Polysplenia syndrome with preduodenal portal vein

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Polysplenia syndrome is a heterogeneous disease that primarily affects the asymmetric organs, including the heart, lungs and bronchi, liver, intestines, and spleen [1]. It manifests mainly in childhood, 40% of the patients reach 2 years of age and the majority dies before 5 years of the age due to cardiac anomalies [2]. 5-10% of the patients lack cardiac involvement, which allows them to reach adulthood [3]. The precise etiology of polysplenia is unknown. Embryonic, genetic and teratogenic components have all been implicated as causative factors in polysplenia [4]. Although polysplenia syndrome has a wide range of abnormalities, there is no single pathognomonic abnormality that characterizes this rare entity. The range of anomalies include multiple spleens of equal volume, visceral heterotaxia, right-sided stomach, a left-sided or large midline liver, malrotation of the intestine, a short pancreas, preduodenal portal vein and inferior vena cava anomalies [5].

We report a 50-year-old female presented to our outpatient department with chief complaints of right upper quadrant pain for the last 3 months. At a local hospital, she was noted to have polysplenia and cholelithiasis. She was referred to our institution for further evaluation and treatment. An abdominal computed tomography (CT) and ultrasonography showed cholelithiasis with polysplenia, the portal vein was located anterior to the duodenum and there was associated malrotation of gut (Fig. 1A). The surgical procedure for cholelithiasis began with a thorough exploration of the abdomen. The portal vein was detected in front of the first part of the duodenum (Fig. 1B). The gallbladder was hugely distended with a 2 cm stone impacted at the neck. The common bile duct was located posterior to the portal vein. The presence of multiple small spleen and one normal size spleen was confirmed on the left side of the upper abdomen. There was malrotation of the gut with the entire right colon located in the left upper quadrant along with left the colon in normal position. The pancreas was short with deficient body and tail. The gallbladder was opened at the fundus, stone removed, cystic artery and duct identified, ligated and cut between ligature and gallbladder removed through liver bed. Ladd’s procedure was added to correct malrotation of gut. The postoperative course was uneventful, and the patient was discharged on 5th postoperative day.

Reports indicate that most cases of preduodenal portal vein (PDPV) in adults involve surgery for cholelithiasis leading to the hypothesis that PDPV may be responsible for the formation of gallstones due to chronic compression of the common bile duct by the portal vein, leading to stasis of bile. A similar opinion was also expressed by Seo et al [6] and Low et al [7]. When surgery is required, care must be exercised, especially for procedures involving the upper abdomen. If PDPV is not detected prior to surgery, it can cause severe complications, such as hemorrhage and vascular ligation [8]. Such accidents can be prevented by performing careful diagnostic imaging.

Figure 1 (A) Contrast-enhanced computed tomography showing the preduodenal vein (long black arrow) traversing the duodenum anteriorly and abnormally coursing towards fissure for ligamentum teres. Multiple spleens (short black arrows) are present in left hypochondrim (small arrows). Gallbladder wall is thickened and shows intraluminal hyperdense focus (calculus). Superior mesenteric vein is on the left side (star) and jejunal loops are not present in usual left hypochondrim consistent with malrotation of the gut. (B) Intraoperative photograph showing the portal vein (arrow) crossing the duodenum anteriorly.
in advance, such as CT, and especially noting the possibility of PDVP in cases of polysplenia syndrome [8].

References


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Acute massive gastric dilation: an unusual presentation of metastatic urinary bladder cancer

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Acute massive gastric dilation represents the extreme form of acute gastric dilation (AGD). Obstruction of distal duodenum can cause AGD with bilious nasogastric drainage. Most common causes are pancreatic neoplasms and pseudocysts, annular pancreas, superior mesenteric artery syndrome, retroperitoneal fibrosis and tumors, duodenal neoplasms and hematoma [1]. AGD complicated with gastric perforation, severe gastritis and abdominal compartment syndrome constitutes a surgical emergency. However, in the setting of non-complicated AGD, gastric decompression, parenteral nutrition, antibiotics and appropriate treatment of associated diseases may be sufficient for several days before undertaking repair [2]. Herein, we present a case of urinary bladder carcinoma that had advanced in the retroperitoneum causing distal duodenal obstruction and massive uncomplicated AGD.

