Unexpected Intussusception Caused by Giant Inflammatory Fibroid Polyp: Case Report

Mehmet Öncü, Serdar Altınay, Ali Solmaz, Meral Uyar, Esra Bilgi, Hakan Yiğitbaş

ABSTRACT
Inflammatory fibroid polyps (Vanek’s tumor) are non-frequent benign lesions, described by Josef Vanek in 1949, that originate in the submucosa of the gastrointestinal tract and usually reach 1 to 3 cm. They are generally seen in the stomach and the small intestines, and are less common in the colon and the esophagus. Inflammatory fibroid polyps (IFPs) are one of the rare conditions leading to intestinal obstruction in adults. A differential diagnosis includes gastrointestinal stromal tumor, small intestine lymphoma, and small intestine cancer. We presented here a case with a giant inflammatory fibroid polyp of the terminal ileum causing intussusception.

Key words: intussusception, giant, inflammatory polyp, intestine

INTUSSEPSİYONA NEDEN OLAN DEV İNFLAMATUAR FİBRİOD POLİP; OLGU SUNUMU
ÖZET
İnflamatuar fibroid polipler (Vanek’in tümörü), Josef Vanek tarafından 1949 yılında tanımlanan, gastrointestinal traktnın submukozasından köken alan ve genellikle 1-3 cm ölçülerinde olan sık rastlanılmayan iyi huylu lezyonlardır. Genellikle mide ve ince bağırsakta daha az olarak kolon ve özofagusta görülürler. İnflamatuar fibroid polipler erişkinlerde barsak tıkanmasına yol açan nadir benign durumlardır. Ayrıca tanısı gastrointestinal stromal tümör, ince bağırsak lenfoması ve ince bağırsak kanserlerini içerir. Biz burada intussepsiyona yol açan terminal ileumun dev inflamatuar polip olgusunu sunduk.

Anahtar sözcükler: intussepsiyon, dev, inflamatuar polip, bağırsak

Intussusception is an uncommon cause of intestinal obstruction in adults (1). Neoplasms are the most frequent cause of intussusception in the adult population. These benign tumors often are pedunculated, which results in the development of an intussusception. Malignant tumors such as carcinomas or lymphomas rarely intussuscept because they infiltrate the bowel wall and not the lumen (2). The most common initial symptoms of inflammatory fibroid tumor of small intestine are obstruction or intussusception. To date, the pathogenesis is unknown and IFPs are considered reactive and non-neoplastic lesions. The presence of platelet-derived growth factor receptor (PDGFRA) mutations questions the reactive nature of IFPs and raises the possibility of a neoplastic process (3,4). In addition, Schildhaus’s discovery has provided strong evidence of clonal proliferation and suggests a neoplastic nature of the inflammatory fibroid polyp (3).
Case presentation

A 42-year-old female presented at the emergency department with an acute abdominal pain, nausea and vomiting. The physical examination revealed rebound and tenderness especially at the right part of the abdomen. The laboratory results revealed 14,000 white blood cell count. An abdominal X-ray showed a few air-fluid levels at the right part of the abdomen. An intravenous and rectal contrast abdominal computed tomography demonstrated an ileoileal intussusception with 6 cm diameter polypoid terminal ileal mass (Figure 1). Due to the obstructive signs and symptoms a laparotomy was performed. A segmental resection of the affected small bowel segment was done. Macroscopically, the specimen was 23 cm in length terminal ileum and 11 cm diameter ileocecal segment. After opening the lumen; a solid, ivory, mucoid 5.5x4.5 cm diameter polypoid mass projecting into the lumen was found (Figure 2).

Histologically the polyp tissue showed cellular proliferation which was likely to originate from the submucosa and was composed of fibrous and edematous stroma, containing many various in size blood vessels, spindle cells and diffuse inflammatory cells infiltrate including eosinophils, plasma cells, lymphocytes, macrophages and mast cells. Immunohistochemistry using a panel of antibodies including CD34 (Clone QBEnd/10, Thermo, UK), c-kit (c-kit, Clone SP26, Thermo, UK), Bcl-2 (Moab Clone 124, Dako, Denmark), S100 (Clone 4C4.9, Thermo, UK) protein and Vimentin (Clone SP20, Thermo, UK) was performed. The spindle cells were positive for CD34 and S100, but negative for c-kit and bcl-2. The lesion demonstrated diffuse expression of vimentin (Figure 3). In the light of these findings, the final diagnosis was inflammatory fibroid polyp.

