An Unusual Cause of Abdominal Pain: Duodenal Cystic Lymphangioma

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Cystic lymphangiomas are rare in children. They commonly present as neck or axillary masses and only rarely involve the gastrointestinal tract. The clinical presentation of these tumors depends on their size and location. They are often discovered incidentally on imaging studies. Children with abdominal lymphangiomas may complain of abdominal pain or urinary symptoms. If large enough, lymphangiomas can present as palpable masses. Duodenal cystic lymphangiomas are extremely rare. We report a case of a duodenal cystic lymphangioma in a girl age 6 years.

Case Report

A girl age 6 years of white race with a lifelong history of regurgitation underwent an elective esophagogastroduodenoscopy (EGD), which revealed normal-appearing esophageal, gastric, and duodenal mucosa and a small hiatal hernia. A Bravo capsule was placed in her distal esophagus during the EGD. There were no immediate complications, and the patient was subsequently discharged home. Later the same day, she returned to the hospital with bilious vomiting and colicky epigastric and left upper quadrant abdominal pain. She was afebrile and otherwise asymptomatic. Results of a complete blood count test with differential and comprehensive metabolic profiles were normal, as were amylase and lipase levels. Chest and abdominal radiographs were unremarkable, except that the Bravo capsule was seen in her esophagus. A barium swallow study revealed a round filling defect in the third portion of the duodenum. An abdominal ultrasound showed a heterogeneous complex fluid collection anterior to the lower pole of the left kidney. This appeared to be separate from the kidney and adjacent to the tail of the pancreas. In an attempt to further
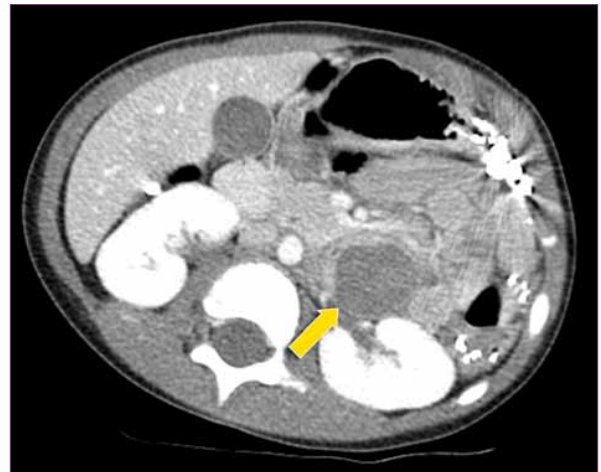
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Figure 1. An abdominal computed tomography scan showing the cystic mass (arrow).

Figure 2. The 5.5-cm duodenal resection specimen contains a well-defined, smooth-surfaced lymphangioma that divides the submucosa (yellow arrow) from the muscularis propria (red arrow).

