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Development of Desmoid and Mesenteric Fibromas following Total Colectomy for Adenomatous Polyposis Coli in Gardner's syndrome

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ABSTRACT

We describe a case of polyposis coli, which was followed by development of desmoid in the rectus abdominis muscle and fibromas in the mesentery during an interval of two years. This case supports the hypothesis that, in Garder's syndrome, the traumatic injury by colectomy triggers an unusual fibrous proliferation in the peritoneal cavity and incision site under the possible genetic basis.

Key Words: Gardner's syndrome, Mesenteric fibromas, Desmoid, Polyposis coli

INTRODUCTION

Gardner's syndrome, as initially described by Gardner in the early 1950, is a complex disorder with multiple colorectal polyposis associated with various soft and hard tissue tumors including osteomas, fibromas and epidermoid cyst[1]. In particular, the former three lesions are the most common ones and have been recognized as the triad of Gardner's syndrome. Fibrous tumor, in Gardna's syndrome including desmoid & mesenteric fibromatosis, however, should be considered as an impressive component in aspect to not only their association with trauma but also their management. There is a single case report of Gardner's syndrome in Korean literature[2].

We add a case of desmoid in the rectus abdominis and fibromas in the mesentery following colectomy for adenomatous polyposis coli in a Gardner's syndrome patient.

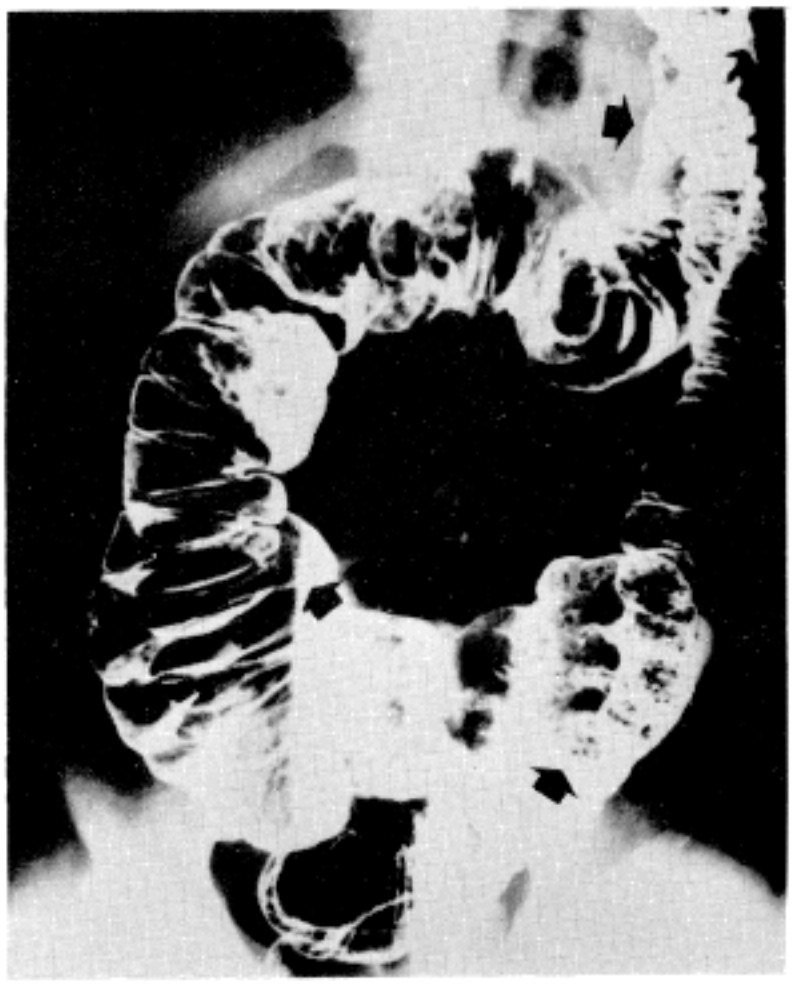

Fig. 1. Barium enema showing numerous filling defects (arrow) on the entire colon.

