Occurrence of Desmoid Tumors following Surgery for Familial Adenomatosis Coli

—Report of Two Cases—

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Summary: Twenty-five patients with adenomatosis coli were subjected to operations, and two in which desmoid tumors appeared after the operation on familial adenomatosis coli are described here. These patients were 26 and 37 year-old males. After the total colorectomy, each tumor was associated with soreness and was palpated in the lower abdominal wall, and operations were performed. The histological findings from the resected specimens, 12 cm and 34 cm in size, indicated desmoid tumors. Poliposis of the large intestine with desmoid tumors, as a complication, has been reported in 23 previous cases in Japan.

Key words: desmoid tumor—familial adenomatosis coli—familial polyposis—gardner's syndrome—diagnosis—treatment

Introduction

It is known that osteomas, desmoid tumors and brain tumors complicate adenomatosis coli, as parenteral concomitant lesions. In addition, desmoid tumors occasionally develop and repeatedly recur in the mesentery and in abdominal surgical wounds after surgery.

Two patients developed desmoid tumors out of 25 (8%) that had undergone surgery for poliposis of the large intestine. In this paper the two cases are described along with some discussion of the pertinent available literature.

Case 1

Patient: 26 year-old male

Chief complaint: Lower right abdominal tumor.

Familial history: No abnormalities are noted.


Present illness: There was a palpable 50 mm tumor in the lower right abdomen in October, 1988. This rapidly increased in size to about 120 mm in February, 1990: and the patient was admitted to the hospital because of an accompanying spontaneous pain.

Clinical conditions upon entering the hospital: The patient was of medium physical constitution and well nourished. From the abdominal findings, the previous surgical wound was noted in the
median, and a fist-sized lump was palpated in the lower right abdomen (Fig. 1). No abnormalities were noted in the general and biochemical blood examinations.

Echographic findings: A solid tumor with distinct borders was circumscribed in the abdominal wall and a heterogeneous pattern was presented internally (Fig. 2).

Computed tomography scan: Presented a heterogeneous low density mass with a distinct border in the lower abdomen. As compared to the June, 1989 CT scan, this mass had increased markedly and was forcing the straight muscle of the abdomen dorsally (Fig. 3).

Pelvic artery angiography: The main feeder was the superficial circumflex iliac artery, and a distended blood vessel was observed in the periphery: but new angiogenesis was not observed and this was not considered to be a suspicious finding in view of the malignancy.

Based on the above examination findings: this mass suspected to be a desmoid tumor combined with a familial adenomatosis coli, and surgery was performed on April 10, 1990.

Surgical findings: The tumor had infiltrated the straight muscles of the abdomen, the internal and external obliques, and the peritoneum, and a combined resection was performed.

Resected specimen findings: The tumor was 120 × 100 × 70 mm in size, weighed

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Fig. 1. From the abdominal findings, the previous surgical wound was noted in the median, and a fist-sized lump was palpated in the lower right abdomen.

Fig. 2. An echograph showing a solid tumor with distinct borders circumscribed by the abdominal wall. A heterogeneous pattern was presented internally.

Fig. 3. (a): A 1990 computed tomography scan of the abdomen showing a heterogeneous low density mass with a distinct border, and (b): A June, 1989 CT scan with a large mass that forced the straight muscle of abdomen, dorsally.
Fig. 4. The tumor was 120 × 100 × 70 mm in size, weighed 370 g, was elastic hard, and had a grayish-white multinodular surface. The cut surface exhibited a uniform structure that is probably rich in a grayish-white fibrous component.

370 g, was hard, and had a grayish-white multinodular surface. The cut surface exhibited a uniform structure that was probably rich in a grayish-white fibrous component (Fig. 4).

Pathological findings: Histologically, long spindle shaped cells that appeared fascicular, irregular, and interlacing were present and an increase in fibrous connective tissue was noted (Fig. 5).

Postoperative course: The patient was discharged on May 10, 1990, and no remarkable changes have been observed in the abdominal findings for six months, postoperatively.

Case 2

Patient: 37 year-old male

Chief complaint: Lower abdominal tumor.

Familial history: Father's brother has undergone total colorectectomy for adenomatosis coli.

Patient history: Underwent a total colorectectomy on September 12, 1989, because of a diagnosed familial adenomatosis coli complicated by colorectal cancer.