Discussion

Adult intussusception is a very rare state accounting for 1% of all adult bowel obstructions and occurs in only 5% to 16% of all intussuscepted cases (5). Intussusception occurs when a proximal segment of the bowel (intussusceptum) telescopes into an adjacent distal segment (6). The precise mechanism of intestinal intussusception remains unclear. However, it is believed that any lesion in the bowel wall within the lumen that alters normal peristaltic activity, forming leading edges for the intussusceptum, may initiate invagination (7).

Inflammatory fibroid polyps are rare polypoid lesions of the gastrointestinal tract. They are generally seen in the antral part of the stomach and the small bowel, less commonly
in the rectum and the esophagus (8). Inflammatory fibroid polyp was first described by Josef Vanek in 1949 as 'gastric submucosal granuloma with eosinophilia' and a variety of names, such as eosinophilic granuloma, hemangiopericytoma, polypoid fibroma, gastric fibroma with eosinophilic infiltration, eosinophilic gastroenteritis, polyp with eosinophilic granuloma and inflammatory pseudo tumor, all synonymous for the same lesion (9).

Although the exact pathology remains unknown, due to eosinophilic infiltration, some authors think that inflammatory polyps may develop due to an allergic background. However none of the patients reported history of an allergy. Another possibility is that due to some kind of mucosal injury or bacterial infection, the immune system exaggerate local reaction and polyps occur (9,10).

Although immunohistochemical and electron microscopic studies can not describe the histogenesis of the lesion, authors emphasize that fibroid polyp is non-neoplastic lesion. But last studies of inflammatory fibroid polyps showed an expression of platelet derived growth factor receptor (PDGFRα) and oncogenic PDGFRα mutations in a majority of analyzed tumors, which suggested that the inflammatory fibroid polyp is a neoplasm driven by activated PDGFRα (3,4). Unfortunately, we couldn't performed immunohistochemistry or molecular analysis for PDGFRα in this case.

Macroscopically it is seen as sessile or pedunculated polypoid lesion that is generally unencapsulated and whose surface is ulcerated. In our case the lesion was solitary and ulcerated. Microscopically the polyp was composed of uniform cells covered by edematous stroma and spindle-shaped fibroblasts with vascular proliferation. Fusiform cells around the vessels arranged for an 'onion like' appearance. Another important feature was an eosinophilic infiltration (10). In this case eosinophilic leukocytes were obvious but the patient did not have an allergy.

Beside histologic features which provide it allocate from eosinophilic gastroenteritis, being a mass lesion is an important parameter. IFP express Vimentin, actin and CD-34 immunohistochemically (1). Although CD-34 positivity preoccupies histogenetic relation between GIST and IFP, electron microscopic studies point out that IFP are originated from fibroblasts (11). Positive staining for CD-34
differentiates IFP from inflammatory myofibroblastic tumors, while negative staining for c-kit and Bcl-2 differentiate IFP from gastrointestinal stromal and solitary fibrous tumors (10, 11).

Preoperative diagnosis of intussusception is controversial. The diagnosis is based on medical history, physical examination and radiological modalities. The clinical symptoms depend on the location and the size of the tumoral mass. Abdominal pain is the most common symptom. An X-ray shows obstructive signs that are air-fluid levels proximal to the obstructed segment. Ultrasonography has a sensitivity of 98-100% and a accuracy of 88-89% for the diagnosis of intussusception (10, 11). In obstructed cases due to the gas in the intestines, an USG may not be so beneficial for the diagnosis. Nowadays computed tomography (CT) is considered the most sensitive technique for confirming intussusception. In reported CT cases, bowel-within-bowel configuration, a crescent of mesenteric fat, and the returning intussusceptions form three concentric layers (10).

Consequently, IFP can mimic benign and malignant lesions radiologically. Not only clinical symptoms, but also radiologic findings can distinguish IFP from malignant lesions, histopathologic verification is needed.

Conflict of interest
Authors declare that there is no conflict of interest.

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References