*This paper was presented at the annual meeting of Korea Society of Pathologists in October 21, 1988.

CASE REPORT

A 31-year-old man was admitted to the Seoul National University Hospital in July, 1986 because of hematochezia and chronic diarrhea for about 5 years. At that time colonoscopy and barium study demonstrated multiple polyposis involving the entire

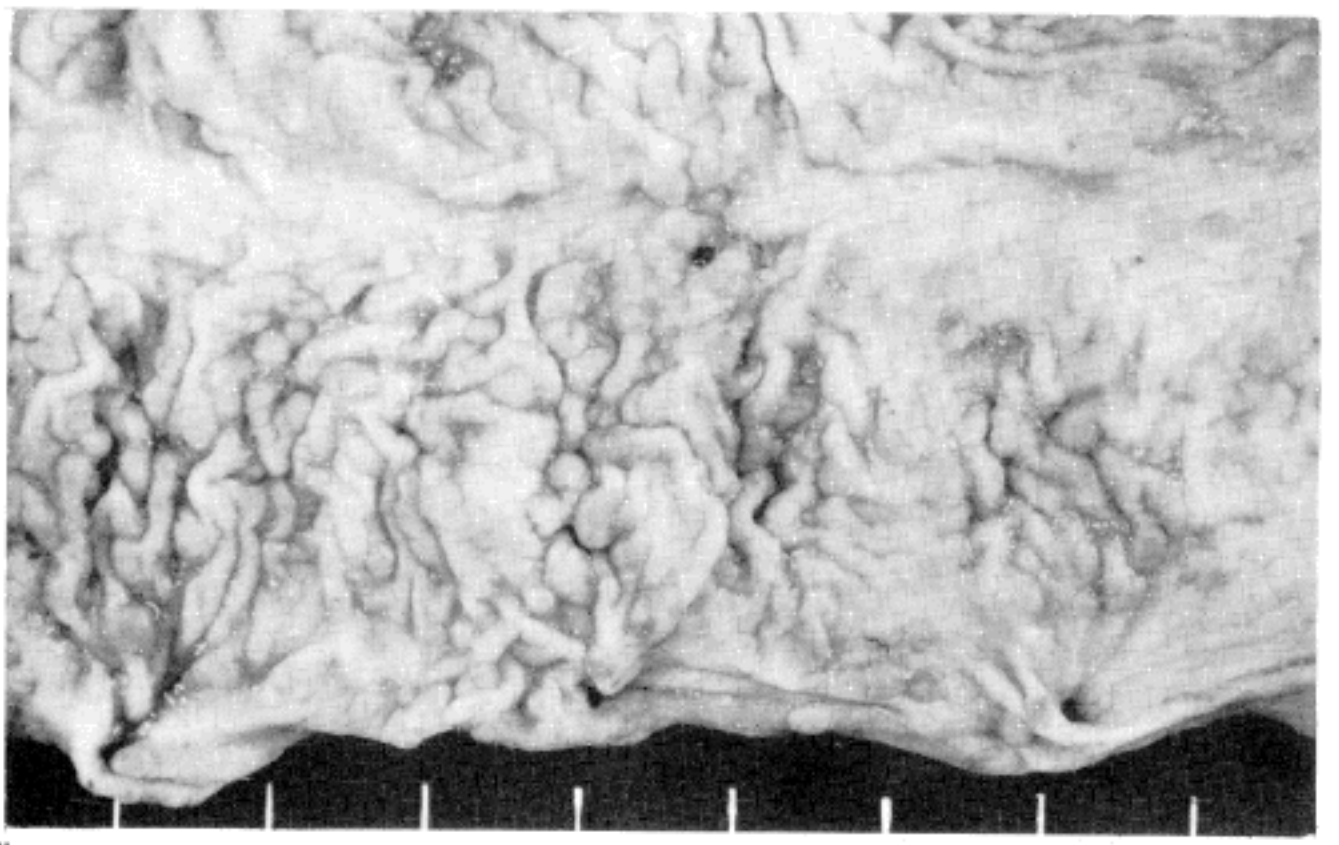

Fig. 2. There are numerous polyps in variable size, measuring 0.2 cm to 2,5 cm, scattered on the entire colon.

