Inflammatory Myofibroblastic Tumor of the Ileum Causing an Unusual Ileocecal Intussusception

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Inflammatory myofibroblastic tumor (IMT), also known as inflammatory fibrous polyps or inflammatory pseudotumor, is a rare mass-forming lesion characterized by fibroblastic or myofibroblastic spindle cell proliferation with varying degrees of inflammatory cell infiltration. IMTs usually present in children and young adults. IMTs occur only rarely in the gastrointestinal tract, and these usually occur in the stomach as well as in the small and large bowel. When localized in the small bowel, the presenting symptoms are colic abdominal pain and obstruction. Intussusception due to IMT is uncommon. We report one case of an IMT of the terminal ileum in a 50-year-old female with presentation of ileocecal intussusception and intestinal obstruction.

Key Words: ileum; inflammatory myofibroblastic tumor; intussusception

Introduction

Inflammatory myofibroblastic tumor (IMT) is sometimes known as inflammatory fibrous polyps, inflammatory pseudotumor or plasma cell granuloma, and these terms have been used interchangeably for rare proliferated lesions, which clinically resemble a malignant neoplasm occurring in both pulmonary and extrapulmonary tissues. The etiology of these lesions remains unclear. Both human herpesvirus 8 and overexpression of interleukin 6 and cyclin D1 in myofibroblasts have been implicated in the tumorigenesis of IMT. Other causes postulated to induce IMT from infection include Mycobacterium avium-intracellulare, Campylobacter jejuni, Bacillus sphaericus, Coxiella burnetii, Escherichia coli, Corynebacterium equi and Epstein-Barr virus, although the evidence is inconclusive. IMT is more common...
Inflammatory myofibroblastic tumor in children; there is no sex predilection, and it occurs most commonly in the lung and mediastinum in all age groups. Extrapulmonary IMTs are rare and occur in a wide variety of extrapulmonary body sites but are the most common in the mesentery and omentum with a predilection for children and young adults. In the alimentary tract, it is commonly found in the stomach and also in the small and large bowel, originating from submucosa and growing as a polypoid mass. Alimentary IMT is seldom diagnosed before an operation owing to its variable presentations. Clinical symptoms and presentations of IMT depend on its location and growth pattern. IMT seldom presents in the ileum, and intussusception is a rare complication of IMT; in particular, ileocecal intussusception has infrequently been reported. We report an intramural ileal IMT in a 50-year-old woman with the particular presentation of ileocecal intussusception.

Case Report

A 50-year-old female presented with a 10-day history of intermittent and worsening colicky abdominal pain with progressive abdominal fullness. The pain was located over the suprapubic area with cramping and was not related to position or meals. Nausea and vomiting were accompanied by bilious vomiting. A bloody stool was also noted 2 days before her hospital admission. Her past surgical history included bilateral ovarian tumor after bilateral salpingo-oophorectomy. On clinical examination, she was a relatively obese patient, was dehydrated and had tachycardia (pulse rate, 114/min). Physical examination revealed a distended abdomen with a mildly tender mass over the right lower quadrant on palpation and decreasing bowel sounds on auscultation. Laboratory investigations showed a raised white blood cell count of 17,900/μL. The other laboratory data were within normal limits. Abdominal X-ray showed dilated small bowel segments. A computed tomography scan of the abdomen demonstrated a typical intussusception complex, including a well-defined intraluminal mass of soft-tissue density, accompanied by a target sign, and dilated loops of small bowel proximal to the tissue mass (Figure 1). We made the decision for operative intervention.

At laparotomy, the patient was found to have a large intussusception in the distal ileum just proximal to the ileocecal valve with a polypoid mass in the distal ileum (Figure 2). During the operation, manual reduction of the intussusception and segmental resection of the terminal ileum were performed, and the bowel was anatomized in an end-to-end fashion. A cut section of the tumor showed that the tumor arising from the submucosa of the ileum and its surface was covered with ulcerated mucosa (Figure 3), and in the tumor and the pericolic region, several lymph nodes were found. The pathology report showed that the tumor was an IMT measuring 4 × 3 cm, with focal mucosal erosion. Microscopically,
the ileum showed a polypoid lesion composed of proliferative vasculature, spindled stromal cells, and mixed inflammatory cells in the myxoid stroma (Figure 4). The mucosal surface was extensively ulcerative with necrotic cellular debris accumulation. The blood vessels near the ulcerated surface showed blue-purple particles deposited on the wall, with infiltration of inflammatory cells. Immunohistochemically, the myofibroblastic spindle cells were negative for cytokeratin, positive for smooth muscle actin (Figure 5), negative for CD34, and focal positive for Bcl-2. Gram, Gomori’s methenamine silver and Warthin-Starry silver stains failed to demonstrate any microorganisms. Histopathologically, the lesion was compatible with IMT. All regional lymph nodes showed reactive hyperplasia. The postoperative course was uneventful, and the patient was discharged 9 days after the operation.

