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A rare case of jejunal gastrointestinal stromal tumor

Gastrointestinal stromal tumors(GISTs) are rare neoplasms of the gastrointestinal(GI) system originating from the mesenchyme. GISTs mostly develop in the stomach and small intestine. We present here a case of jejunal GIST which is the rarest subtype. A 54-year-old man presented with lower right side abdominal pain. On workup, images showed a 7cm solid-cycstic lesion adjacent to ascending colon. On surgical exploration, a large jejunal tumor en bloc resected and jejuno-jejunal primary anastomosis was performed. Pathologic results showed a 9cm jejunal GIST with 5% proliferation index. Immunohistochemistry results demonstrated high expressions of CD117, whereas CD34 negative. The patient was discharged uneventfully. GISTs should be considered in patients with abdominal pain. The mainstay treatment of the jejunal GIST is complete surgical resection. The definitive diagnosis of GISTs is by immunohistochemical stains.

KEY WORDS: GIST, Stromal tumor, Jejunum

Introduction

Mazur and Clark in 1983 was described gastrointestinal stromal tumors(GISTs) as a rare mesenchymal tumors of the gastrointestinal system [1]. They correspond to 0.3 to 0.5% of all GI tumors. However, they can be found anywhere along the alimentary tract, mostly develop in the stomach and small intestine [2]. In this case report, we present a jejunal GIST with abdominal pain. On the other hand, we will discuss the diagnostic tools and the management of GISTs.

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Case Report

A-54-year old man investigated over 3 years for abdominal pain. A physical examination was significant for mild tenderness on the right side of the abdomen. His past surgical history was positive for right inguinal hernia repair. Abdominal ultrasound revealed a 7cm solid-cystic lesion in the right lower abdomen. Magnetic resonance imaging(MRI) and computed tomography(CT) demonstrated 7x7.5 cm mass filling the right side of the abdomen and communicating with ascending colon (Fig. 1).
When the patient was taken for laparotomy, there was a 9 cm tumor arising from anti-mesenteric side of the mid jejunum (Fig. 2).
The large jejunal tumor resected en bloc and end-to-end jejuno-jejunal anastomosis was performed. Pathology demonstrated a CD 117 positive jejunal gastrointestinal tumor with negative margins (Fig. 3).
Macroscopic size of the tumor was 9x7x6 cm in diameter. The tumor showed hemorrhage, but necrosis was not found. On histologic examination, the tumor showed expansive growth pattern. Furthermore, the tumor had low risk according to Fletcher's criteria with mitotic rate 3 per 50 per higher field (HPF), composed of epithelioid and spindle cells (Fig. 4).

