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Myxoglobulosis, A Rare Variant of Appendiceal Mucocele: Case Report and Review of the Literature

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A B S T R A C T

Introduction: Appendiceal mucocele is a term referring to the abnormal accumulation of mucus into the lumen of appendix regardless of the underlying cause. Mucocele alone is not considered a rare finding in routine appendectomy. However, the coexistence of multiple small intraluminal globoid bodies as "myxoglobulosis" or "caviar appendix" is a rare finding.
Case report: We report a 54 year old woman with complaint of abdominal pain who subequnetly had a surgery with the final diagnosis of peritonitis due to perforated peptic ulcer. During the operation, a jejunal mass and appendiceal dilation were observed incidentally. By histopathologic examination, a diagnosis of submucosal leiomyoma for jejunal mass, nonspecific ulceration for peptic specimen and cystadenoma with myxoglobulosis for appendix specimen was made. She had an uneventful postoperative course and after five months, she reported that she had not experienced any discomfort or pain.
Conclusion: this report presents a rare case of appendiceal myxoglobulosis secondary to mucinous cystadenoma, which was incidentally found in patient with peritonitis due to peptic ulcer disease.

Keywords: Appendectomy; Mucocele; Appendix

1. Introduction

Appendiceal mucocele is a term used to describe the abnormal accumulation of mucus into the lumen of the appendix regardless of the cause (1). Symptoms of Myxoglobulosis-a rare variant of appendiceal mucocele-include chronic abdominal pain or pain episodes simulating acute appendicitis (2). The preoperative diagnosis of this condition (i.e. in mucinous cystadenoma) is very important, considering that it has malignant potential (3). Mucocele alone is not a rare finding in routine appendectomy. Whereas, the coexistence of multiple small intraluminal globoid bodies, accompanied by mucoid material resembling fish eggs, constitutes a special type of mucocele described as "myxoglobulosis" or "caviar ap-

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Implication for health policy/practice/research/medical education:
Mucocele by itself is not considered a rare finding after the routine appendectomy. However, the coexistence of multiple small intraluminal globoid bodies as myxoglobulosis or caviar appendix is considered a rare finding. This report presents a rare case of appendiceal myxoglobulosis secondary to mucinous cystadenoma, associated with peritonitis and suggests that these phenomena should be considered in the differential diagnosis of peritonitis.

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pendix". Myxoglobulosis is a rare finding (4, 5) where the incidence of appendiceal mucocele with myxoglobulosis constituting only 0.35% to 0.8% of mucoceles (6, 7).

In this article we describe a patient who was referred to our hospital with abdominal pain symptoms due to perforated peptic ulcer and during the surgical operation, a mass in the jejunum and caviar appendix were found incidentally.

2. Case Report

A 54 year old woman with abdominal pain for two days was referred to Razi general hospital of Guilan university of medical sciences in September, 2011. At first, the pain was only in epigastrium but was gradually spread to the whole abdomen by the second day of admission. She also had nausea, vomiting and constipation. The patient did not mention any history of previous hospitalizations or major illnesses. She only remembered a mild gastrointestinal disease five or six years before. She did not have any history of smoking and alcohol consumption.

Physical examination revealed generalized tenderness, rebound tenderness and involuntary guarding. According to the history, a broad differential diagnosis was considered such as gastroduodenal, hepatobiliary, and gynecologic diseases but based on the physical examination an acute abdomen diagnosis was suggested by the surgeon. Eventually, peritonitis due to perforated peptic ulcer was confirmed by sub-diaphragmatic air in the chest X-ray. She was transferred to the operating room for laparatomy.

During the surgical exploration, a mass in jejunum and appendiceal dilation and inflammation were seen in addition to generalized peritonitis. After abdominal cavity lavage, resection and gastroraphy of prepyloric rupture, jejunal mass excision, appendectomy and gastric ulcer biopsy was done. On the gross examination, jejunal mass excision consisted of a portion of small intestine measuring 11 cm in length and 2 cm in diameter. Fibrofatty tissue was attached to the mass and a creamy colored submucosal mass of 2 cm in length and 1.1 cm in width and 2.3 cm thick was protruding from the serosal surface (*Figure 1*). Appendectomy showed a dilated appendix measuring 5 cm in length and 1 cm in diameter. On the cut section, appendiceal lumen was dilated by mucinous material and numerous pearl-like spheroids ranging in size from 2 to 7 mm in diameter, which were resembling fish eggs (*Figure 2*).

