Inflammatory Fibroid Polyp of the Terminal Ileum Presented with Ileus: A Case Report

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Abstract: Inflammatory fibroid polyp (Vanek tumor) is a rare benign lesion which originates from the submucosa of the gastrointestinal tract. The most common localization is the gastric antrum. Although the etiopathogenesis is unknown, it is thought that inflammatory fibroid polyps are the result of reactive inflammatory processes because of their eosinophil-rich stroma. Microscopically, fibrous connective tissue, inflammatory cells and blood vessel networks are observed in the stroma. Differential diagnosis involves a large number of benign and malignant lesions such as gastrointestinal stromal tumor, schwannoma, leiomyoma, and leiomyosarcoma. In this case, we present a 60-year-old patient with oversized inflammatory fibroid polyp in the terminal ileum which caused ileus.

Keywords: Inflammatory fibroid polyp, Terminal ileum, Submucosal tumor, Ileus.

INTRODUCTION

Inflammatory fibroid polyp (IFP) is a rare benign tumor that appears grossly as localized submucosal polypoid mass in the gastrointestinal tract [1-3]. IFP can occur in any part of the gastrointestinal tract. The most common site is the gastric antrum but it can be seen in esophagus and in the small and large intestines, too [1-6]. It occurs in both children and adults, with the average age being 60 [7]. Males have a slightly greater predominance [8]. IFP was first described by Vanek in 1949 [9]. Microscopically, it is characterized by eosinophil-rich inflammatory cells, myxoid stroma with spindle-shaped cells that are concentrically arranged around the vessels [10]. The etiopathogenesis is still unclear. But because of the eosinophil-rich inflammatory infiltration, it is thought to be reactive inflammatory process contributory [1-6]. Surgical resection is usually necessary to confirm the diagnosis and alleviate the symptoms of the big-sized tumor. In this case, we present a 60-year-old patient with oversized inflammatory fibroid polyp in the terminal ileum which caused ileus.

CASE REPORT

A 60-year-old woman was referred to our hospital with a suspected ileus. An ilioileal-type intussusception and a solid lesion in terminal ileum was described at abdominal computerized tomography (Figure 1). The patient was immediately operated. 28-cm-long terminal ileum resection material was sent to our pathology department. In the gross examination, the specimen contained a 4.5x4x3.3 cm sized, gray colour, submucosal polypoid tumoral lesion (Figure 2). The tumor was as large as the lumen. The surface mucosa was normal. Microscopic examination showed that the lesion was localised in the submucosa and contained loose fibrous-myxoid connective tissue, especially eosinophil-rich inflammatory cells, spindle stromal cells, which were concentrically arranged around the vessels (Figure 3). There were no pleomorphism, atypical mitosis and necrosis in the tumor. Immunohistochemically, the spindle-shaped cells are strong positive with vimentin, CD34 and smooth muscle actin (SMA), negative for S-100 and CD117 (Figure 4, 5). The case was reported as inflammatory fibroid polyp with these histopathological and immunohistochemical features.

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Fig-1: Abdominal CT images from a 60-year-old woman who presented with generalized abdominal pain, nausea and vomiting. (A, B) Axial contrast-enhanced CT shows ileoileal intussusception as target sign (black arrow), and dilatation in distal ileal segments (long white arrow). Solid mass (short white arrow) located at terminal ileum which causes intussusception is seen.

Fig-2: Gross appearance of inflammatory fibroid polyp, well-defined submucosal polypoid mass

Fig-3: Loose connective tissue, especially eosinophils rich inflammatory cells and spindle stromal cells(H&E stain x100)
DISCUSSION

IFP is a benign and rare submucosal polypoid tumor that is seen most commonly in the sixth decade of life, arises from the gastrointestinal tract, usually in the stomach and small and large bowel [1-7]. In the past, inflammatory fibroid polyp was described under names such as eosinophilic granuloma, inflammatory pseudotumor, granuloblastoma, neurofibroma, and hemangiopericytoma [11]. Vanek [9] defined this lesion as “gastric submucosal granuloma with eosinophilic infiltration” in 1949. Then, in 1953, Helwing and Ranier [10] used the term “inflammatory fibroid polyp” for this tumor.

Macroscopically, IFPs are localized, firm submucosal sessile or pedunculated masses, with varying sizes. Tumor’s clinical symptoms depend on the localization and size and can vary from incidental findings to obstruction [8]. Ethiopathogenesis is still unknown [1-6]. Due to the histological nature of the lesion, it is thought as a reactive process or a reparative response [10, 12]. Takeuchi et al. [13] have proposed that IFP is caused by a parasitic infection. Some other authors have taught that IFP may be caused by Helicobacter pylori infestation [14, 15]. On the other hand, Nakase et al [12]. supported that IFP is a benign reactive process with myofibroblastic morphology. In our case there was neither morphological nor serological evidence for any parasitic infestationan. The serum electrolytes were normal and there was no peripheral blood eosinophilia.

Microscopically, the tumor consists of spindle-shaped stromal cells, prominent large and small blood vessels and an inflammatory infiltration composed of eosinophils, plasma cells, lymphocytes and mast cells. Spindle stromal cells are arranged concentrically around the vessels with an “onion-skinned” view [1-10].

Differential diagnosis of the tumor includes spindle-shaped mesenchymal, submucosal and mural tumors of the gastrointestinal tract like leiomyoma, schwannoma, gastrointestinal stromal tumor and solitary fibrous tumor. Absence of pleomorphism, atypical mitoses and necrosis are important clues to exclude a malignant condition. The histomorphological nature of the lesion and immunohistochemical findings are helpful for the differential diagnosis. The most important differential diagnosis is gastrointestinal stromal tumor because of the the management and prognosis involved [8]. Lesion’s morphological nature and CD117 immunostain negativity is useful to exclude GIST. We could distinguish the IFP from schwannoma with S-100 protein negativity, from leiomyoma with...
SMA negativity and CD34 positivity. Eosinophilic gastroenteritis may also be considered for differential diagnosis because of the IFP's eosinophil rich stroma. But eosinophilic gastroenteritis doesn't present as a mass or tumor. However, some parasitic infections may cause intense granulomatous reaction. Therefore, absence of parasitic eggs and larvae is important for the diagnosis of IFP.

Clinical symptoms vary depending on the location and size of the lesion [16]. The two most common complications of ileal IPF are ileus and invagination. Surgical excision is usually necessary because of the size and locatication of the tumor and difficulty of diagnosing the lesion with endoscopic biopsies [2, 12]. Because of IFP’s submucosal localization, endoscopic biopsies are not always helpful for diagnosis. Our patient underwent an ileal surgical resection.

CONCLUSION

Intestinal IFPs must be kept in mind as a rare entity in the differential diagnosis of the gastrointestinal masses which cause to invagination and ileus. Additionally, we think that detailed histopathological examination and using immunohistochemical stains are helpful to confirm the diagnosis of these tumors and to differentiate IFPs from other mesenchymal neoplasms.

REFERENCES