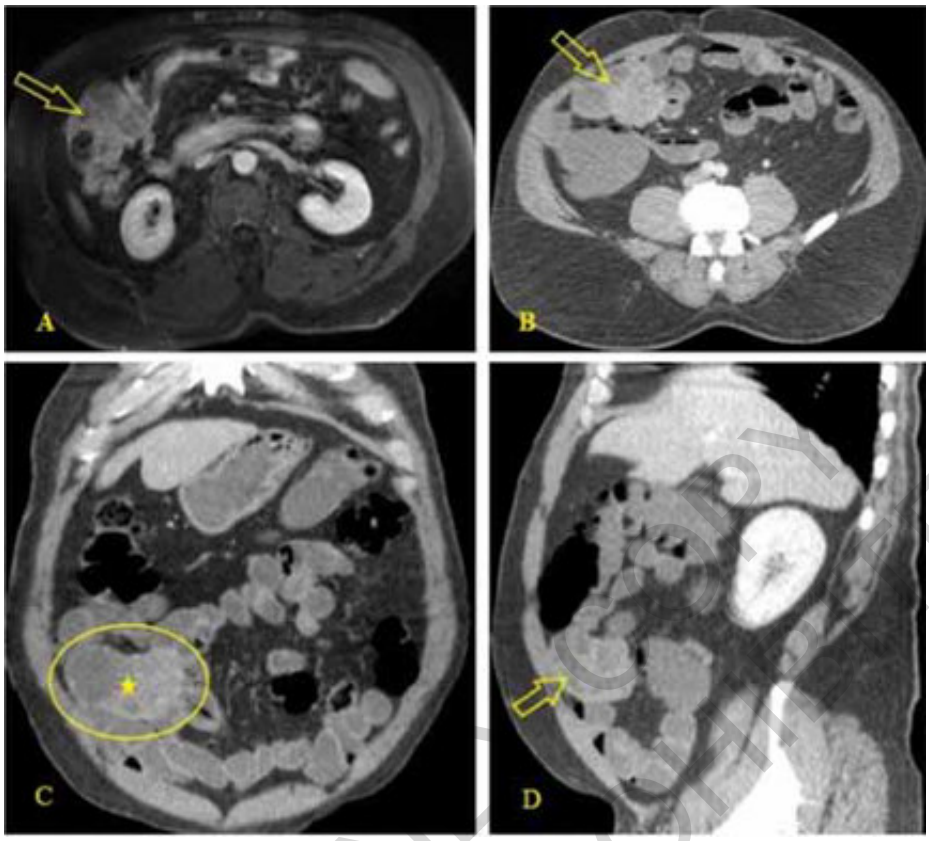

Fig. 1: Preoperative imaging of jejunal GIST. Axial image in abdomen MRI (A) and in abdomen CT (B) shows the heterogeneous lesion (arrow). Coronal (C) and sagittal (D) view in abdomen CT with the tumor. Asterisk in centrum of circle (C) and arrow (D) shows the lesion.

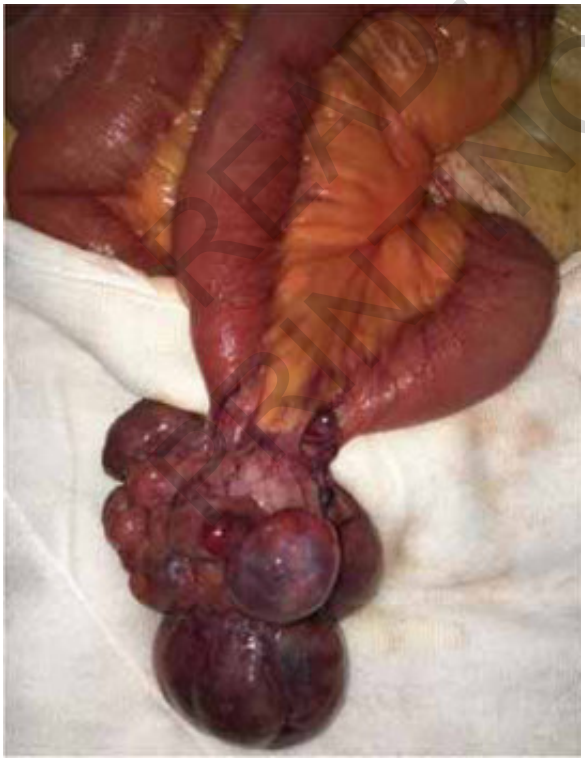

Fig. 2: Intraoperative picture of the mass (9x7x6 cm)

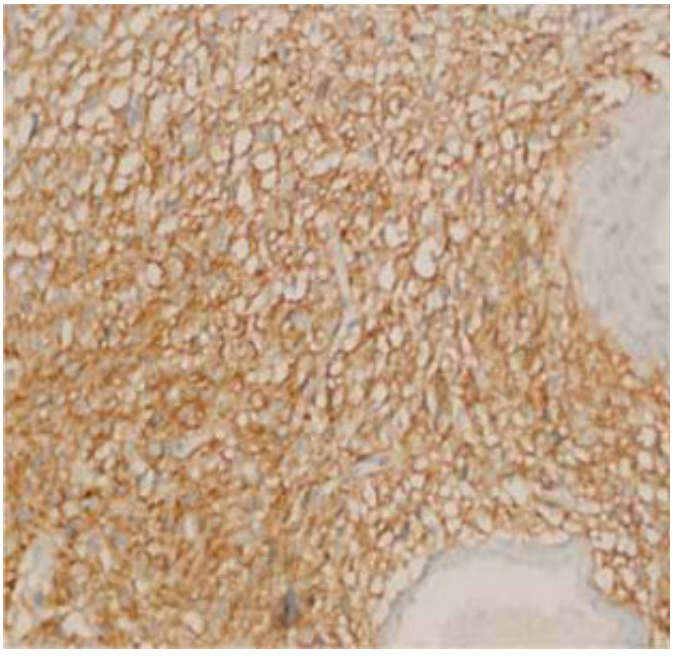

Fig. 3: Strong and diffuse expression of CD 117 (x40)