On microscopic evaluation, gastric tissue revealed a nonspecific ulcer with fibrinoleukocytic exudates. Submucosal jejunal mass composed of benign spindle cell fascicles. Immunohistochemical staining of these tumoral cells revealed negative reaction to S-100 and positive reaction to SMA (*Figure 3* and *Figure 4*). Also tumoral cells of jejunal mass were negative for CD117 (c-kit) and CD34. Therefore these evidences support a smooth muscle origin and submucosal leiomyoma of jejunum.

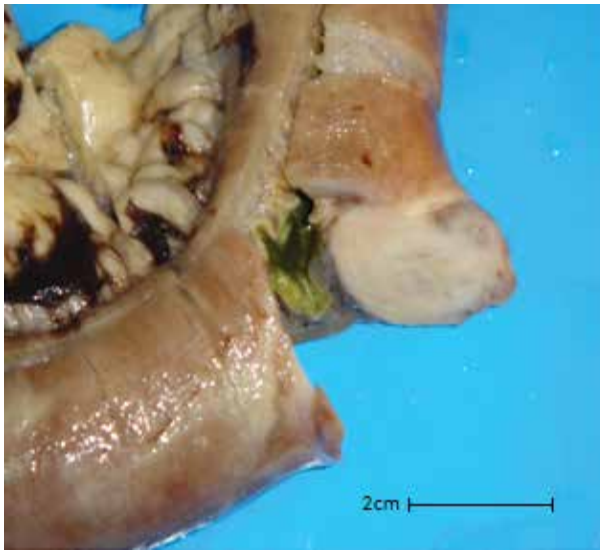

Figure 1. Jejunal Mass Excision Consististing of a Portion of Small Intestine With Attached Fibrofatty Tissue. A Creamy Colored Submucosal Mass is Observed Protruding from the Serosal Surface

Figure 2. Mucinous Material and Numerous Pearl-like Spheroids Resembling Fish Eggs

Appendiceal lumen was focally lined by proliferating mucin secreting columnar epithelium with focal atypia along with numerous intraluminal myxoglobulins that was composed of compact mucinous material without cellular components (*Figure 5*). Pathologic investigation revealed a diagnosis of submucosal leiomyoma for jejunal mass, nonspecific ulceration for peptic specimen and a diagnosis of cystadenoma with myxoglobulosis for appendix specimen.

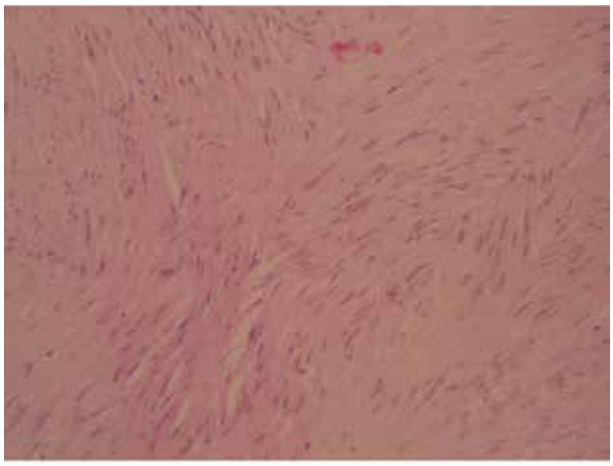

Figure 3. Submucosal Jejunal Mass, H&E Stain Showing the Mass is Composed of Benign Spindle Cells Fascicles

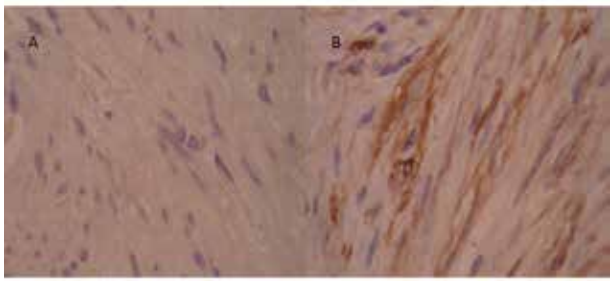

Figure 4. Submucosal Jejunal Mass. IHC Staining Shows the Tumoral Cells With Negative Stainig to S-100 and Positive Reaction to SMA (A,B)

