THE UNCOMMON TUMORS OF THE LARGE INTESTINE

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CARCINOMA is the commonest malignant tumor of the large intestine. Sarcoma occurs considerably less frequently. These two types of malignant growth represent by far the large majority of new growths which come under the observation of the diagnostician and the surgeon. Adenomatous polyps of the colon and rectum are, themselves, of sufficiently frequent occurrence to warrant description and discussion by themselves. Various aspects of these lesions have been reported previously. As diagnostic methods improve and routine examination of the colon and rectum are undertaken in all cases in which there is any form of intestinal dysfunction, other less common tumors are encountered, many of which are benign.

At first thought it might seem that these less common tumors are so rare, and, since they are usually benign, so innocuous as hardly to deserve mention, but when it is recalled that occasionally benign tumors assume immense proportions, cause obstructive phenomena, may be associated with anemia and bodily depletion and a patient may be denied adequate treatment occasionally because such tumors were thought to be malignant and inoperable, a careful study of a group of such tumors seems timely.

Some of these rare tumors may be malignant. In such instances the malignant process usually progresses slowly and causes injury by extension into adjacent tissues. The rare benign tumors of the large intestine may remain symptomless until they assume proportions sufficient to interfere with normal mechanical function of the bowel. Occasionally they produce such complications as intussusception and consequent obstruction. Therefore, it is conceivable that patients have harbored such growths during their entire lives, have never had symptoms from them and have died of other causes with these growths undiscovered. Other such growths, however, have been brought to our attention very forcefully, and occasionally, because of their size and the other features mentioned, have been thought to be incurable.

Unusual pathologic types observed include: fibroma, fibromyoma, fibromyxoma, fibromyxomyoma, adenofibromyoma, fibroleiomyoma, myoma, adenomyoma, angioma, lipoma, cholesteatoma, paraffinoma, papilloma or villous tumor, teratoma, glioma, chordoblastoma, endothelioma, dermoids and cysts of the cecum. Each type of tumor originates from the part of the intestinal wall which corresponds to the histologic structure of that tumor: for example, adenomas from the mucosa, fibromas from the submucosa and serosa, lipomas from the fat cells of the external coat and from the appendices epiplöicae, angiomas from the blood vessels of the intestinal wall, and myomas, according to most investigators, from the muscularis or the muscularis mucosa. Of the benign tumors reported in the literature, adenomas are the most frequent, angiomas the most infrequent and myomas comparatively infrequent.

SYMPTOMS

The group of tumors being discussed has no characteristically distinguishing symptoms. Their presence is usually unsuspected until there occurs mechanical occlusion of the intestinal lumen, until intussusception of the tumor into that part of the bowel which is distal to it takes place, or until volvulus of the involved segment occurs. All of the varieties occasionally produce intestinal obstruction; this is more commonly met in its partial or intermittent form. The partial type of obstruction may merge into an acute complete variety, or may be ushered in by an acute attack which subsides, only to recur at varying periods and then evidenced by paroxysmal cramps, abdominal distention or tenesmus. The attacks of obstruction vary in no way from the attacks of intestinal obstruction resulting from any mechanical cause, and usually characteristic, cramp-like attacks of colic occur, resulting from direct occlusion of the intestinal lumen by the tumor itself, from intussusception caused by the effort of the bowel to rid itself of the tumor, or from torsion and volvulus. The number of tumors varies from a single growth to involvement of all the colon, as it is involved in adenomatous polyposis. The tumors also vary in size from small, sessile elevations to huge pedunculated, solitary tumors which encroach on the intestinal lumen. When the growths are in the ascending or the transverse colon, often the first symptoms are those of obstruction; when they are in the sigmoid or upper part of the rectum, such growths are always more productive of discomfort and of derangement of function. Frequently they cause a sensation of fullness and produce dull pain or soreness in the left part of the abdomen. With growths in the lower part of the colon, and particularly in the rectum, urgency and frequency of desire to defecate, with unsatisfactory and incomplete evacuation, occur. Pedunculated rectal growths are likely to be extruded through the anus and, with extrusion, may be associated partial or complete prolapse of the rectum.