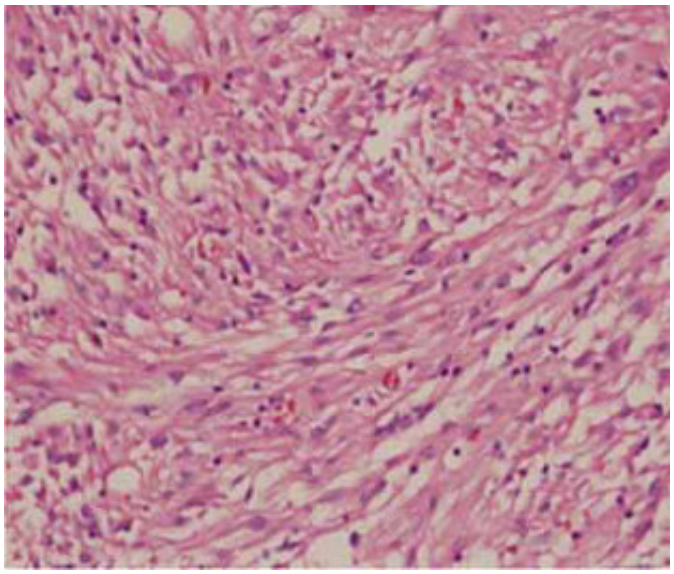

Fig. 4: Spindle cells forming short bundles (H&Ex40).

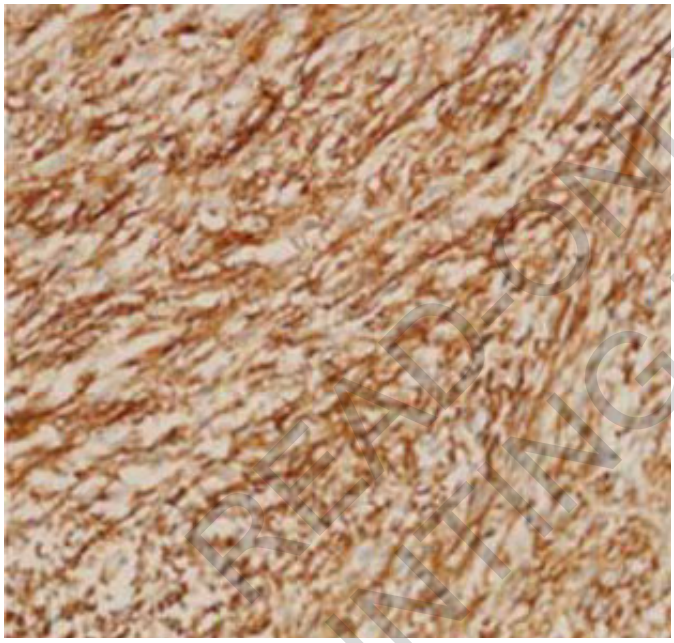

Fig. 5: Diffuse and strong cytoplasmic positive reaction with DOG1 (x40).

CD 34, S100 and Desmin negative, on the other hand DOG1 positive (Fig. 5) and α-smooth muscle actin (SMA) focally positive in the immunohistochemistry staining. Ki-67 proliferation index was %5. The patient was discharged uneventfully.

Discussion

GIST is the most common sarcoma of the alimentary tract that usually occurs in adults over 40 years old. It is equally observed in women and men. Only a few studies reported a slightly higher incidence of males [3]. Jejunal GISTs with small diameter are usually asymptomatic. As it grows, it may occur with signs of GI bleeding or obstruction [4].

CT scans identify 87% and MRI scans nearly 100% of GISTs. The definitive diagnosis is made by histopathology and immunochemistry stains.

Immunohistochemistry based on proto-oncogen c-kit(CD 117) (an epitope of the KIT receptor tyrosine kinase) and CD 34 positivity. Nearly 95% are positive for c-kit mutation(CD 117). GISTs are mostly originating from the interstitial cell of Cajal. Tumor morphology of GISTs contains mostly spindle cells(70%) and epithelioid cells(%20) or mixed spindle and epithelioid cells(10%), respectively. Such as leiomyomas, leiomyoblastomas and leiomyosarcomas may show similar morphologic features. For this reason, both immunohistochemistry staining and morphologic histology should be done for the final diagnosis [4,5]. In this study, the tumor has high expressions of CD 117, whereas CD 34 is negative.