A 78-year-old male presented with acute abdominal pain and a progressively distended abdomen. Direct questioning revealed a 3-month history of abdominal discomfort, weight loss of 22 kg and 2 episodes of hematuria. The only relevant past medical history was type 2 diabetes mellitus treated with vildagliptin/metformin for 3 years.

On admission, inspection revealed a massively distended abdomen. A succussion splash in the upper abdomen and diffuse tenderness were present. Blood pressure was 85/50 mmHg, heart rate 125 beats/min, SaO2 88% on air and axillary temperature 37.6°C. Laboratory tests were remarkable for WBC count 14100/mm3, absolute neutrophil count 12150/mm3, hemoglobin 12.6 g/dL and creatinine 2.4 mg/dL.

A nasogastric tube was placed without difficulty. The impressive volume of 9.5 L of bilious material was withdrawn. After gastric decompression the patient became hemodynamically stable. Clinical, laboratory findings and electrocardiographic abnormalities (sinus tachycardia with T-wave inversion in leads V1–V2) improved. Abdominal x-rays showed no findings of pneumoperitoneum. Intra-abdominal pressure was 12 cmH2O (grade I intra-abdominal hypertension according to the Burch system). Upper GI images with Gastrograin® showed a massively dilated stomach, no extravasation, normal pyloric channel and an abrupt stricture at distal duodenum. A contrast-enhanced CT scan revealed: a) an enhanced mass along the left lateral bladder wall with obstruction of the left ureteric orifice and invasion to the perivesical fat; b) para-aortic, para-caval, retrocrural lymphadenopathy; c) an infiltrative mass in the retroperitoneum causing obstruction of the third portion of duodenum (Fig. 1); and d) multiple pulmonary metastases in lower zones (T3b/N3/M1 disease). Cystoscopy revealed a solid bladder tumor (poorly differentiated transitional cell carcinoma), with obstruction of the left ureteric orifice and diffuse diverticula. Bilateral ureteral stents were placed. Unfortunately on day 2, the patient developed an episode of atrial fibrillation, which failed several attempts of cardioversion and he finally expired.

In summary, the advanced stage of the primary urinary bladder cancer prompted the diagnosis of metastatic retroperitoneal tumor with infiltration of distal duodenum. Several cases of massive AGD have been described worldwide but to our knowledge this is the second case due to bladder cancer metastasis to the duodenum [3]. What is interesting in our case is that the stomach was massively distended; however no perforation occurred. To our knowledge this is the first case reported in which 9.5 L of gastric fluid were initially aspirated without gastric rupture [4]. Another remarkable fact is that our
patient was unable to vomit. Although emesis is the dominant symptom, inability to vomit has also been reported and can be explained by occlusion of the gastroesophageal junction by the distended fundus, which angulates the esophagus against the right crus of the diaphragm [5].

References


Polypoid and hyperplastic heterotopic gastric mucosa in the jejunum as a cause of recurrent subocclusive episodes

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Gastric heterotopia occurs throughout the entire gastrointestinal tract, from the oral cavity to the anorectum, and also involves the gallbladder, biliary tract, umbilicus and scrotum [1-10]. The presence of this lesion beyond the ligament of Treitz with recurrent intestinal subocclusive episodes is uncommon [1-10].

We report a case of a 21-year-old woman who had a one-year history of intermittent hypogastric abdominal pain, vomiting and nausea. A physical examination revealed abdominal tenderness with reduced bowel sounds. An abdominal X-ray and a CT scan showed gastric and proximal small bowel distention with multiple air-fluid levels.