Discussion

Gastrointestinal tract IMT is a pseudotumor with a peculiar polypoid growth pattern; it was first described in 1939 as a gastric submucosal granuloma with eosinophilic infiltration and has been given many different names. IMT has been previously referred to as inflammatory fibrous polyps, inflammatory pseudotumor, plasma cell granuloma, atypical fibromyxoid tumor, polyp with eosinophilic granuloma, fibromyxoid tumor, and pseudosarcomatous. The most commonly affected sites are the lung and mediastinum in all age groups, but it is more common in children without sex predilection. The lesion has a distinctive histologic appearance and usually presents as a benign, non-metastasizing proliferation of myofibroblasts with a potential for recurrence and persistent local growth. However, the most common sites of extrapulmonary IMT are the mesentery and omentum. IMTs of the gastrointestinal tract are rare with the most common site in the stomach, but there are rare cases in the colon and small bowel. The present symptoms and signs depend on the location and growth pattern of the primary lesion. These patients often present with abdominal pain and fullness, fever, palpable mass, weight loss, and other vague nonspecific symptoms. Obstruction or intussusception is the most frequent initial symptom when an IMT is located in the small bowel. The tumor often mimics a malignant tumor clinically and radiologically even through magnetic resonance imaging. Abdominal computed tomography scans have been used in the diagnosis of IMTs, with a 50–80% diagnostic rate with a typical “target” or “bulls-eye” sign, although this is not absolutely specific. We performed a computed tomography scan of the abdomen that revealed a typical intussusception complex, including a well-defined intraluminal mass of soft-tissue density mimicking a malignant mass with a typical target or bulls-eye sign.
The pathogenesis of IMTs is unknown. Our patient had a history of intra-abdominal surgical procedures performed several years previously, suggesting that trauma may have initiated the process. Another feature of note is that our patient was a relatively obese female. A female predominance has been reported, but an association with obesity has not been described in the literature. Since IMTs have no distinctive radiologic and clinical findings, histologic confirmation is necessary in all cases to exclude malignancy. Surgical resection of the specimen is preferred to an endoscopic biopsy for diagnosis owing to the submucosal origin of the lesion, which usually shows ulceration in the overlying mucosa. IMTs of the intestine are potentially invasive and composed of spindle cells, which proliferate in a myxoid to collagenous stroma with a prominent inflammatory infiltration and are typically uniform with predominant myofibroblastic characteristics. Differential diagnosis should include a spindle-cell lesion from mesenchymal neoplasms or other malignant masses in the abdomen including leiomyosarcoma, inflammatory fibrosarcoma, idiopathic sclerosing mesenteritis, spindle-cell carcinoid rhabdomyosarcoma, and malignant fibrous histiocytoma. Aggressive en bloc resection of the tumors remains the best treatment for patients with IMTs. Otherwise, IMTs of the ileum may lead to ileocecal intussusception, causing partial or total small bowel obstruction, which is a surgical emergency. Laparoscopic surgery is reported to have a role in the management of adult intussusception. Intracorporeal resection may be simple or complex, ranging from reduction to more complicated or even colonic cases. However, laparotomy with extracorporeal resection is usually indicated in acute presentation of intussusception causing partial or total small bowel obstruction. Several recent studies have reported that recurrence of IMTs is extremely infrequent following complete excision of a solitary lesion. These patients should undergo total resection, depending upon the patients’ symptoms and whether intussusception is present. Some patients with IMT have a past history of intra-abdominal surgical procedures, suggesting a possible mechanism of injury of the gastrointestinal tract that may have initiated the process.

IMTs are classified as tumors of intermediate biologic potential by the most recent World Health Organization classification owing to the tendency for local recurrence and a small risk of distant metastasis. The recurrence rate varies with the anatomic side and is 25% for extrapulmonary lesions. Distant metastasis of IMTs is rare. The most common sites of metastasis are the lung and brain, followed by liver and bone. Metastatic disease is usually identified at presentation or within a year of diagnosis; but occasionally, patients develop metastases up to 9 years following excision.

The search for prognostic indicators in IMT has focused on a combination of genetic and morphologic studies. IMTs should be regarded as potentially local recurrent lesions with uncertain malignant potential. Therefore, a long-term follow-up is important to recognize the recurrences earlier, and the optimal management of local aggressive and recurrent disease should be decided individually for each patient.

References