Patient come back to surgery clinic one week later and H-pylori dependent gastric ulcer diagnosis had been confirmed with H-pylori assay of the gastric ulcer specimen. On gastric ulcer specimen, moderate H-pylori coloniza

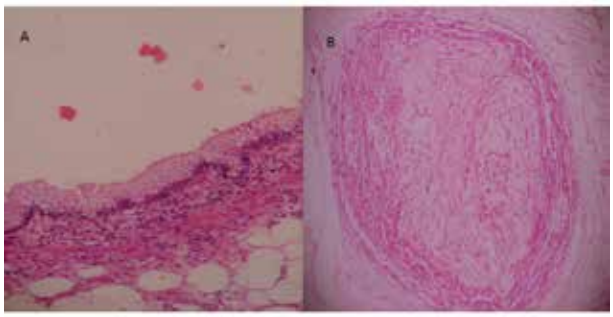

Figure 5. (A) H&E stain reveals that Appendix Lumen Is Focally Lined By Proliferating Mucin Secreting Columnar Epithelium With Atypia (B): Myxoglobulins, Which Is Composed of Compact Mucinous Material Without Any Cellular Component

tion was present on adjacent gastric foveola. Addressing this diagnosis, treatment with the four drugs protocol of H-pylori was prescribed for one month. In further follow up, she had no complaint and proton-pump inhibitor therapy was continued for him. She had an uneventful postoperative course and after five months of follow-up, she did not express any uncomfortable feeling.

3. Discussion

Mucocele of the appendix, regardless of the underlying cause, is characterized by dilatation of the appendiceal lumen due to abnormal accumulation of mucus (8). Myxoglobulosis of the appendix, a subtype of appendiceal mucocele, is identified by the presence of mucinous, often calcified, pear-like globules in the lumen of the appendix (4). Myxoglobulosis occurs most frequently in the sixth or seventh decades of life with a female preponderance (7). The incidence of appendiceal mucocele is estimated at 0.2% to 0.3% of all appendectomies, and the incidence of appendiceal mucocele with myxoglobulosis constitutes 0.35% to 0.8% of mucoceles (6, 7). Latham described the first case of myxoglobulosis in an autopsy in 1897 (4).

Reviewing the past literature, we found only few case reports that in most of them myxoglobulosis has been an incidental finding at autopsy or laparotomy while in few of them it has presented clinically as a “surgical abdomen”, most commonly acute appendicitis (6, 9). Most common complication is generally considered to be either peritonitis or pseudomyxoma peritonei, and perforation of the appendix has been a reported only rarely (6, 9). Our patient was a 54 year old woman which showed similar clinical features.

Although various hypotheses have focused on the formation of a core, which then acts as a nidus for the concentric deposition of mucin, as the initiating event in the pathogenesis of the globules, the exact factors leading to the transformation of mucin into the globular bodies of myxoglobulosis are not known. Some authors propose the hypothesis of bacterial and necrotic epithelial debris origin and that small mucinous masses putatively formed in dilated glandular crypts (6, 9). The most conceivable hypothesis is proposed by Lubin and Berle who suggest that the core of each globule is a necrotic fragment of granulation tissue broken off the appendiceal wall (10). In our case, the globules composed of compact mucinous materials without cellular components too.

Viswanath et al. (7) reported a case of myxoglobulosis secondary to an occlusive membrane. e Brustmann et al. (11) described another case associated with a proximal carcinoid. Aroukatos (4) described a case of myxoglobulosis associated with ruptured diverticulum and Guionmet et al. (12) reported a case of mucinous cystadenoma of the appendix with myxoglobulosis associated with cecal adenocarcinoma. Padhy et al. (13) described a case of myxoglobulosis of the appendix with the clinical diagnosis of recurrent appendicitis. However, the case presented here was novel as it was a case of myxoglobulosis

associated with mucinous cystadenoma.

In the current case, considering the non-ruptured appendiceal wall and absence of mucinous discharge inside the peritoneal cavity, it seems that the patinet'd peritonitis and acute abdominal pain are caused by perforated peptic ulcer and not by the patient's appendiceal pathology. Still, it remains unclear whether the presence of a mass in the patient's jejunum had resulted in increased pressure inside the gastrointestinal tract and caused accelerated perforation of peptic ulcer. In conclusion, this report presents a rare case of appendiceal myxoglobulosis secondary to mucinous cystadenoma, which was incidentally found in patient with peritonitis due to PUD.

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Authors' Contribution

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