An abdominal laparotomy was performed with the following findings: small bowel adhesions and the presence of a large intraluminal tumor affecting the jejunum. The tumor was totally resected.

Upon a macroscopic examination, the specimen was a 25-cm segment of the jejunum containing a large and soft polypoid mass of 15 cm length (Fig. 1). A histological examination revealed that the tumor consisted of gastric type epithelium with hyperplastic foveola and oxyntic glands covered by parietal, chief and neuroendocrine cells.

To analyze cell cycle proteins we counted positive cells per 100 epithelial cells in five randomly selected microscopic fields. p27 was positive in 97% of foveolar cells, in 68% of mucous neck cells and in 20% of glandular cells. p21 was positive in 82% of foveolar cells, in 10% of mucous neck cells and in 2% of glandular cells. p16 and p57 were negative. Cyclin D1 was positive in 87% of foveolar cells, in 75% of mucous neck cells and in 2% of glandular cells. Ki67 was positive in 20% of foveolar cells and 99% of mucous neck cells and was negative in glandular cells.

Several hypotheses have been suggested to explain the origin of gastric heterotopia. Wacrenier et al [4] and Soule [5] believed that gastric heterotopia arose from the epithelium of the primitive gut, which was separated from the primordial stomach and underwent hyperplasia over time due to unknown pathways. Skandalakis et al [6] proposed that heterotopic gastric mucosa originated from the metaplasia of pluripotent endodermal cells of the foregut. Abel et al [7] proposed that this lesion was of vitellointestinal tract origin. Other authors proposed the ability of endodermal cells of the primitive gut throughout the gastrointestinal tract to differentiate and undergo hyperplasia or physical movement of the gastric epithelium due to unknown pathways [2,7,8].
In the immunohistochemical analysis of cell cycle molecule expression, we showed that p21, p27 and cyclin D1 were highly positive on the foveolar surface. In the neck, a site of cellular replication, p21 was low, and p27 and cyclin D1 were high; p16 and p57 were negative throughout the gastric mucosa. Ki67 was positive in the neck zone, with few positive cells in the foveolar region.

We propose that heterotopic gastric mucosa is associated with an alteration in the expression of cell cycle molecules (the Cip/Kip family), stimulated by unknown growth factors that induce high cellular proliferation.

References


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Ulcerative colitis following diagnosis and successful cure of Hodgkin’s disease: description of a case

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Hodgkin’s disease (HD) is a malignant neoplasm of lymphoreticular cell origin, characterized by the presence of large mononucleated Hodgkin and giant multinucleated Reed-Sternberg cells [1]. Although it represents the most common subtype of malignant lymphoma in young people it has very
rarely been described in combination with ulcerative colitis (UC). Basic-Jukic et al described a patient who had suffered from relapsing UC for 6 years before he developed HD [2]. Vieites et al described the case of a patient with UC who developed a cutaneous Hodgkin-type lymphoproliferative lesion 6 months after the initiation of treatment with anti-tumor necrosis factor-α agent [3]. Beaugerie et al found only one case combining UC and HD among 19,486 patients with inflammatory bowel disease (IBD) [4].

All cases of HD described so far, appeared after the establishment of the diagnosis of UC. However, HD preceding the diagnosis of UC has never been described. We report hereinafter the case of a young patient who developed left-sided UC, 3 years after the diagnosis and successful treatment of HD. A woman, aged 35, was admitted to our department in 2011 for investigation of bloody diarrhea. From her past history she mentioned diagnosis of HD (stage Ila) in 2008. From 21/11/2008 to 04/03/2009 she was submitted to 4 cycles of chemotherapy, consisting of adriamycin, bleomycin, vinblastin and dacarbazine followed by 14 courses of radiation therapy. The response was excellent and the disease was considered to be completely healed. UC started 5 weeks before admission with 2-4 bowel movements/d accompanied by abdominal pain. Laboratory investigation was compatible with left-sided UC of mild-to-moderate degree. She responded well to the combined per os and rectal administration of mesalamine. A new flare-up of moderate degree appeared in March 2012 which was settled promptly with oral administration of predniizolone and rectal and oral administration of mesalamine. At present there are no clinical or laboratory signs of recurrence of the hematological malignancy. She is on maintenance treatment with MMX mesalazine per os.