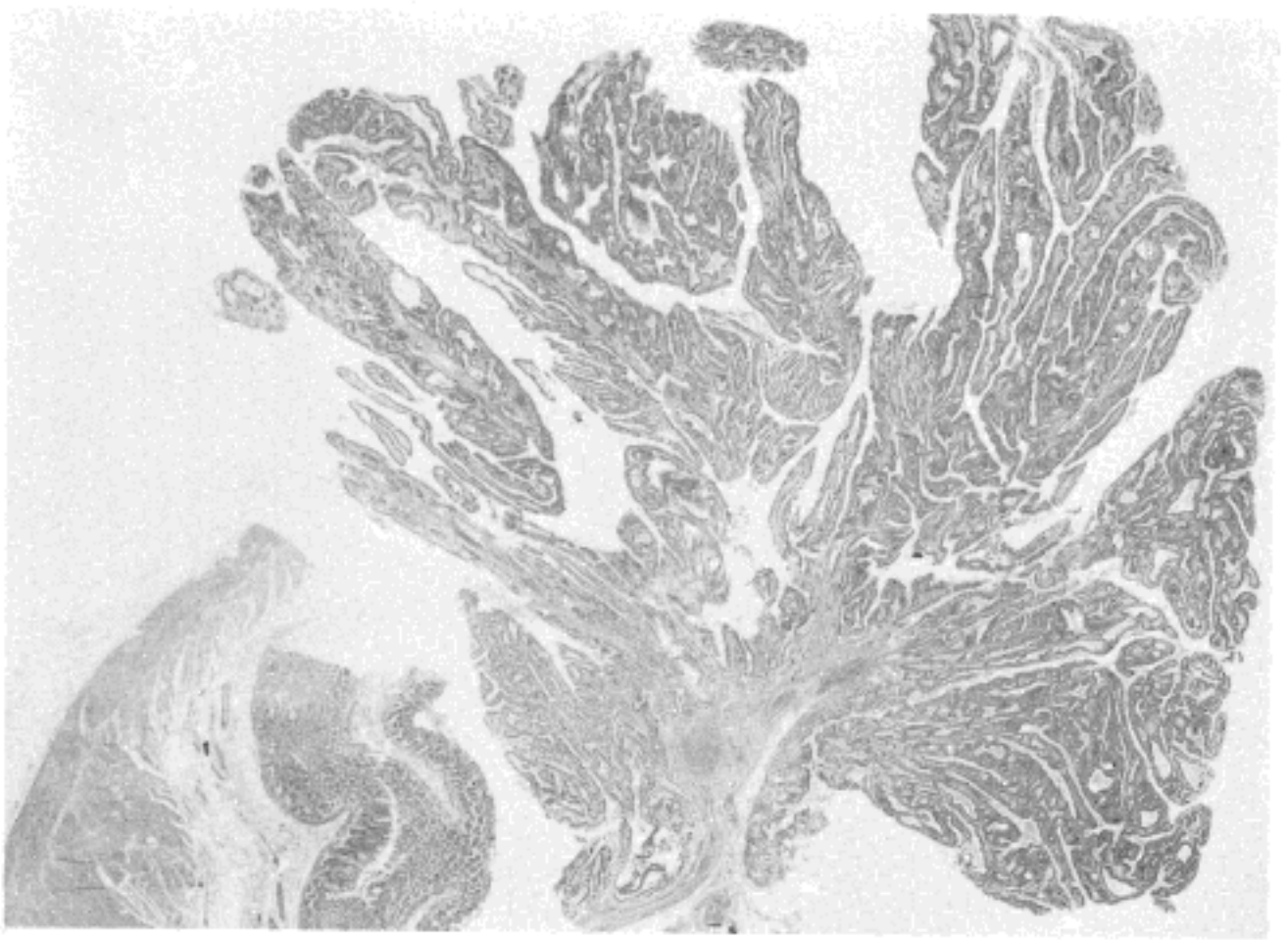

Fig. 3. Photomicrograph of the colonic polyp featuring villo-tubular adenoma (H-E, ×1).

colon (Fig. 1). There was no family history of colonic polyposis, although pedigric information was incomplete. He had received an excision of lipoma in the neck 20 years ago. He had another 3 cm-sized mass on the posterior neck which was not cofirmed by biopsy. Under the preoperative clinical diagnosis of familial polyposis coli, a panproctocolectomy with ileostomy was performed. Grossly, numerous sessile or pedunculated polyps of varying size, ranging from 0.2 cm to 2.5 cm, were scattered on the entire colon, rectum and the terminal ileum (Fig. 2). Histologically, polyps were villotubular adenomas and there was no carcinomatous change (Fig. 3).

