si recava al Pronto Soccorso per comparsa di febbre e dolore addominale. Durante la degenza si eseguiva esame TC che mostrava infiltrazione del mesentere digiuno-ileale con aree di colliquazione. Non riscontrando miglioramenti con la terapia medica si decideva per l'intervento chirurgico d'urgenza. All'ingresso in addome si evidenziava infiltrazione delle anse digiunali con ostacolo al transito intestinale. Non si rendeva possibile la completa escissione della neoplasia e si procedeva pertanto ad una resezione parziale del digiuno e a drenaggio delle aree colliquate.

DISCUSSIONE: I tumori desmoidi, nonostante siano considerati neoplasie benigne, hanno un comportamento altamente aggressivo con infiltrazione locale e facile tendenza alla recidiva dopo resezione. Vengono classificati in base alla loro localizzazione in extra- ed intra-addominali; questi ultimi hanno la prognosi peggiore. Circa il 5% di questi tumori si presenta associato alla FAP e rappresentano la seconda causa di morte, dopo i tumori colon-rettali, di questa malattia. Il trattamento di queste neoplasie è ancora in discussione, in quanto in letteratura non ci sono studi clinici randomizzati sull'argomento. Nella FAP le indicazioni per la chirurgia delle lesioni intra-addominali sono riservate a condizioni d'urgenza come dolore addominale, occlusione intestinale o perforazione; questo per l'alto rischio di recidiva legato al fatto che una completa resezione non è quasi mai possibile

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