characterize the lesion, both an abdominal computed tomography (CT) scan (Figure 1) and magnetic resonance imaging (MRI) were performed. These showed the lesion adjacent to the duodenum but separate from the pancreas. During an exploratory laparotomy, the lesion was found to involve the second and third portions of the duodenum and mesenteric vessels. It was resected en bloc (Figure 2).
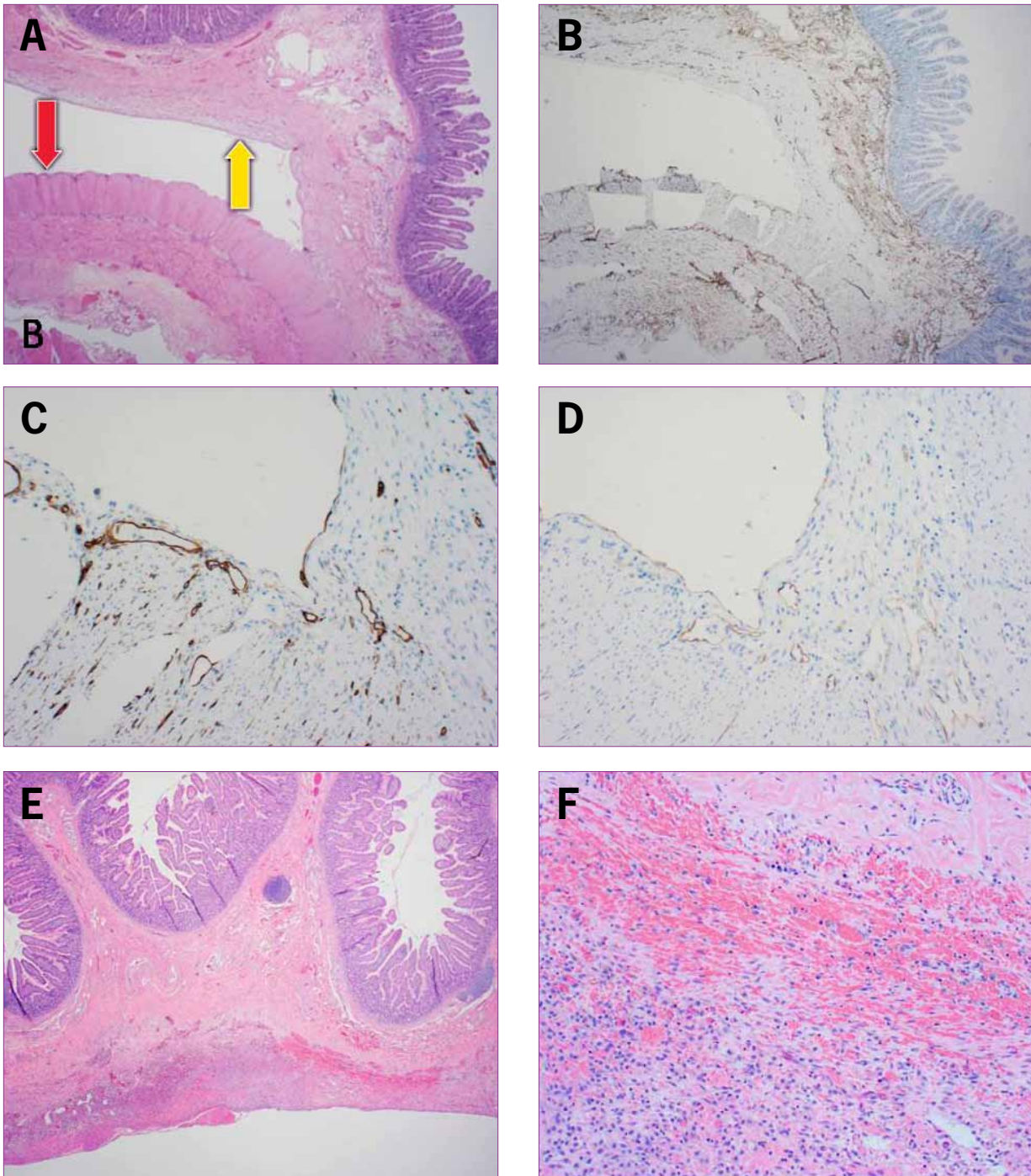
Figure 3. A: The lymphangioma is circumscribed by the submucosa (yellow arrow) and muscularis propria (red arrow). The unremarkable mucosa is present to the far right and on top. The vascular immunohistochemical marker CD34 (**B** and **C**) and the lymphatic immunohistochemical marker D2-40 (**D**) highlight the endothelium. Smaller endothelial-lined spaces are seen in the submucosa (**B–D**) that may be small tributaries of the main cyst. **E** and **F**: The cyst wall has extensive reactive changes consisting of granulation tissue, hemorrhage, and hemosiderin deposition. The duodenal mucosa is unremarkable. (Hematoxylin and eosin stain: **A**, **E**, and **F**; 10× magnification: **A**, **B**, and **E**; 100× magnification: **C**, **D**, and **F**.)

Histologically, it had the appearance of a duodenal cystic lymphangioma (Figure 3).

Postoperatively, the patient's vomiting and abdominal pain resolved. The patient currently experiences intermittent episodes of gastroesophageal reflux that have been well controlled with proton pump inhibitor therapy.

Discussion

Epidemiology

Lymphangiomas account for about 5% of benign tumors seen in infants and children.[1,2] The sites most commonly involved are the neck (75%) and axillae (20%).[3,4] Less than 1% of lymphangiomas are found in the gastrointestinal tract. The incidence of intra-abdominal lymphangiomas has been estimated to be less than 1 per 100,000 individuals.[5] Since the first description of these mesenteric cysts in 1507, fewer than 1,000 cases have been reported in the medical literature.[6] No sex predominance has been confirmed, although some authors have suggested that the incidence may be slightly higher in females.[3]

Etiopathogenesis

The way in which cystic lymphangiomas develop remains unknown, although several hypotheses have been postulated regarding their origin. Acquired factors that may contribute to the development of these cysts include trauma, inflammation, or degeneration of the lymphatic ducts.[7] A congenital cause may be the early sequestration of lymphatic vessels that fail to establish connections with normal draining lymphatics.[8] These lymphatics may dilate under the pressure of accumulating lymph, forming large cysts. This hypothesis is the most widely accepted one, although it fails to explain the invasive nature of many cystic lymphangiomas.