Han IW et al. retrospectively analyzed 101 consecutive patients with GIST located in duodenum or small intestine. They have found ≥ 60 years, advanced tumor(T) size (≥ 10cm), high mitotic rate (>5 per 50 HPF), presence of epithelioid cell morphology, presence of tumor rupture and/or necrosis as poor prognostic factors [6].

We present a 54-year old man with jejunal GIST. Our case histologically composed of mixed spindle and epithelioid cells. In this case, mitotic rate is 3 per 50 HPF and necrosis is not found. In addition, the tumor size is 9 cm in diameter (T<10cm). According to the literature our case does not have poor prognostic factors. In 2006, Miettinen&Lasota established a new risk classification for gastric and small intestine GISTs. This classification is based on tumor size and mitotic rate. According to the classification, pathologic result shows intermediate risk (group 3a) for this case [7].

Ki-67 index, as well as other parameters, is one of the most prognostic tool for GIST recurrence and important for assessment of malignant potential of disease [5]. There was no relationship between mitotic rate and Ki-67 index. In this case, the patient has low proliferative index based on Ki-67 expression (5%).

Sunkey RE et al. presented a 70-year old male with small bowel obstruction. Emergency small bowel resection was performed. Similarly with our case, the tumor composed both spindle and epithelioid cells and CD 117, DOG1 and SMA was positive in immunohistochemistry examination [4].

Manxhuka Kerliu-S et al. reported a 30-year old woman in their study [5]. The tumor was detected in her duodenojejunum, resected en bloc and performed primary anastomosis. The tumor size was 10 cm in diameter, composed of spindle and epithelioid cells. Mitotic rate >5 per 50HPF, Ki-67 expression was 10% which means

low proliferative index. CD 117 and CD 34 were positive, Desmin and S100 were negative. Taking into consideration, poor prognostic factors was found such as high mitotic index, size of the tumor(10cm) in their case.
Interestingly, Yuval BJ et al. demonstrated a 64-year old women presented with active GI bleeding. Gastroscopy and colonoscopy was normal. CT angiography revealed a hypervascular mass in the jejunum and distal coil embolization was performed. After embolization the patient underwent laparoscopic resection and primary anastomosis. Selective angiography is important for finding the localization of bleeding and allows treatment at the same time [2].
Complete surgical resection is the mainstay treatment modality for jejunal GISTs. Although there is no evidence for optimal resection margins size in the literature, it is crucial to ensure negative margins. 5-year survival after complete resection low-to-intermediate risk GISTs is 95% [4]. Therefore, R0 and minimal resection are essential and sufficient [8].

Conclusion

Jejunal GISTs are mostly asymptomatic but, if the patient has non-specific abdominal pain, GI bleeding and obstructive symptoms we should consider GISTs. The mainstay treatment of jejunal GISTs is complete surgical resection. Immunohistochemical staining and morphology is vital for the final diagnosis of GISTs.

Riassunto

I tumori stromali gastrointestinali (GIST) sono rare neoplasie del sistema gastroenterico (GI) originate dal mesenchima. I GIST si sviluppano principalmente nello stomaco e nell'intestino tenue. Presentiamo qui un caso di GIST digiunale che è il sottotipo più raro. Un uomo di 54 anni presentava dolore addominale nella parte inferiore destra. Allo studio le immagini hanno mostrato una lesione cistica solida di 7 cm adiacente al colon ascendente. All'esplorazione chirurgica, è stato resecato un grande tumore digiunale in blocco e è stata eseguita l'anastomosi primaria digiuno-digiunale. I risultati istopatologici hanno mostrato un GIST di 9 cm con un indice di proliferazione del 5%. I risultati di immunoistochimica hanno dimostrato alte espressioni di CD117, mentre CD34 negativo. Il paziente è stato dimesso senza incidenti. I GIST devono essere considerati in pazienti con dolore addominale. Il trattamento principale del GIST digiunale è la resezione chirurgica completa. La diagnosi definitiva di GIST è mediante colorazioni immunoistochimiche.

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