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Case report

Intestinal submucous fibrovascular hamartoma: A case report ☆,☆☆,★

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ABSTRACT

Intestinal submucosal fibrovascular hamartoma is considered as a rare intestinal lesion. We present the case of a 63-year-old female with abdominal symptoms, bleeding, and increased serum tumor markers. The abdominal ultrasound revealed that the left abdominal intestinal wall and mesentery were thickened with enlarged multiple lymph nodes, suggesting intestinal obstruction. Other imaging findings confirmed the ultrasound findings. Histopathology of the removed lesion provided the diagnosis of intestinal submucosal fibrovascular hamartoma with hemorrhage, inflammation, and amyloidosis. Intestinal submucosal fibrovascular hamartoma is a hemorrhagic lesion with macroscopic tumor due to the abnormal mixing of the organ's normal components, which still remains a challenge for clinicians and pathologists. We consider routine abdominal ultrasonography and contrast-enhanced ultrasonography (SICUS) to be safe and effective in the diagnosis of intestinal neoplastic lesions.

Introduction

Intestinal submucosal fibrovascular hamartoma is a benign lesion induced by the abnormal proliferation of muscles, nerves, and vascular elements in small intestine, which was first reported in 1982 by Fernando and Mc Govern [1]. Patients with this disease usually have abdominal pain, obstruction, occult gastrointestinal bleeding and iron deficiency anemia. The lesion consists of a cluttered mass of benign primary cells leading to submucosal dysplasia of the muscles, nerves, and vascular components (hemangioma). It affects the intestine's variable length, resulting in stenosis, for which surgical resection is the curable treatment method [2].

Abbreviations: SICUS, small intestine contrast-enhanced ultrasonography; CT, computed tomography.

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★ Patient Consent: Written informed consent was obtained from the patient for publication of the case details and accompanying images.

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Furthermore, a wide range of chronic reactive diseases exhibits histological features similar to intestinal submucosal fibrovascular hamartoma. Some researchers claimed that this lesion is not truly hamartoma, but a "burnt-out" phase of Crohn's disease or other chronic inflammatory processes [3–5]. The proliferation may be related to diaphragm disease, radiation enteritis, ischemic enteritis, and Crohn's disease.

Intestinal submucosal fibrovascular hamartoma is a rare intestinal hemorrhagic lesion with occult gastrointestinal bleeding as main symptom, which still remains a challenge for clinicians and pathologists [6]. Small intestine contrast-enhanced ultrasonography (SICUS) is the optimal method for diagnosing intestinal submucosal fibrovascular hamartoma with features of high accuracy, excellent tolerance, and radiation-free [7].

Case presentation

A 63-year-old female was admitted to the hospital with complaints of abdominal distension, abdominal pain, and hematemesis. On admission, the patient underwent a physical examination, high-resolution computed tomography (HRCT), routine blood test, fecal occult blood test, coagulation test, renal function test, and liver function test. The physical examination was as follows: temperature 36.6°C, pulse 75 beats/min, respiration 15 beats and/or min, and blood pressure 80/55 mmHg. The patient had abdominal distension for two years without obvious inducement, systematic diagnosis or treatment. The patient had severe abdominal pain with hematemesis and hematochezia for 10 days. A palpable mass was detected in the left lower abdomen, while the liver and spleen were not palpable. She had no medical history of taking non-steroidal anti-inflammatory drugs (NSAIDs) or radiation exposure. The results of the various tests performed were as follows: blood cells: white blood cells(WBC): 3.80(4.0˜10.0) × 10^9/L,neutrophil(NE)%:62.0(50˜70)%, hemoglobin(HGB):93(110˜150)g, platelet (PLT): 181(100˜300) × 10^9/L; liver function: total protein (TP): 51.3(60˜80)g/L, albumin (ALB): 25.2(40˜55)g/L; serum tumor marker cancer antigen (CA)12–5:39.40(<35)U/mL; the occult blood test was weakly positive; glutamic acid (GLU): 4.59(3.89˜6.1)mmol/L; coagulation tests: prothrombin time (PT): 15.2(11.0˜13.0)s,prothrombin time activity (PTA):76(75˜100)%, international normalized ratio (INR):1.18(0.8˜1.5). The abdominal ultrasound scan revealed thickening of the left abdominal intestine wall, and the mesentery was thickened with enlarged multiple lymph nodes, suggesting intestinal obstruction (Fig. 1). The enhanced CT scan of the small intestine suggested that inflammatory lesions likely occur in stomach and small intestine owing to incomplete obstruction of the small intestine and thickening of the intestinal wall in the left iliac fossa (Fig. 2). A barium enema examination suggested the presence of intestinal obstruction (Fig. 3). Before surgery, the patient was diagnosed with obstructive space-occupying lesions in the small intestine.