Present illness: A palpable tumor of about 30 mm was noted in the lower abdomen in January, 1990, and it has increased in size. In addition, the patient was admitted to the hospital because of pain since the first ten days of March.

Clinical conditions upon entering the hospital: the patient had a medium physical constitution and was well nourished. In the abdominal findings, the previous surgical wound was noted in the median, and a 40 mm, elastic hard, poorly mobile
A lump was palpated in the lower abdomen. No abnormalities were noted in the clinical laboratory data.

Echographic findings: Because a solid lump with indistinct borders was circumscribed in the lower abdominal wall, a low echo pattern was presented internally (Fig. 6).

Computed tomography scan: A low density area with a distinct border was noted in the median of the lower abdominal wall, and a marked enhancement effect was presented (Fig. 7).

Based on the above examination findings: the lump was suspected to be a desmoid tumor combined with familial adenomatosis coli, and surgery was performed on April 3, 1990.

Surgical findings: The tumor was localized within the abdominal wall and had infiltrated the lower abdominal straight muscle, fatty tissues, and peritoneum, and was resected as a single mass. In addition, an elastic hard $20 \times 10$ mm tumor was noted in the mesoileum about 150 cm from the Tritz ligament, and was resected.

Resected specimen findings: The tumors were $34 \times 28$ mm and $20 \times 10$ mm, and each was elastic hard and grayish-white. The surface was nodular, and the cut surface had a uniform structure believed to be rich in a grayish-white fibrous component.

Pathological findings: Histologically, hyperplasia of the fibrous connective tissue was noted.

Postoperative course: The patient was discharged on May 1, 1990, and no remarkable changes have been observed in the abdominal findings for six months, postoperatively.

Discussion

Desmoid tumors are known to be a concomitant disease of adenomatosis coli, but among 201 cases of adenomatosis coli at the Mayo Clinic, seven cases (3.5%) complicated by desmoid tumors have been reported, and at St. Mark’s Hospital, of 141 poliposis patients, eight cases (5.7%) have been reported. In Japan, 23 cases of desmoid tumors have been reported to have developed after surgery for adenomatous poliposis of the large intestine. They usually develop within two years after surgery for this disease, and these recent cases also developed nine to sixteen months, postoperatively.
DESMOID TUMOR

The region of development, according to Kinoshita et al. (1984) is primary in three locations, outside the abdominal wall, in the abdominal wall, and within the intraperitoneal cavity. Among these, the abdominal wall is reported as the most frequent location, where 11 out of 15 cases that developed in the area of the surgical scar, 1-3 years postoperatively.

Furthermore, as Smith (1966) pointed out, many desmoid tumors develop in the abdominal wall scar after poliposis surgery, this was also true for the cases we encountered. On the other hand, most of those that developed in the intraperitoneal cavity were in the mesentery, and because adenomatous poliposis was complicated by colorectal cancer and the development of desmoid tumors.

More basic studies on the therapeutic approach to desmoid tumors need to be conducted. Although they are composed of benign cells histologically, they can diffuse and become invasive. Any tumor cells remaining will almost certainly lead to a recurrence. Consequently, tamoxifen, cyclic AMP antagonists, non-steroidal anti-inflammatory drugs and radiation therapy have been used as maintenance therapy, but the results were not always satisfactory.

On the other hand, even when exirpation has been carried out, the postoperative recurrence rate reaches 50-62%. Thus, wide resections, involving intact tissues have been performed, and the repair of abdominal wall defects using Gore-tex sheets and Marlex mesh have been reported. In addition, Khorsand et al. (1980) reported cases in which recurrences were not observed for five years after the combined use of resection and radiation therapy. What type of therapy would be the best, when the masses are growing very rapidly? Bussey et al. (1975) reported that desmoid tumors with adenomatosis coli do not grow at a uniform rate. There are those that grow rapidly, those that stop growing after a certain size, and those that are unchanged after 15 years. However, there is a report of a desmoid which developed in the mesentery after a total colorectomy and led to the complication of hydronephritis due to a compression of the ureter from the growth pattern. It became unresectable. Also there is a report of a desmoid tumor that grew to 230 × 220 mm and caused a pressure necrosis of the duodenum. On the other hand, there are also occasional report in which spontaneous healing has been observed. Caldwell (1976) described a case in which a desmoid tumor, that developed after a normal delivery, regressed after it had grown to 170 × 150 mm. Even so, the best treatment is a complete resection while the tumor is small.

References