Myxoglobulosis of Appendix A Rare Entity

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Abstract We describe a case of the extremely rare entity of myxoglobulosis of the appendix in an 18-year-old man who underwent laparoscopic appendectomy with the clinical diagnosis of recurrent appendicitis. Transaction of the appendix after ligation resulted in extrusion of numerous whitish egg-like globules ranging in size from 0.2 to 0.7 cm in diameter. It was initially thought to be hydatid of appendix, resulting in emergent usage of povidine iodine as scolicidal after suction of all the cysts/eggs. On microscopic examination, the globules consisted of faintly eosinophilic laminations of mucin surrounding an amorphous granular core were found. The mucin was identified by positivity with histochemical mucin stains. After thorough microscopic examination of the appendix, our case was diagnosed as myxoglobulosis due to mucosal hyperplasia, associated with luminal obstruction with possibly a membrane.

Keywords Laparoscopic appendicetomy · Membrane · Mucocoele · Myxoglobulosis

Case Report

An 18-year-old man presented with right lower abdominal pain. He had few episodes of such pain in the past. His general physical examination was unremarkable, except deep tenderness over the right Iliac region. He had a normal TLC count. His abdominal radiograph was normal, while the USG did not reveal any abnormality other than probe

tenderness over the right Iliac region. We did not feel it necessary to do a CT abdomen since the patient was poor.

He was provisionally diagnosed as a case of recurrent appendicitis and planned for laparoscopic appendectomy the next day.

Laparoscopic appendicetomy was performed using supraumbilical 10 mm optical and two 5 mm left iliac ipsilateral ports and observations are given as follows:

- A short 5 cm×3 cm turgid appendix in subcaecal position.
- Bipolar coagulation and transaction of mesoappendix and appendicular artery.
- Double proximal ligation of the stump with 1-0 catgut.
- We routinely use a 5 mm telescope through the inferior-most port while extraction of the appendix is done through the 10 mm umbilical port using a 10 mm gall bladder extractor. This facilitates easy extraction of the most inflamed appendix. The distal stump of the appendix is held with a grasper while the appendix is cut distal to the ligatures. The session is then replaced by the gallbladder extractor and the cut appendix is transferred to it midair, which is then extracted out. We consider this to be our innovation.
- On section of the appendix, multiple small globular egg-like structures escaped from both the cut ends. They were found to be colaced with some kind of mucinous jelly-like substance. Since we never expected this kind of presentation, we were initially confused about the diagnosis. We thought it to be a hydatid of the appendix with escape of daughter cysts (Figs. 1 and 2).
- We did a through suction of all the “daughter cysts” and injected about 20 ml of 5 % povidine iodine as a scolicidal and kept it for 10 min. After which it was sucked out. The whole abdominal cavity was inspected and procedure terminated. The appendix and the egg-like structures were sent for histopathology (Figs. 3 and 4).

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Fig. 1 Multiple small eggs after appendectomy

- The patient made a normal recovery with oral intake after 24 h and he was discharged on the third day after he had started taking oral intake regularly.

Discussion

Myxoglobulosis is an unusual variant of appendiceal mucocoele [1–3]. The initial description of myxoglobulosis of the appendix dates back to 1897 by Latham [1, 2], nearly 55 years after the first description of appendiceal mucocoele by Rokitanski [4]. Since then at least 68 cases have been reported [1]. The incidence of appendiceal mucocoele is estimated as 0.2–0.3 % of appendectomy specimens, with myxoglobulosis constituting 0.35–0.8 % of mucocoeles [1, 5, 6].

The factors leading to the transformation of mucin into globular bodies of myxoglobulosis are unknown. Various hypotheses have focused on the formation of a core, which

Fig. 3 Instillation of povidine iodine as scolical

then acts as a nidus for the concentric deposition of mucin, as the initiating event in the pathogenesis of the globules. Some authors propose the hypothesis of bacterial and necrotic epithelial debris origin and small mucinous masses putatively formed in dilated glandular crypts [7–9]. Lubin and Berle [10] in a report of two cases proposed that the core represented an organizing mass of mucin and granulation tissue originating in the appendiceal wall that broke off and underwent necrosis. In our case, examination of all the globules revealed inflammatory cellular elements. The same general mechanisms requisite in the production of appendiceal mucocoele are involved in the pathogenesis of myxoglobulosis [11, 12]. These mechanisms include (a) partial or complete obstruction of the lumen of the appendix and (b) continued mucin production by a normal or altered—for example, neoplastic—epithelium [13].

Most cases of myxoglobulosis have been incidental findings at autopsy or laparotomy. Patients with myxoglobulosis present clinically in a similar manner to those with mucocoele of the appendix, the disorder occurring most commonly in

Fig. 2 Close up view of the eggs/cysts

Fig. 4 Resected appendix with a cluster of eggs/cysts

the sixth or seventh decades of life with a female preponderance [4]. Patients are asymptomatic and myxoglobulosis is found incidentally at necropsy or laparotomy for other reasons [1, 2, 14]. In a few cases, patients present with episodic right lower abdominal pain with or without features of acute appendicitis as in our case [1, 3, 14]. Complications of myxoglobulosis are the same as mucocoele and include intussusception, bleeding, perforation, peritonitis and pseudomyxoma peritonei [1, 4, 5, 14].

Plain radiographs may show annular, non-laminated calcification of the globules [2, 14], and a barium enema may show a smooth polypoid filling defect in the caecum, with features of an intramural extramucosal lesion [1, 4, 6, 14]. Colonoscopy may show a smooth glassy submucosal or extramucosal caecal mass moving in and out with respiratory movement; this endoscopic sign has been described as the trapped balloon sign [4]. Ultrasound and computerized tomography can be complementary in establishing the diagnosis [6]. The clinical and radiological differential diagnoses include other lesions of the appendix, such as polypoid adenoma, adenocarcinoma, lipoma, lymphoma, prolapse and intussusceptions [15, 16]. Treatment is essentially by appendicectomy, but in cases of rupture or suspected malignancy a standard right hemicolectomy is needed [2–4, 11].

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