The partial resection of the small intestine was performed under general anesthesia. During the operation, the obvious

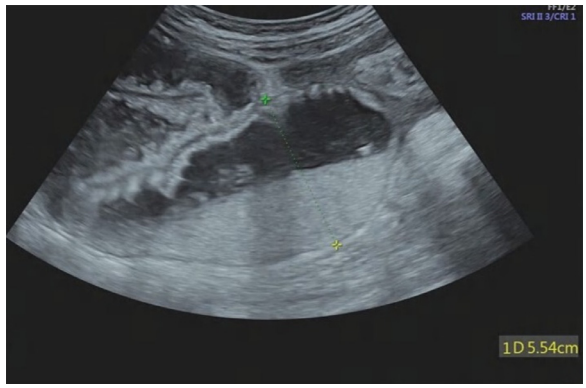

Fig. 1 – The intestinal dilatation is observed in the left upper abdomen with a maximum width of approximately 5.5 cm. Liquid contents are seen in the intestine, and reverse peristalsis is observed. The wall thickness is about 0.28 cm with a clear layer.

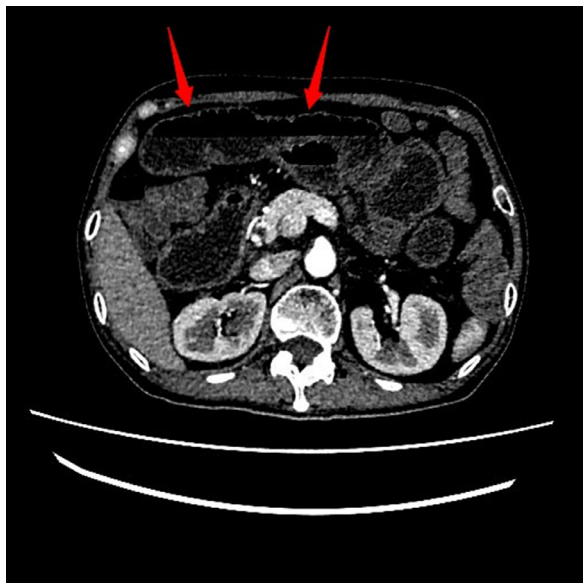

Fig. 2 – The diffuse edema and thickening of the small intestine wall, local effusion, and dilatation of the intestinal cavity with the plane shadow of short gas fluid, and local thickening of the small intestine wall with the obvious enhancement of the left iliac fossa is observed. A bag-shaped gas shadow (red arrow) can be seen in the descending part of the duodenum. (Color version of figure is available online)

dilatation and congestion of the small intestine were located 150cm underneath Treitz ligament. The intestinal canal was stiff, and adhered to the left abdominal wall. Hemorrhagic effusion of the mesentery and surface of the small intestine were found, and the severely damaged intestinal canal, mesentery and necrotic small intestinal canal about 100cm in length were removed. After the operation, the patient was diagnosed with small intestinal space-occupying lesions or inflammatory stenosis with obstructive changes. Intestinal decompression indicated that the small intestine was filled with