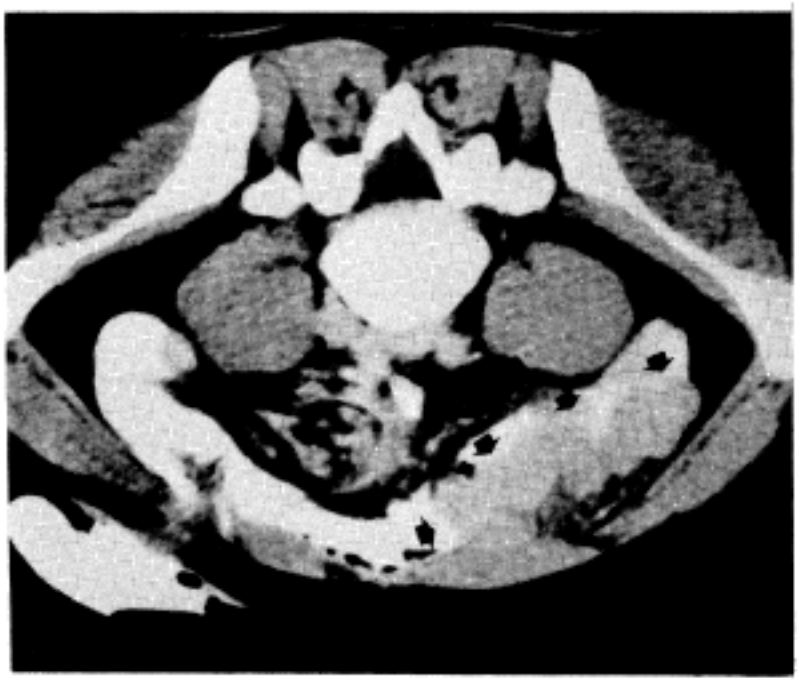

Fig. 4. Three ill-demarcated masses (arrows) overlying the ileum and the abdominal wall are seen on abdominal CT.

On April 1988, he was readmitted because of abdominal mass (Fig. 4). On operation there were three well circumscribed masses, one in the rectus abdominis muscle and two in the mesentery overlying the ileum. Removal of the tumors along with a segmental resection of ileum was performed. The resected masses measured 7×6×5 cm, 5×5×5 cm and 4×4×4 cm, respectively. They were all firm and solid, and showed whitish tan, coarsely trabeculated cut surfaces. The intestinal mucosa of the segmentally resected ileum showed no polypoid lesions.

