1130-0108/2009/101/11/813-819 Revista Española de Enfermedades Digestivas Copyright © 2009 Arán Ediciones, S. L.

Rev Esp Enferm Dig (Madrid) Vol. 101, N.° 11, pp. 813-819, 2009

Letters to the Editor

Desmoid tumor arising in a laparoscopic trocar site after cholectomy

Key words: Desmoid tumor. Familial adenomatous polyposis. Port site.

Dear Editor,

Desmoid tumors are solid masses of the fascia or of musculoaponeurotic tissue showing proliferation of well-differentiated fibroblasts or miofibroblasts. They are rare in the general population but their incidence ranges between 11-20% in case of patients with familial adenomatous polyposis (FAP) (1,2). Here we report the case of a female patient with FAP who developed a desmoid tumor in a trocar site after total laparoscopic colectomy. So far, this is the 2nd case reported in the literature of desmoid tumor arisen in a laparoscopic trocar site (3).

Case report

Patient is a woman who was diagnosed of attenuated familial adenomatous polyposis. Laparoscopic total colectomy was performed when she was 34 years old. Twenty moths later she began to feel abdominal mass. We observed a mass in the anterolateral abdominal wall located at the right laparoscopic trocar site. TC and MR confirmed the existence of an abdominal mass compatible with a desmoid tumor. In order to reduce the size of the tumor before surgery, patient was treated with indometacine 75 mg/24 h, tamoxifene 10 mg/12 h for two months. After this time a new abdominal MR revealed the growth of the tumor and consequently the lack of response to therapy (Fig. 1). The tumor and the anterolateral abdominal

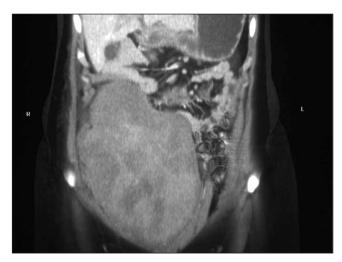


Fig. 1. Abdominal MR. Tumor of 22 x 14 cm occupying the entire right anterolateral abdominal wall.

wall were resected with negative margins more than 1 cm. We also removed 40 cm of the distal jejunum with and we performed manual end-to-end anastomosis as we found a new desmoid tumor of 6 x 4 cm in the mesojejunum whith intestinal serosa affected. The abdominal wall was reconstructed using a Proceed® prostheses of 20 x 30 cm.

Discussion

Desmoid tumors are benign mesenchymatous neoplasms which are locally aggressive and have limited tendency to metastasize. They are more frequent among female patients 2:1 (1,4). The etiology of this type of tumors is not well established. Hereditary and genetic factors are involved such as the APC gene or beta-catenine mutation in patients with FAP and which would determine their predisposition to suffer from these tumors (4). Hormonal alterations also affect the etiopathogenia LETTERS TO THE EDITOR

of these tumors as they can express estrogen and progesterone receptors (4).

The incidence of desmoid tumors is very low in general population (2-4 cases per million inhabitants) and account for 0.03% of all tumors (6). Their most common location is extraabdominal and mainly affects the lower limbs, thorax, head and neck.

A second group, where we will include our patient, comprises patients with FAP, 11-20% of whom develop desmoid tumors. This is the most common extraintestinal manifestation of the disease and the second cause of mortality following colorectal cancer. As regards tumor location in this group, 50% are intrabdominal (mesentery and small intestine) and around 30% are located in the abdominal wall and the mesentery, as was the case of our patient. In the case here reported, the tumor appeared in the right laparoscopic trocar site 15-20 months after surgery. According to the review of the literature, this is the second case of a desmoid tumor arising in a trocar scar (3).

Desmoid tumors are normally silent from a clinical point of view but symptoms appear when their growth causes compression or less frequently infiltration of adjacent structures. The small intestine and ureters are frequently affected. The finding of an abdominal mass is usually the first and only symptom as was the case of our patient (4,5).

Diagnosis is based on imaging test. CT images, most tumors are hyperintense to muscle and show well-defined margins. In MR T1 images the tumor has low signal intensity to muscle and T2 images show variable or intermediate intensity signals (5,6).

In our case, imaging tests were useful to confirm the diagnosis of desmoid tumor in abdominal wall as well as to analyse its size and affectation of adjacent structures, but they did not detect the tumor in the mesojejunum.

Recommended treatment is surgical resection with negative margins. Mesenteric location, tumor size and the involvement of resection margins are considered the most relevant risk factors for recurrence which, generally and even in the case of a R0 resection, occurs in 25-65% of the cases (4). Eighty per cent of these recurrences occur in the two years after surgery. Adjuvant therapies have been described to treat desmoid tumors, but there are not meta-analyses or randomized prospective studies proving their efficacy. Hormonal therapy with tamoxifene or NSAIDs has proved effective occasionally. Radiation doses between 40-60 Gy has been reported by several authors as an effective treatment and low toxicity (1,6).

G. Suárez-Artacho, R. Jiménez-Rodríguez¹, J. M. Díaz-Pavón¹, J. Sánchez-Gil¹ and J. Vázquez-Monchul¹

Unit of Hepatobiliopancreatic Surgery and Trasplantation. 'Unit of Coloproctology. University Hospital Virgen del Rocío. Sevilla, Spain

References

- Oguz M, Bedirli A, Gultekin A, Dursun A, Mentes BB. Desmoid tumor arising at the colostomy site after abdominoperineal resection for rectal carcinoma: report a case. Dis Colon Rectum 2006; 49(9): 1445-8
- Soravia C, Berk T, Mcleod RS, Cohen Z. Desmoid disease in patients with familial adenomatous polyposis. Disease Colon Rectum 2000; 43(3): 363-9.
- 3. David B, Kaplan MD, Edward A, Levine MD. Desmoid tumor arising in a laparoscopic trocar site. The American Surgeon; 1998. p. 64-5.
- Ferenc T, Sygut J, Kopczynsky J, Mayer M, Latos A, Dziki A, et al. Agressive fibromatosis (desmoid tumor): definition, ocurrence, pathology, diagnostic problems, clinical behavior, genetic background. Pol J Pathol 2006; 57(1): 5-15.
- Dequanter D, Gebhart M. Desmoids tumors. J Chir (Paris) 2002; 139(4): 236-9.
- 6. Privette A, Fenton SJ, Mone MC, Kennedy AM, Nelson EW. Desmoid tumor: a case of mistaken identity. Breast J 2005; 11(1): 60-