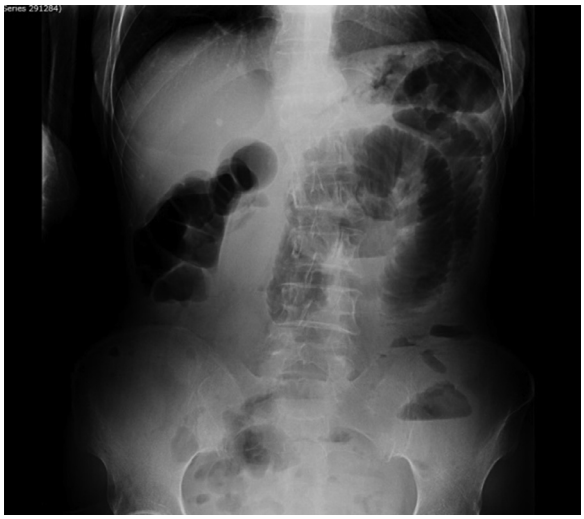

Fig. 3 – The surfaces of both diaphragms are smooth, no free gas shadow is found under the diaphragm; the stomach bubble shadow is observed under the left diaphragm; the intestinal tube of the left upper abdomen is slightly expanded, and the gas-liquid plane shadow is detected inside.

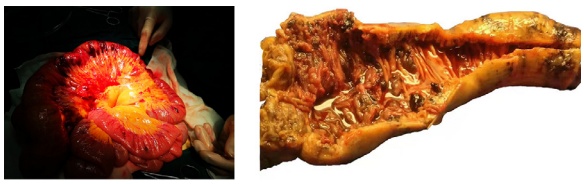

Fig. 4 – (A) Hematodes exudate on the surface of the mesentery and small intestine. (B) The resected intestine is approximately 35 cm in length. The gray-red cauliflower-shaped masses with an unclear boundary and a diameter of 5 cm can be observed at 18 cm from one side of the broken end. The intestinal wall is covered with polyps after the intestinal tube is incised. (Color version of figure is available online)

approximately 1500 mL of the fluid containing blood. The patient developed symptoms of severe anemia (Hb<70g/L) during the operation, and the blood transfusion was performed.

Considering the pathological findings, a segment of the resected intestine approximately 35 cm in length was selected. The gray-red cauliflower shaped masses with an unclear boundary and a diameter of 5 cm were observed at 18 cm from the broken end. The intestinal wall was covered with polyps after the incision of the intestinal tube (Fig. 4). Microscopically, intestinal submucosal fibrous tissue and vascular hyperplasia, local lumen dilatation and hyperemia, local hemorrhage with inflammatory cell infiltration, and amyloidosis could be observed (Fig. 5). Immunohistochemistry revealed CK (epithelium +), CD3 (scattered +), CD20 (scattered +), Pax-5 (scattered +), Bcl-2 (scattered focal +), CD21 (–), CD10 (scattered +), CD23 (–), CD5 (scattered +), Cyclin D1 (–), CD56 (–), CD4 (scattered +), CD8 (scattered +), CD30 (–), Ki-67 (30% +), EBV (–) CD138 (scattered +), CD38 (diffused +), Lambda (scattered +), and kappa (diffused +). The complementary staining and special staining: SMA (+) Congo red (+) HMB45 (–) Actin (–) CD34 (–) CD117 (–) Dog1 (–) S100 (–). The patient was diagnosed with intestinal submucosal fibrovascular hamartoma, local bleeding, inflammation, and amyloidosis.

The patient was followed up for half a year. Abdominal CT and X-ray showed small intestinal obstruction, blood routine displayed long-term anemia, and cardiovascular ultrasound revealed different degrees of deep vein thrombosis in upper and lower limbs.