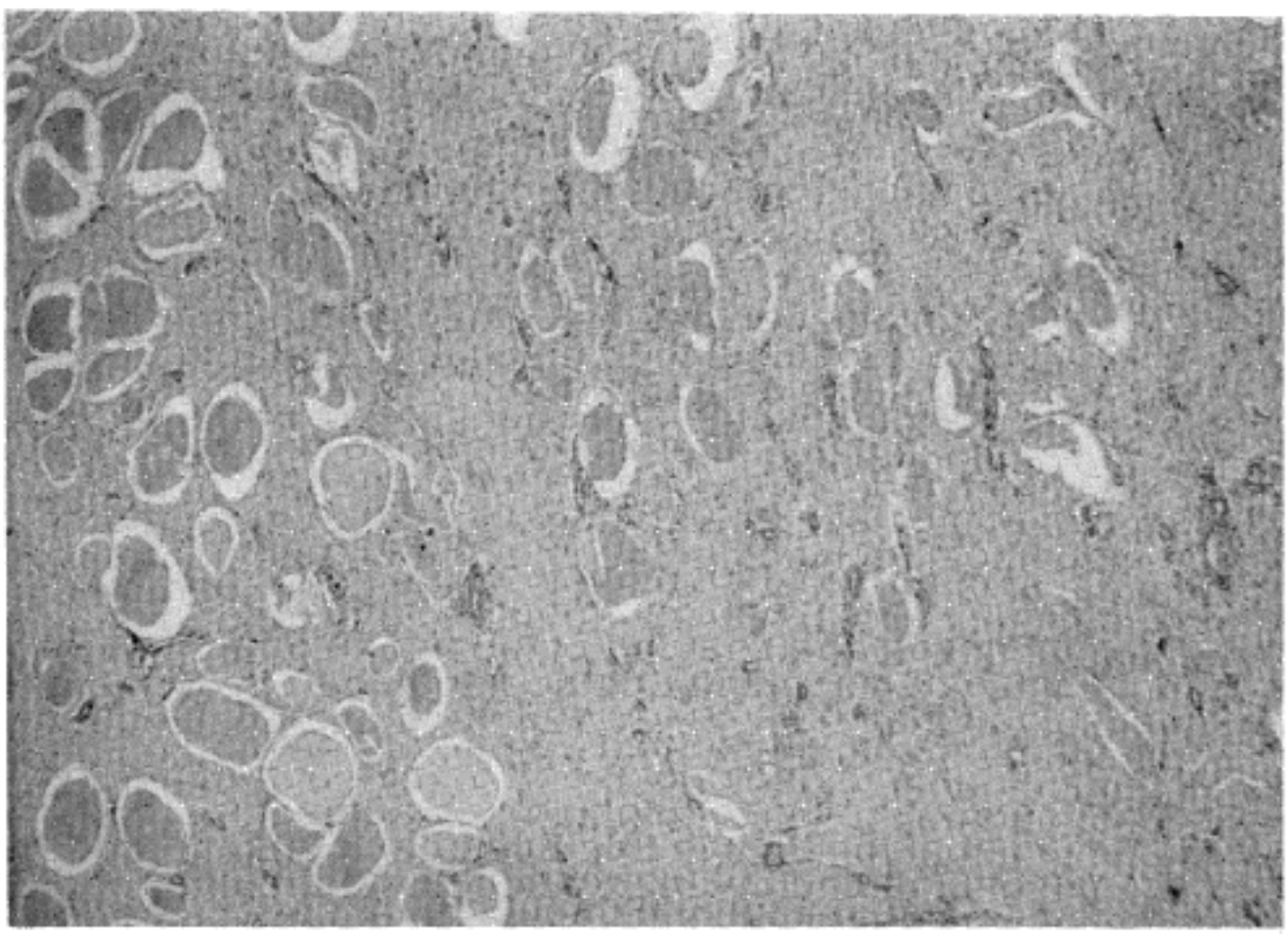

Fig. 5. The desmoid of the rectus abdominis. The fibrous tissue extends between fascicles, eventually isolating groups of muscle fibers (H-E, ×40).