Diarrhea, in one form or another, usually is present at some time during the growth of a benign tumor; it

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varies in severity from intestinal irritation, with several stools per day, to a violent attack with ten or twelve evacuations daily. Usually blood is not present in the stools, although in a case of lipoma it is noted oftener than with other varieties of benign tumor. This is caused by ulceration of the mucous membrane overlying the growth and often results in profound anemia. A number of cases has been reported in which the clinical picture of malignancy has been mimmicked by a benign tumor, usually of the lipomatous or angiomatous variety. Here, the secondary anemia, loss of weight and cachexia, which are incidental to disturbances of gastro-intestinal function, probably are largely responsible for the clinical picture.

TYPES OF TUMOR

Fibroma. Fibromas of the large intestine are extremely rare. They arise from the fibrous tissue found in the submucous coats of the bowel; types such as fibromyoma, fibroleiomyoma, adenofibromyoma, fibromyxoma and fibromyxomyoma have been described. Jansen, in 1886, probably reported the first true fibroma, a hard, mobile tumor, 8 cm. in diameter, in the descending colon of a woman aged thirty-five years. It was successfully resected. To the seven cases of fibroma previously reported from the Mayo Clinic, we should like to add one more striking case.

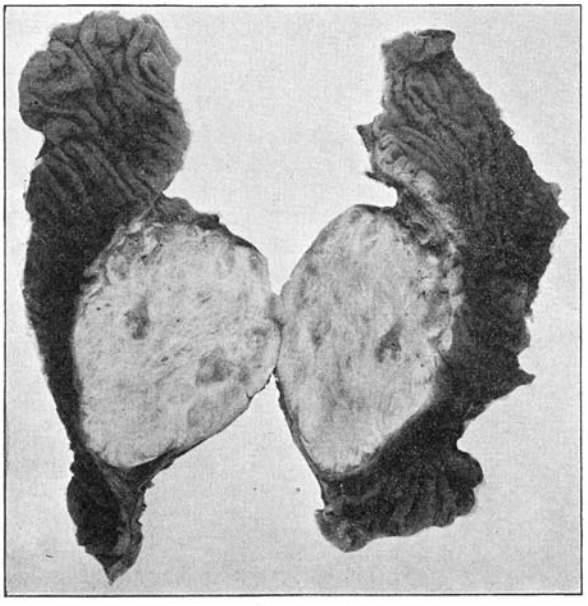

Fig. 1. Gross appearance of fibroma of cecum.

Case 1. A married woman, aged forty-six years, came to this Clinic November 13, 1931, because of a tumor in the right, lower quadrant of the abdomen, and occasional attacks of pain in this region. The pain had been present for about three years, never severe, occurring on quick motion or when turning in bed. In August, 1929, abdominal exploration was carried out elsewhere, with partial oöphorectomy. The woman seemed somewhat better after the operation, but the infrequent pains in the right lower quadrant continued. She felt positive about the presence of a tumor a year before her admission and felt that it had grown somewhat during that time. There were no associated digestive symptoms and the woman made no other complaints.

At the time of her admission, blood pressure was 120 mm. of mercury systolic and 80 diastolic; temperature 98.2 degrees F., weight 166 pounds (75.3 kg.), which was her usual normal. There was a firm, smooth, movable tumor in the right lower quadrant, about 12 cm. in diameter. We were struck with its unusual mobility. We were able to slide the tumor along in the left loin below the kidney; the latter could be felt separately. *Roentgenologic* examination disclosed a hyperplastic lesion involving the cecum.

November 21, 1931, ileocolostomy was performed. January 23, 1932, right hemicolectomy was performed for a fibroma of the wall of the cecum, not involving the mucosa; the tumor was 6 cm. in diameter (Fig. 1). Recovery was uneventful and the patient has remained well.

Myoma. Pure muscle tumors are exceedingly rare; most tumors designated as such contain varying amounts of muscle and fibrous tissue. The first true myoma of the rectum was reported by Vander Espt in 1881. Hunt, reviewing the literature back to 1872, classified twenty cases as examples of myoma or myofibroma of the rectum. He added reports of four cases encountered at the Mayo Clinic. Of this total of twenty-four patients, eleven were males and thirteen females; the youngest patient was aged twenty-one years; the oldest, eighty-five years. These tumors may assume great proportions; the largest we found was 15 cm. in diameter. Marked constipation, hemorrhage, and rectal pain were the predominating symptoms. Complete obstruction occurred in three instances. Malignant change has been known to develop in these tumors. The tumors are cellular and tend to recur after enucleation. Furthermore, after repeated removal, it has been noted that successive examination of the specimen discloses increasingly greater cellular infiltration and malignant change. Such a case was reported in 1932[17] and another follows:

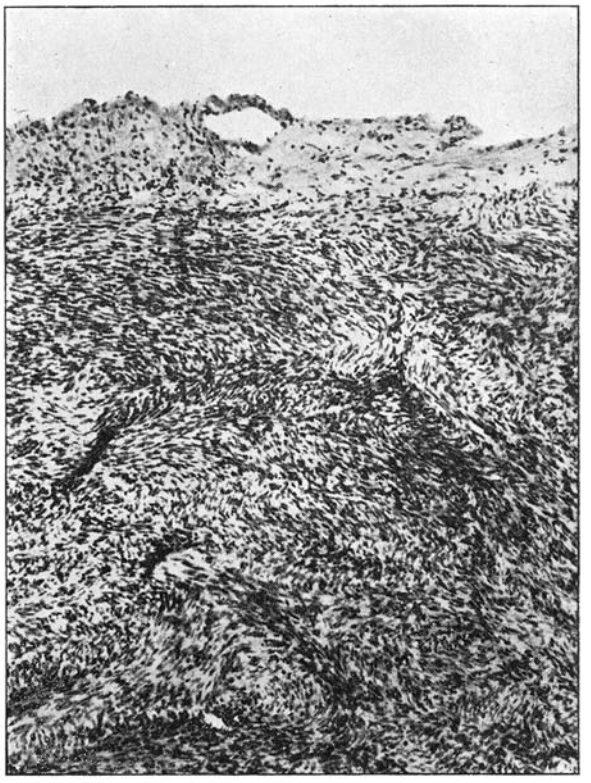

Fig. 2. Microscopic section of myosarcoma (x 75).

Case 2. A man, aged fifty-one years, a confectioner, came to this Clinic, April 10, 1931, because of a small lump behind the anus. He had first noticed the lump about 1922, and it had slowly, gradually, enlarged to a diameter of about 7 cm. by August, 1929. The tumor did not cause pain. At that time it was removed elsewhere and was designated as a "shell tumor, suggestive of cancer". The wound healed properly; there was no drainage and no recurrence of the tumor for eighteen months. About that time, the man noticed a painless lump the size of a walnut in the same region as the first tumor. When the patient was examined at this Clinic several months later, the same rounded tumor was found at the posterior wall of the anus, and measured about 3 by 3 cm. The lesion appeared to be beneath the submucosa; there were no ulcers and no bleeding. Over it was the scar of the former operation.

April 14, 1931, under sacral anesthesia, the tumor was removed by resecting the left half of the anus. Pathologically, the lesion was a myosarcoma, graded 2.

The man returned in February, 1932, at which time the re-

current lesion was about as large as it had been in April, 1931. Single-barrelled colostomy was performed, and the entire rectum and its perirectal structures were removed by combined abdominoperineal resection. Pathologically, the lesion was 3 cm. in diameter and again was a myosarcoma graded 2 (Fig. 2). There was no involvement of lymph nodes. The patient made an uneventful recovery.

Adenomyoma. Spencer, in discussing a paper by Lockyer, in 1913, probably was the first to refer to a case of adenomyoma of the sigmoid. In 1920, Cullen reported four cases of adenomyoma of the sigmoid, two from own experience and two from the literature. Ten cases were reported from this Clinic in 1932.[17] Nine of the adenomyomas occurred among women between the ages of twenty-seven and forty-eight years; the tenth one occurred in a man aged eighty years. The tumors of the women were at the sigmoid, and might have been implants or extensions from former pelvic lesions. All were found in relation to pelvic disease, and in several cases, by their annular nature, produced obstruction of the sigmoid. Several of the tumors were possibly of ovarian origin. Enucleation was possible in some cases, but resection of the sigmoid was necessary in others. Adenomyoma of the recto-vaginal septum occurred fairly frequently, but consideration of them has been omitted here. The adenomyoma of the man aged eighty, occurred as a hard, postanal tumor that was diagnosed preoperatively as carcinoma.