Appearance

Grossly, lymphangiomas have been classified as capillary, cystic, or cavernous.[9] Capillary, otherwise known as "simple," lymphangiomas consist of small, ill-defined, dilated, lymphatic vessels with rich cellular stroma. Cystic lymphangiomas consist of several cystic formations of serous, serosanguinous, or chylous fluid separated by fibrous septae. Most abdominal cystic lymphangiomas are multilocular, although unilocular cysts may occur and can be mistaken for mesenteric cysts. Cavernous lymphangiomas are large cystic masses that usually present as compressible tumors.

Histologically, cystic lymphangiomas consist of dilated lymphatic channels with a single layer of flattened endothelium consisting of fibrocollagenous and lymphoid tissue. The content of the cyst varies from serous to chylous fluid, based upon the degree of stasis, whether or not communication with surrounding lymphatics exists, and the presence of coexisting infection or hemorrhage. In our

patient, hemorrhage was noted within the cyst following resection. This likely occurred secondary to trauma from the endoscope sustained during the endoscopic procedure.

Clinical Features

Intra-abdominal cystic lymphangiomas most commonly occur in children younger than 10 years. Ninety percent of children with these lesions receive a diagnosis before age 2 years.[10] Rarely, the lesion presents in adolescence or adulthood. Within the gastrointestinal tract, the small bowel mesentery is the most common site involved, followed by retroperitoneal sites.[11] The clinical presentation of these lesions depends on their size and location. They are most frequently found incidentally. Occasionally, they may cause abdominal pain, vomiting, and/or alterations in bowel habits due to intestinal compression, obstruction, and/or intussusception. Although acute complaints tend to occur in children, adults with these lesions tend to experience more chronic symptoms.[12] The abdominal pain and vomiting that our patient experienced most likely developed secondary to partial duodenal obstruction caused by hemorrhage into the cyst cavity and resultant enlargement of the cyst.

Diagnosis

Although plain abdominal radiographs typically fail to show these lesions, they may identify bowel displacement or obstruction. Ultrasonography is very sensitive at detecting cystic masses and can be used to better define them. Cystic lymphangiomas may appear as multilobular masses with internal septations and anechoic fluid. The identification of intracystic and/or intercystic echogenic septae within the lesion is an important element of the diagnosis.[10] One limitation of ultrasonography in defining these lesions is that their relatively large size may interfere with the evaluation of their borders and sites of origin. CT scans can be used to better define the location and extension of the cystic masses. This imaging modality can also determine the attenuation coefficient of the cystic fluid, defining it as chylous fluid, blood, or pus. MRI is more sensitive and specific for the diagnosis of hemorrhagic complications but otherwise offers little information not already provided by the combination of ultrasonography and CT.[10,13] Arteriography, lymphangiography, and radionuclide scans are rarely useful in the evaluation of intra-abdominal cystic lymphangiomas.[10,14]

Treatment

Surgical resection is the definitive treatment for abdominal cystic lymphangiomas.[14,15] Depending on the extent and location of the cysts, bowel resection may be required. This is often the case in the management of retroperitoneal cystic masses. In our patient, the

intimate adherence of the cystic mass to the second and third portions of the duodenum and mesenteric vessels necessitated partial small bowel resection. Complete en bloc resections usually result in excellent outcomes and prognoses for patients.[11,14,16] A 10% recurrence rate has been described in patients who have undergone incomplete resections.[14] In children, ultrasonography is the preferred imaging modality for postoperative surveillance. There are no published guidelines regarding the frequency at which surveillance ultrasounds should be performed.

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Review
Toward an Understanding of Lymphatic Malformations

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Vascular anomalies are diverse and represent a broad spectrum of lesions of multiple pathologies that occur at many anatomic locations. These complex lesions impact a variety of medical and surgical specialties.[1] Historically, the field has been fraught with a litany of confounding medical terminology that often leads to misdiagnosis and inappropriate management.[1] For example, the term hemangioma is often used to describe a wide variety of vascular lesions with variable presentations, imaging characteristics, treatments, complications, and recurrence. The suffix -oma is often applied to many vascular anomalies regardless of pathology, which gives the appearance of an underlying proliferative process for all vascular anomalies.[2]

To minimize confusion and simplify the nomenclature, Mulliken and Glowacki developed a general classification scheme of vascular anomalies based on physical findings, clinical presentation, and cellular and biologic characteristics in which lesions are classified as vascular tumors or vascular malformations.[3] Vascular tumors, of which the benign hemangioma is the most common entity, are caused by abnormal endothelial cell proliferation. On the other hand, the pathogenesis of vascular malformations is related to vascular dysmorphogenesis, in which lymphatic, venous, or arterial vessels are abnormal in structure and connectivity.[2]

Combined lesions that involve multiple vessel types also may occur. Lesions may further be characterized as either "fast-flow lesions," in which the lesions have an arterial component, or they may be "low-flow lesions," which involve capillary, lymphatic, or venous channels. In this classification scheme, "lymphangioma" may be classified as a low-flow macrocystic (>1 cm in diameter), microcystic (<1 cm in diameter), or mixed lymphatic malformation depending on the size of the cystic lesions. This terminology reflects the biologic character of the lesion. Diagnosis and treatment of the lesions often require coordinated care from a variety of medical fields, including primary care providers, medical specialists, radiologists, interventionalists, and surgeons.