The described case raises some questions concerning, among others, the reasons lying behind the extremely low frequency of HD in young patients with IBD, the possible (if any) common etiopathogenetic link between these two disorders in case of concurrent appearance, and the role of medical treatment in the course and natural history of both diseases. Epstein-Barr virus is the only infectious agent that has been consistently associated with HD, and notably EBV-encoded RNA is detected in the Hodgkin and Reed-Sternberg cells in up to 40% of cases [5]. This virus has also been linked positively to the development of lymphoproliferative disorders in patients with IBD. Patients with HD are usually treated with chemotherapy alone or combined therapy with external beam radiation. Our patient was successfully treated with 4 cycles of combined chemotherapy achieving complete remission of HD till now. However, there are no data concerning the influence of chemotherapy administered for a given malignancy on the colon mucosa in patients with UC. It also remains unanswered what would the influence of corticosteroids and/or immunosuppressives be on the previous hematological malignancy and the possibility of appearance of a new one in the upcoming years. We conclude that clinicians must bear in mind that UC could appear after diagnosis and successful treatment of HD. Treatment of UC can be succeeded by using the same therapeutic modalities as in ordinary cases.

References


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Restless legs syndrome in patients with Crohn’s disease

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A recent prospective multicenter study by Weinstock et al indicated that restless legs syndrome (RLS) is commonly found in patients with Crohn’s disease (CD), with incidence and prevalence rates of 42.7% and 30.2% respectively [1]. This disease entity, even though it has a great impact on both sleep disturbances and on quality of life, has not yet been thoroughly investigated in patients with CD.

RLS is a common, but significantly underestimated and misdiagnosed, neurological disorder affecting about 5-10% of the general population in Europe and the United States [2]. It is mainly characterized by a sense of discomfort and an urge to move focused on the legs; the diagnosis can be set with the standard diagnostic criteria that have been established from the International RLS Study Group since 1995 (Table 1).

Although iron deficiency anemia is considered as a secondary cause of RLS, Weinstock et al found that current iron deficiency in patients with CD was not related with a higher incidence of RLS symptoms. However, documented iron deficiency in the past was significantly related with the occurrence of RLS symptoms at the time of the study [1]. Inadequate iron stores, with a ferritin level below 50 mcg/L,
Table 1 The essential diagnostic criteria of the restless legs syndrome [7]

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<td>1</td>
<td>An urge to move the legs, usually accompanied or caused by uncomfortable and unpleasant sensations in the legs (or other body parts, in addition to the legs)</td>
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<td>2</td>
<td>The urge to move or unpleasant sensations begin or worsen during periods of rest or inactivity (such as lying down or sitting)</td>
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<td>3</td>
<td>The urge to move or unpleasant sensations are partially or totally relieved by movement (walking or stretching), at least as long as activity continues</td>
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<td>4</td>
<td>The urge to move or unpleasant sensations are worse in the evening or at night than during the day, or only occur in the evening or night</td>
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have been associated with a greater intensity of RLS symptoms and subsequent sleep disturbances in a retrospective study of patients with RLS by Sun et al [3].

Apart from iron deficiency anemia, both CD-related polyneuropathy [4] and bacterial overgrowth [1] have been hypothesized to be involved in the pathogenesis of RLS in patients with CD as well as micro-element deficiencies. In patients with irritable bowel syndrome Weinstock et al have found that small intestinal bacterial overgrowth has been related with RLS symptoms and a significant RLS improvement has been observed in the subgroup that was treated with a long-term antibiotic therapy [5,6]. RLS amelioration has also been reported in 44.5% of CD patients with overall symptom improvement, further supporting the relation between RLS and CD [1].