Angioma. This is perhaps the rarest type of tumor found in the large bowel, and varies in size from a small nevus to a pedunculated type which grossly may attain considerable size. Apparently the tumors are of congenital origin. Involvement of the whole colon in an angiomatous process has been noted, but it is extremely unusual to find this type of growth in the colon except in its lower segments. The tumors usually arise in the submucosa, but Buie and Swan found the origin of the tumor in the serosa in one of their cases. Ulceration occasionally takes place with resultant hemorrhage, but there is otherwise little about these lesions to suggest their pathologic nature. The angiomas described have been hemangiomas. Of those cases of angioma for which operation was performed at this Clinic, one of the tumors was in the wall of the cecum; it was accidentally discovered in the course of an operation for gallstones. One occurred in the splenic flexure, one in the sigmoid, one at the recto-sigmoid and one in the rectum. The latter three were diagnosed as polyps preoperatively, and the one at the splenic flexure as carcinoma because of the type of roentgenographic filling defect.

Lipoma. Lipomas of the large intestine should be considered to be only relatively rare, for after adenoma (not considered in this paper) lipoma is the most frequently encountered benign tumor of the gastrointestinal tract. However, as Comfort pointed out in 1931, only seventy-four symptom-producing lipomas of the large intestine had been reported in all medical literature available to him. The diagnosis has been made before operation only in exceptional cases. It is rare that the diagnosis can be more accurate than that of a benign tumor, although in some instances, in which the tumor is visualized through the proctoscope, or in which it has prolapsed to the outside, its character is apparent.

The age of patients with lipomas, the frequent appearance of blood in the stools, and the loss of weight, together with the attacks of obstruction, make difficult the differential diagnosis between lipoma and carcinoma.

Cholesteatoma. One variety of tumor, apparently never before reported as having been found in the large bowel, namely, cholesteatoma, was first recorded by Humiston and Piette, in 1925. In their case, the tumor was situated in the cecum; in a review of the literature they failed to find a report of any other tumor of this epidermoid variety in this situation. A cholesteatoma is described as a "cystic tumor with thin epithelial lining, and a content which is formed of epithelial cells and cholesterol". Tumors of this variety presumably originate from misplaced, embryonic, epidermoid germ cells. Similar changes occasionally are produced by chronic inflammation.

The cecal cholesteatoma, described by Humiston and Piette, was found in a young man who presented typical symptoms of subacute appendicitis; at exploration there was found an inflamed appendix with an encysted tumor in the outer wall of the cecum, which tumor did not connect with the lumen of the bowel and was easily enucleated. Gross and microscopic examinations confirmed the diagnosis of true cholesteatoma of the cecum.

Dermoids. True dermoids of the rectum occur very rarely, although dermoid cysts and tumors in the perirectal spaces are not uncommon. Our contribution is not concerned with the postrectal dermoids, those of the rectovaginal septum, nor those invading the rectum from nearby structures, as for example, from the ovary. True dermoids may arise in the rectal wall; for this reason they produce symptoms relatively early. They may be pedunculated and so protrude from the anus, or produce obstruction by invagination of the wall of the bowel. The first sign may be protrusion of hair from the anus. The patients may suffer from constipation of obstructive type, marked tenesmus, and, at the time of defecation, the tumor may present at the anus. The tumor may consist of skin covered with hair and sebaceous follicles, enclosing fat and fibrous tissue. Dermoid cysts have been adequately described by Bensuade and Rachet, Port, Saphir, Maingot, and Danzel.

Teratoma. Although there is no sharp line of demarcation between dermoids and teratomas, in the strictest sense, a teratoma is a tumor containing tissue and fragments of viscera and without the cystic fluids of dermoids. In Fried and Stone's case, the lesion was a teratoid mass of mixed tissue, and instead of being a cavity lined with skin, it was a solid mass covered by skin. The tumor swung freely in the lumen of the bowel, attached by its pedicle to the posterior rectal wall. It was comma-shaped, with a smooth surface, and covered with small patches of brown hair. At one point, a hard, pearly-white object projected; it resembled a tooth. Microscopically, it was surrounded by cornified squamous epithelium, and it contained smooth muscle, connective tissue, fat, sweat glands, bone and hair follicles.

Cysts of the cecum. Few instances of this condition have been reported. McAuley reviewed the literature up to 1923, and found reports of eleven cases; he reported another case. Bazin, in 1925, added still another. McAuley argued that the condition should be considered as a definite entity. In the literature there is a fair number of references to enteric cysts and other cysts of the intestine. Such cysts produce symptoms of obstruction, or are discovered at necropsy or operation. The condition they cause usually is diagnosed as irreducible intussusception; the diagnosis is not established until the resected specimen is examined.

