Lipohyperplasia of Ileocecal Valve, Causing Recurrent Intussusception

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Lipohyperplasia of the ileocecal valve is a fairly common entity that occasionally behaves as an intestinal tumor, causing obstruction, sometimes with intussusception, or bleeding, which may be acute or chronic. Most often occurring in middle-aged or elderly women, it may be mistaken clinically and radiologically for carcinoma or other neoplasms. Its differentiation is best made by endoscopy, confirmed if possible by biopsy. Differentiation is important, as the treatment is a limited ileal and cecal resection, instead of a blind hemicolectomy. A recent study suggests possible associations with other medical conditions. We describe the clinical and pathological findings of this entity in a patient who had recurrent bouts of intussusception over a period of seven years, causing gastrointestinal symptoms and a palpable mass, eventually requiring surgical treatment. (Henry Ford Hosp Med J 1990:38:259-61)

Case Report

A 48-year-old woman presented as an emergency at Macomb Hospital Center in October 1988, with progressively worsening abdominal pain, vomiting, and rebound tenderness in the right lower abdominal quadrant. She had a five-year history of episodic bouts of pain with cramping in the right lower abdominal quadrant, accompanied by nausea but no vomiting. The cramps were sometimes followed by diarrhea and a palpable, hard, “small egg-sized” lump in this region which often persisted long after the pain had gone and once “almost refused to go away.” There was no other significant medical history.

Because of persistent lower abdominal pain and leukocytosis, appendicitis was suspected. Laparoscopic examination revealed a yellow, subserosal mass bulging from the cecum in the region of the appendix. At this time the differential diagnosis included a walled-off, perforated appendix or a carcinoid tumor. At laparotomy, a congested-looking but otherwise normal appendix was removed. A “mass” separate from the appendix was palpated in the ileocecal region. Ultrasound of the pelvis and abdomen in the immediate postoperative period showed a questionable pelvic mass to the right of midline. Computed tomography (CT) two days later showed a filling defect in the cecum and at its junction with ascending colon. Endoscopic examinations three weeks later and in January 1989 and January 1990, all done at times of recurring symptoms, revealed on each occasion a smooth, yellow, rounded mass, intussuscepting and filling the ileocecal lumen (Fig 1). The differential diagnosis in order of likelihood was lipohyperplasia of the ICV, a carcinoid tumor, or some other submucosal neoplasm. Carcinoid was included in the differential diagnosis because these tumors are characteristically submucosal and yellow and sometimes present with intestinal obstruction or intussusception. Moreover, 60% to 80% of carcinoid tumors occur in the midgut, with the appendix and distal ileum as their most common sites (4). More than 40 years ago, Horne (5) stressed the ICV as an important primary site for carcinoids and described two examples in this location.

Colonial mucosal biopsies, taken at our patient’s endoscopies in 1988 and 1990, showed mild, nonspecific chronic inflammation and congestion, respectively, with no evidence of carcinoid or other neoplasm. Because of recurring symptoms, the patient underwent a partial right hemicolectomy in February 1990. The pathological findings are illustrated in Figs 2 and 3. She was asymptomatic when last seen in November 1990 and is considered cured of her condition.

Discussion

The first published series of intestinal lipomas was by Comfort (6) in 1931. Only two of his 181 cases had “lipomas” situated at the ICV. Golden (7) is credited with first describing
roentgenographic changes that help to distinguish pathological enlargement of the ICV from malignant neoplasm. He noted that lipohyperplasia presents as a smooth, radiolucent, lobulated mass, with the lucency being an important diagnostic criterion. Current techniques of CT or barium enema may show a filling defect or distortion in the region of the ICV, which may be difficult to distinguish from a malignant neoplasm.

In 1948, Kelby (8) was the first to differentiate clearly the pathologic entity of “lipomatosis of the ICV” from encapsulated and pedunculated lipoma. Shortly thereafter, Edwards and Zangara (9) used the term “lipomatous hypertrophy” in describing two cases in which clinical symptoms and roentgenographic changes were present. In 1953, Zettergren (10) suggested that the lesion should be called lipohyperplasia rather than lipomatosis, to avoid confusion in terminology. Lasser and Rigler (11) studied over a period of several years 18 patients (16 of whom had symptoms) with enlarged ICV revealed by roentgenography and coined the term “ileocecal valve syndrome.” Their patients were all women over 45 years of age who had experienced crampy abdominal pain, gas, and bloating that had aroused suspicions of gall bladder disease. These authors postulated that the symptoms were due to chronic intussusception or partial obstruction. In 1956, Seabrook et al (12) emphasized that “lipomatosis of the ICV” may cause exsanguinating hemorrhage. In 1959, Cabaud and Harris (13) studied ten cases and noted an association with local inflammatory disease, such as diverticulitis or adhesions, in many of their patients. Elliott et al (14) further defined the clinical and radiological features of ICV lipohyperplasia in 1968.

The medical literature concerning the frequency of ICV hyperplasia is confusing. Morson and Dawson (3) and Cooper (15) state that it is a fairly common entity usually found in operative specimens removed for other conditions. We suspect that ICV hyperplasia may not always be included in the pathological diagnosis of colectomy specimens, particularly if it is not severe and only an incidental finding. This impression is supported by a recent report by McGregor and Tawfik (16). They studied a series of 30 consecutive autopsies to determine the frequency of ICV lipohyperplasia and any associated pathologic entities. In estimating valvular lipohyperplasia, 7 (23%) of the studied colons were found to have none, 8 (26.7%) mild, 11 (36.7%) moderate, and 4 (13.3%) marked. The magnitude of lipohyperplasia appeared to correlate with degree of right ventricular and pancreatic fatty infiltration and with “greater body weight” but not with left ventricular, adrenal, or lymph node fatty infiltration or hepatic fatty change. The patients’ clinical histories recorded various gastrointestinal symptoms and lesions accompanying lipohyperplasia. Despite this, with the exception of one case of marked lipohyperplasia associated with marked mucosal necrosis and acute inflammation of ICV, no definite causal relationship was established. These same authors also reviewed seven cases of ICV lipohyperplasia diagnosed in surgical specimens (six resections and one biopsy). Three cases had right lower quadrant pain and ICV mass (identified either by barium enema or by operative examination), and two cases had ICV mucosal acute inflammation and necrosis. The other two were incidental findings in resections for appendiceal neoplasms. The one case diagnosed on endoscopic biopsy showed excessive adipose tissue in the submucosa (personal communication, Dr. Douglas H. McGregor).

**Summary**

ICV hyperplasia is a fairly common entity resulting from an excess of adipose tissue in the submucosa of the ileocecal valve. The thickened valve protrudes into the lumen and may occasionally be associated with clinical symptoms caused by intestinal obstruction, intussusception, or hemorrhage. Clinically, the
Pig

Cross section of ileocecal valve (hematoxylin-eosin stain). Note the excess of adipose tissue (arrow) producing a circumferential thickening and pouting of the valve which protrudes into the cecum. The excess adipose tissue is not encapsulated and fades away on both sides of the valve. The overlying mucous membrane is intact in this instance.

lesion may be confused with an adenoma, carcinoid tumor, or carcinoma at radiologic or endoscopic examination. Preoperative diagnosis is important when surgical treatment is being considered for symptoms, as a limited ileal and cecal resection is preferable to a blind hemicolecction. Endoscopic examination with biopsy is highly recommended as it remains the best method of diagnosing lipohyperplasia.

References