Lymphatic malformations, or lymphangiomas, are a common form of vascular malformation that most commonly present as cutaneous lesions in the head and neck region.[1] As noted in the case reported by Rana and col-

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leagues, intra-abdominal lymphatic malformations are rare and may present both in the retroperitoneum and peritoneal spaces.[4] Although these lesions are congenital and present at birth, they may manifest clinically at any age. Presentation may be facilitated by an intralesional or remote infectious process, trauma, or hormonal changes. Lymphatic malformations can vary extensively in size and may be focal or diffuse; the microcystic lesions in particular may cross traditional anatomic planes. Additionally, they may be intimately associated with or involve both hollow visceral and solid organs, which can significantly complicate therapeutic interventions.[5]

Histologically, lymphatic malformations are thin-walled vessels lined by lymphatic endothelial cells that are immunohistochemically positive for endothelial markers D2-40 and lymphatic vessel endothelial receptor 1.[1,6] Typically, the anatomic location will dictate the presenting clinical signs and symptoms. Duodenal and periduodenal (eg, retroperitoneal, intestinal, and pancreatic) lymphatic malformations may present with pain, obstruction, or ulceration and bleeding.[5,7] Anomalous lymphatics in the gastrointestinal tract may also cause ascites as well as hypoalbuminemia and diminished immune function from a chronic protein-losing enteropathy.[1]

These symptoms may lead to an extensive diagnostic evaluation, which often includes cross-sectional imaging and endoscopy. Ultrasonography of a lymphatic malformation may demonstrate cysts of variable sizes.[8] These cysts may be differentiated from bowel and intestinal duplication cysts by the lack of a mucosal ultrasound signature.

The differential diagnosis of intra-abdominal lymphatic malformations also includes cystic oncologic processes of the abdominal, hollow visceral, and retroperitoneal organs as well as infectious processes. Magnetic resonance imaging (MRI) is an excellent modality for defining the nature and extent of lymphatic malformations that show T2 hyperintensity, fluid-fluid levels from protein and blood layering, and septal enhancement, although MRI may be less useful for intestinal lesions.[2,8,9]

Computed tomography with bolus intravenous contrast may also be useful in defining the nature and extent of the malformation.[9] Radionucleotide studies are less helpful in the diagnosis of lymphatic malformations. Rarely, contrast lymphangiography may be useful as an adjunct for determining the location and nature of a lymphatic leak from the malformation.[6] Endoscopy, esophagogastroduodenoscopy, colonoscopy, and capsule endoscopy may demonstrate malformations, showing a mass effect on the intestinal lumen or stigmata of the malformations if they involve the mucosa.[10]

Management of these complex lesions is often achieved using multimodal therapy including surgical exploration, resection, and direct- or imaged-guided intralesional sclerotherapy.[1,9] In some cases, surgical exploration is mandatory if an oncologic process cannot be definitively excluded. Intralesional sclerotherapy using agents such as ethanol, OK-432, doxycycline, and others may be used to treat large cystic lesions. However, the risk of wall necrosis and intestinal perforation demands extreme caution when using intralesional sclerotherapy as a treatment modality. These potential complications of intralesional sclerotherapy make surgical exploration and resection the preferred options for both macrocystic and microcystic lymphatic malformations. Surgical resection may result in cure, particularly if the malformation is limited to the intestinal wall and is completely resected.

Malformations that involve solid organs may require partial resection of the parenchyma to achieve complete resection of the malformation. Surgical therapy must be tailored to the nature of the lesion and the severity of symptoms. It is important to remember that since the underlying pathology of vascular malformations is dysmorphogenesis and not abnormal endothelial cell proliferation, these lesions will not respond to antiangiogenic agents such as corticosteroids, propranolol, or other chemotherapeutic treatments.

Overall, duodenal and small intestinal lymphatic malformations are very rare and require a high index of suspicion and a broad diagnostic approach to establish an accurate diagnosis. An experienced multidisciplinary team of gastroenterologists, radiologists, and surgeons is helpful in developing an appropriate management strategy that provides the best chance of a successful outcome.

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