Rath reported the case of a woman, aged forty years, in which the diagnosis was hydrops of the appendix,

but on exploration the mass was found to be a cyst of the cecum with its base 3 cm. distant from a normal appendix. Flick reported a case in which a large cyst had twisted itself on a pedicle 1.5 cm. long; the cyst contained bloody fluid. This had brought on the clinical picture of acute disease within the abdomen.

In the years for which the records of this Clinic were reviewed, two cysts of the cecum were found.

Papillomas or villous tumors. These tumors have been variously designated as papillary tumors, villous carcinomas, villous polyps, papillary polyps and papillary adenomas. Whether they should be included here is doubtful. We mention them only for the sake of completeness.

Paraffinoma. This tumor results from injection of paraffin. When the tumor is in the lower part of the rectum, the injection has been made for hemorrhoids.

Chordoblastoma and glioma. Although these are not primarily rectal tumors, occasionally the symptoms they cause are referable to the rectum; hence, they should be mentioned in any discussion of unusual rectal new growths.[21]

Endothelioma. Endotheliomas are similar to gliomas in origin and clinical characteristics. They have been found to remain quiescent for many years and then, suddenly, for inexplicable reasons, to grow with great rapidity. The symptoms depend primarily on the size, situation and direction of extenison of the tumor. Usually they are superficial. When they arise in the rectum proper, their symptoms are those of obstruction; their diagnosis usually is suggested by digital and proctoscopic examination. Because of the relatively early invasion of the spinal cord by all tumors which arise from neural structures, pain in the region of the sacrum and coccyx is common, and by impingement on the sacral and lumbar plexuses, the tumors may give rise to sphincteric disturbances and to paresthesia in the perianal region and in the region about the urethra. Sensation of incomplete evacuation, dragging and heaviness in the pelvis and painful defecation also may be observed.

Roentgenograms are of value in revealing spina bifida or other bony defects in these regions or they may reveal bony structures and teeth. At times, these neural tumors result in the erroneous diagnosis of sciatica, coccygodynia, arthritis and rectalgia of other types.

TREATMENT

Treatment of all of these lesions is surgical. In some patients, local excision is all that is required. This is particularly true of those benign tumors which can be seen by the sigmoidoscope. Sometimes lipomas, fibromas and other tumors are on a pedicle; then fulguration with the electric cautery is the treatment of choice. Many times, however, when lesions are above the rectosigmoid, radical resection is the appropriate treatment because the important question of the malignant nature of the tumors cannot be settled before they are removed.

Thus, the rectal fibromas were excised locally, but for the cecal fibromas ileocolostomy and subsequent right hemicolectomy were performed.

In reviewing the records of the cases of myoma, local recurrence with repeated excision was common. Specimens removed on successive examinations were recorded by the pathologists as being examples of myoma, cellular myoma, and malignant myoma. Hence, it seemed wise, in the case of the recurrent myosarcomas, to perform radical resection as described in Case 2.

Angioma, because of the size it may attain and its clinical similarity to carcinoma, usually requires radical excision. Cholesteatoma may be enucleated from the colonic wall without radical resection of the intestine. Because of their tendency to recur, villous tumors are best treated by radical extirpation. The paraffinomas reported in the literature were both resected; the lower end of the anal canal with sphincters was preserved and results were completely satisfactory. For the neurogenic lesions, excision is the usual treatment. In the case of chordoblastoma, excision of the lesion and insertion of radium have given satisfactory results.

When feasible, the treatment of choice of endotheliomas is complete surgical extirpation. The posterior approach, with at times removal of the coccyx and lower sacral vertebrae, may be necessary. Radium and roentgen rays have supplemented surgery with indefinite results. Prognosis in cases of the malignant types of tumors is unfavorable, generally, and is patricularly bad when associated with meningocele.

The treatment of dermoids consists of complete removal and dissection, with the tumor intact. However, removal of the sac of a large dermoid may be facilitated by first evacuating the content. Exposure of the tissues to the content of the dermoid usually will set up a violent inflammatory condition. Every portion of the wall of the cyst must be removed; otherwise the wall will not close and the cyst will re-form. Complete removal and not drainage is essential for permanent cure. Cysts of the cecum, because of the mechanical difficulties which they produce, should be removed surgically.

This brief review of uncommon tumors of the large bowel, if for no other reason, seems justifiable because such tumors occur just often enough to warrant their possible presence being kept in mind when dealing with tumors generally.

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