In conclusion, gastroenterologists treating patients with CD should be aware of the high frequency of CD and RLS comorbidity, as RLS is very often underdiagnosed. Treatment of the underlying inflammatory bowel disease, serum ferritin level monitoring with necessary iron supplementation and adequate control of the intestinal bacterial overgrowth could be the initial steps in the management of CD patients with RLS.

References


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Pyostomatitis vegetans leading to Crohn’s disease diagnosis

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Pyostomatitis vegetans (PV) is a rare oral condition of unknown pathogenesis, usually considered a specific marker of inflammatory bowel disease (IBD), in particular Crohn’s disease [1-5]. PV manifests with yellowish, linear pustules set on an erythematous base that are usually called “snail track ulcerations” [3,4]. Hallepou first introduce the term PV for similar skin lesions in 1898, and McCarthy described the oral analogue as PV in 1949 [6]. The presentation of PV may coincide, precede or follow intestinal involvement [6,7].

We present a rare case of PV that led to the diagnosis of Crohn’s disease, in order to emphasize the association of oral and gastrointestinal (GI) diseases.

A 58-year-old man was referred for evaluation of “white lesions that looked like pus” on the labial gingiva. The lesions peeled off during tooth brushing, without causing any discomfort. They had been initially diagnosed as pseudomembranous candidiass and unsuccessfully medicated with Miconazole nitrate oral gel. The patient’s medical history disclosed that during the last few months he had been suffering from bloody diarrhea. Complete colonoscopy was performed by a GI specialist, and a diagnosis of “hemorrhoids” was given.

Clinical examination revealed numerous yellow-white mucosal pustules on the labial maxillary and mandibular gingiva, as well as the hard palate. They were asymptomatic, set on an erythematous base, had the characteristic appearance of “snail-tracks”, and easily peeled off upon rubbing with...
“Snail-track” mucosal pustules on an erythematous base on maxillary gingiva

gauze (Fig. 1). The rest of the mucosal examination as well as head and neck examinations were within normal limits. The patient did not report the presence of any skin lesions.

A recent complete blood count, performed for a regular check-up, showed anemia [RBC 4.28 (4.50–6.30*10^6/μL), HB 10.7 (14-18 g /100 mL), HCT 34.8 (40-52%), MCH 25.0 (26-32 pg), MCHC 30.7 (32-36 g /100 mL), ferritin 5.20 (25-380 g /100 mL), Fe 44 (65-157 mg/dL), peripheral eosinophilia, EOS 15.6 (1-3%), and increased erythrocyte sedimentation rate 38 (<20 mm/h)].

With the clinical diagnosis of PV, a partial biopsy was performed under local anesthesia. Microscopic examination of 5 μm thick formalin-fixed and paraffin-embedded tissue sections stained with hematoxylin and eosin, showed acanthosis and pseudoepitheliomatoid hyperplasia of the focally detached parakeratinized stratified squamous epithelium, and intraepithelial and subepithelial microabscesses, composed of eosinophil and polymorphonuclear cells. The underlying connective tissue demonstrated a mixed inflammatory infiltrate, consisting of eosinophils, neutrophils and lymphocytes. No Candida hyphae were identified in a PAS stained section.

With the provisional diagnosis of IBD, the patient was referred to a GI specialist. A new complete colonoscopy revealed ulcerative-type pancolitis. Biopsies revealed transmural pattern of inflammation and were interpreted as “consistent with Crohn’s disease”.

The oral lesions persisted 2 months after systemic medication with Mesalazine 500 mg daily, probably due to the low dose of medication. The patient was instructed to rinse his mouth three times daily with chlorhexidine 0.12%, but he was lost to follow up.

In conclusion, PV may be rare, but its association with IBD necessitates its prompt recognition. Thorough evaluation of the patient, in particular when GI symptoms and signs are subtle and may be easily overlooked, is very important, as this could result in early diagnosis of the underlying disease.

References


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