Microscopically, they were composed of mature fibroblasts and densely packed collagen laydown in fasciculating arrangement. At periphery of the mass in the rectus abdominis, the proliferating fibrous tissue extended between muscle fascicles, eventually isolating and engulfing groups of muscle fibers (Fig. 5). But in the mesenteric lesion, there was no extension into the ileal wall. Neither mitotic figures nor tumor necrosis was found. During continuous follow up about 8 months after the second operation, he remains free of recurrent masses or subjective symptoms.

DISCUSSION

The diagnosis of Gardner's syndrome can be made under the following propositus; 1) at least two manifestations of the diagnostic triad in the individual without a positive family history, or 2) one mainfestation and a blood-relative with at least two of the manifestations[3]. The present case meets the diagnostic criteria of Gardner's syndrome by both adenomatous polyposis of the large intestine and subsequent development of fibromatous soft tissue masses in variable intervals despite no definite familial history. The significance of lipoma in the neck removed 20 years ago remains speculative but seems an incidental association.

It has been generally recognized that fibromas and desmoid tumors in Gardner's syndorome frequently arose following a surgical incision. Simpsons et al[4] reviewed 7 cases with mesenteric fibromatosis in Gardner's syndrome and noticed that these patients had a history of previous abdominal surgery. The interval between operation and recognition of the mass varied from 3 months to 5 years, and was usually between 1 and 3 years. Abdominal surgery, especially mesensteric surgery, seems etiologically related to the fibrous mesenteric masses that have appeared in these patients. The desmoid tumors noted by other investigators usually have followed an abdominal incision, and trauma also has been suggested as a factor in other subcutaneous fibrous lesion seen in this syndrome. The above proposal is totally supported by the present case which had recognized mesenteric fibromas and desmoid in the incision site during interval of two years after colectomy for adenomatous polyposis.

Many attempts have been made to understand the pathophysiology of desmoid fibromatosis. Skin fibroblasts from familial polyposis phenotypes were found to have lost serum and density-sensitive growth control in culture[5]. Lack of contact inhibition, elevated levels of plasminogen activator and alteration in intracellular distribution of actin cables were also demonstrated. In "desmoid fibroblastoma" the tumor fibroblasts differ from normal fibroblasts in the production of intracellular collagen and the accumulation of glycogen. Normal fibroblasts pocess an inhibitor which prevents premature cross-linking of the tropocollagen molecule in the Golgi system, while in desmoid fibroblasts this substance may fail to act, allowing collagen to complete its organization the cytoplasm[7,8]. Thus, this molecular defect and abnormal growing properties of fibroblast which might be responsible for the uncontrolled fibroblastic proliferation was proposed as the possible pathophysiologic basis in simple fibromas or other tumors of patients with Gardner's syndrome.

At present the development of post-surgical desmoid or mesenteric fibromatosis poses serious problem beyond polyposis coli itself and illustrate one of the many dilemmas which exist in the management of this unique genetic disorders[9]. Cumulative experiences with long-term follow-up results of the cases will answer the prognostic outcome of this complex disorder.

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= 국문초록 =

선종성 대장 폴립증 수술후의 복직근 및 장간막 섬유종의 발생

— 1예의 Gardner 증후군 —

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조 정 희 · 김 용 일

저자들은 31세 남자에서 선종성 대장 폴립증으로 전 대장 절제술을 시행한 2년 후에 복직근 및 장간막에 섬유종이 발생한 1예를 보고하였다.

본 예는 Gardner 증후군에서 개복술 후 3개월 내지 5년 사이에 장간막 섬유종이 발생되었다는 이제까지의 보고예와 유사한 양상을 보였다. 이는 본 증후군에서의 비정상적 섬유세포의 증식이 선종성 대장 폴립증과 관련된 유전적 배경하에서 개복술에 의한 외상으로 인하여 촉진된다는 가설을 뒷받침해주는 예라고 사료된다.