Discussion

Intestinal submucosal fibrovascular hamartoma is a rare lesion of the small intestine (usually locates in the jejunum or ileum). The clinical symptoms include non-specific abdominal pain, recurrent obstructive symptoms and occult gastrointestinal bleeding. Intestinal submucosal fibrovascular hamartoma can occur as single or multiple stenoses or polypoid masses [3,4], resulting in changes in the intestinal length. Radiologically, the affected intestines may exhibit the same manifestations as stenosis, intussusception, or polypoid masses. Distinguishing intestinal submucosal fibrovascular hamartoma with a series of chronic inflammatory bowel diseases is difficult, which were identified as the cause of chronic intestinal obstruction and gastrointestinal bleeding in this study.

Intestinal submucosal fibrovascular hamartoma is a benign non-epithelial hamartoma which proliferates in mature submucosa, causing tissue disorder, usually in the small intestine. These lesions were characterized by random, abnormal smooth muscle bundles from the mucosal muscle layer, unmyelinated nerve fiber bundles with scattered abnormal ganglion cells, and angiomatous vascular channels [1,3]. Considering that many features of intestinal submucosal fibrovascular hamartoma could been observed in a range of reactive diseases, including Crohn's disease, ischemic enteritis, radiation enteritis, and small intestinal stenosis caused by non-steroidal anti-inflammatory drugs ("diaphragm disease"), there is no consensus on the nature of the hamartoma [3,5, 8]. Some researchers pointed out that the intestinal submucosal fibrovascular hamartoma isn't an independent tumor but represents a "burnt out" phase of Crohn's disease or other chronic inflammatory processes [3–5].

Crohn's disease features diverse and non-specific pathological changes, which are characterized by transmural chronic inflammation, non-caseous granuloma, and lymphatic accumulation in the intestinal wall. Moreover, typical mucosal manifestations include cobblestone fissures and edematous mucosa. Neuromuscular hypertrophy, submucosal fibrosis, and serous fibrosis are common manifestations in the advanced stage of Crohn's disease [3]. Occasionally, the transmural inflammation and hiatus described by a typical Crohn's disease may not exist, and connective tissue changes (such as fibrosis, muscle hypertrophy, neurovascular changes) are residues of the chronic "burnt out" process.

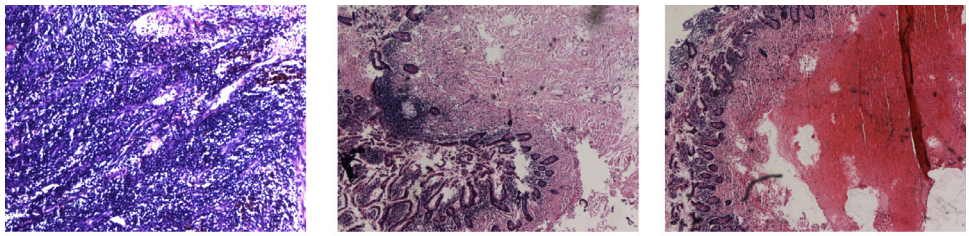

Fig. 5 – Pathological image (HE staining × 100) A and pathological image (HE staining × 40) B: intestinal submucosal fibrous tissue and vascular hyperplasia, local lumen dilatation and congestion, local hemorrhage with inflammatory cell infiltration, and amyloidosis (black arrow). Fig. 5C: immunohistochemical and special staining results: CK (epithelium +), CD3 (scattered +), CD20 (scattered +), Pax-5 (scattered +), Bcl-2 (scattered focal +), CD21 (–), CD10 (scattered +), CD23 (–), CD5 (scattered +), Cyclin D1 (–), CD56 (–), CD4 (scattered +), CD8 (scattered +), CD30 (–), Ki-67 (30% +), EBV (–), CD138 (scattered +) CD38 (diffused +), Lambda (scattered +), kappa (diffused +) SMA (+), Congo red (+) HMB45 (–), Actin (–), CD34 (–), CD117 (–), Dog1 (–), and S100 (–)

Intestinal submucosal fibrovascular hamartoma is a rare lesion, with its presence as an obvious hamartoma lesion or a non-specific response phenomenon remaining controversial. Undoubtedly, intestinal submucosal fibrovascular hamartoma exhibits a macroscopic "tumor" pattern with severe bleeding and obstruction as clinical manifestations due to abnormal mixing of normal intrinsic tissue components. At the initial stage, the lesion formed by hamartoma mass was considered as a neoplastic lesion. However, tumor lesions are extremely rare in the small intestine (2%), with malformations constituting the major part [9]. Therefore, vascular hyperplasia continues to be the most common cause of severe bleeding and obstruction. Intestinal submucosal fibrovascular hamartoma has certain characteristics similar to the common symptoms of inflammatory bowel disease and other reactive diseases; nevertheless, inadequate as indicators of these diseases.Therefore, submucosal fibrovascular hamartoma should be considered in the differential diagnosis of small intestinal stenosis and giant lesions when radiation or ischemic enteritis is clinically suspected while clear histological features of Crohn's disease or diaphragmatic myopathy is inadequate.

In view of the clinical symptoms and pathological changes of intestinal submucosal fibrovascular hamartoma, we put forward some suggestions for the treatment of this case: anti infection; anticoagulant therapy (including oral medication and upper arm vein puncture catheter anticoagulant therapy); parenteral nutrition; follow-up of disease changes, close attention to abdominal signs and imaging examination results. In addition, early recognition and treatment are critical to improve the prognosis of the disease, so it is extremely important to choose the appropriate and efficient diagnosis technology.

As a non-invasive imaging tool, the transabdominal ultrasound is increasingly preferred, which is superior to small-bowel follow-through (SBFT) for the detection of intestinal diseases, with higher cost-effectiveness and enhanced tolerance than magnetic resonance imaging, with minimal radiation exposure as compared to CT images. However, the traditional transabdominal ultrasound is often limited by intracavitary gas and intestinal wall collapse, which may increase the difficulty of diagnosis. Oral contrast medium before the operation can promote the dilatation of the intestinal loop and improve the display of the intestinal wall. Therefore, SICUS is emerging as an alternative to the traditional ultrasound in diagnosing and monitoring intestinal submucosal fibrovascular hamartoma [10,11].

The oral anechoic contrast agent can effectively increase the sensitivity from 91.4% to 96.1% in the diagnosis of intestinal submucosal fibrovascular hamartoma [10]. Compared with conventional ultrasound, SICUS is characterized by a higher accuracy in detecting stenosis and determining the degree of involvement of the small intestine. Moreover, SICUS exhibits a comparable diagnostic rate of conventional enteritis with that of SBFT and CT (k coefficient is 0.88 and 0.91, respectively). Furthermore, there is a substantial consistency between SICUS and ileo-colonoscopy or surgery ($k = 0.62$) [12].

The preliminary data suggested that power Doppler imaging played a critical role in improving the diagnostic accuracy of conventional ultrasonography and SICUS. Moreover, power Doppler ultrasonography is effective in evaluating blood vessels of intestinal wall in inflammation related diseases as while as distinguishing inflammatory stenosis from fibrotic stenosis [12-13].

In conclusion, SICUS can be used as an accurate imaging tool for the evaluation of intestinal submucosal fibrovascular hamartoma with acceptable tolerance and no radiation. The limitation of SICUS is owing to the difference between observers and being a dynamic process, interpretation and comparison of images retroactively are challenging. The diagnostic accuracy of SICUS generally depends on the operator's clinical experience. Collectively, it is imperative to conduct a multicenter prospective study for comparing the results of magnetic resonance enterography with SICUS in such patients [14].

Ethics approval and consent to participate

Not applicable.

Availability of data and materials

All data are available and stored by the authors.

Authors' contributions

Yanjun Liu analyzed and interpreted the patient data regarding the intestinal submucosal fibrovascular hamartoma. Jin Liu was a major contributor in writing the manuscript. Ziyao Ji is responsible for proofreading the article. All authors read and approved the final manuscript.

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