Unusual Entities of Appendix Mimicking Appendicitis Clinically – Emphasis on Diagnosis and Treatment

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ABSTRACT

Background: Abdomen is considered a magic box or a Pandora box where you will get different, unexpected pathologies along with rare entities. Appendicitis is the commonest emergency in surgery which presents challenges to surgeons because of a myriad list of differential diagnosis including both medical and gynaecological pathologies. Preoperative imaging plays an important role in diagnosis and management.

Aims and objectives: To study the rare atypical anatomical and surgical presentations of appendix in patients with clinical features of appendicitis. We focus on the clinical features and the role of investigations for the radiological part and management.

Material and methods: This study was done in M.M. Institute of Medical Sciences and Research, Mullana, Ambala, from November 2014 to July 2016. This was a retrospective study. We found 168 cases with the diagnosis of appendicitis, out of which 19 were with rare entities.

Results: Subjects of both genders were aged between 20 and 60 years. Out of 19, 15 were males and 4 females. Four patients were operated for inguinal hernia but incidentally we found appendix in the hernial sac termed as Amyand’s hernia. Another patient presented with obstruction and appendix was forming a band diagnosed as torsion of appendix. Two most interesting cases were diagnosed as appendicular neuralgia and relieved by appendectomy. Out of 19 cases, 7 cases were operated for appendicitis diagnosed as appendicolith. In all the cases appendectomy was done without encountering any complications. Symptom free patients were operated for appendicular neuralgia. No malignancy was found in mucocele appendix at follow up. There were no complications by the 6-month follow-up.

Conclusion: As we came across with different entities of appendix presented with appendicitis, patients should be investigated before proceeding for surgery. In our study, there were incidental findings for which surgeons were not aware of the diagnosis and even for the patient. In inguinal hernia, ultrasonography was not done, diagnosis being made on clinical basis. Clinical and radiological investigations play an important part in early diagnosis and management.

Keywords: appendix, perforation, inflammation, torsion, hernia, mucocele, neuralgia.

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INTRODUCTION

Appendicitis is the commonest surgical emergency for which appendectomy is the only option by either a laparoscopic or an open procedure. The appendicular neuralgia is a rare cause of chronic right lower quadrant abdominal pain (RLQAP), even though no objective disorder can be determined. This condition can be described as chronic appendicitis or (neurogenic) appendicopathy. Van Rossem et al included 10 patients with chronic RLQAP who underwent an appendectomy. After careful selection, elective appendectomy was performed in their centre for this group of patients (1). Mucinous cystadenoma is a rare cystic neoplasm of appendix that develops as a result of proliferation of mucin-secreting cells in an appendix. It is seen in 0.2–0.3% of resected appendices in Europe and the United States. Even in benign disease such as cystadenoma, dissemination of mucin-producing cells into the peritoneal cavity can cause pseudomyxoma peritonei (2). About 25% of patients are asymptomatic and the condition is found incidentally on imaging or at the time of surgery. Another rare entity, known as Amyand’s hernia, with an incidence of 0.07% to 0.13% of all cases of appendicitis, is an inguinal hernia with appendix as the content of hernial sac (3). Radiological investigations play a major role in diagnosing the disease including appendicitis cases. It is very rare to diagnose it preoperatively on Ultrasonography (USG) as an inflamed appendix in the obstructed inguinal hernia (4). Prompt surgery is required to avoid the complications such as incarceration or strangulation and subsequent morbidity (5). Nowadays, diagnostic laparoscopy (DL) is a valuable adjunct to the early diagnosis and management of this often-confounding condition (6). Regarding imaging as per ACR (American College of Radiology), computed tomography is the most accurate imaging study for evaluating suspected acute appendicitis and alternative etiologies of right lower quadrant pain. USG and contrast-enhanced computed tomography (CECT) help in the investigations to diagnose abdominal injuries (6-8).

MATERIAL AND METHODS

This study was done in M.M. Institute of Medical Sciences and Research, Mullana, Ambala, India, in a single unit, from November 2014 to July 2016. A total of 168 cases that were found retrospectively diagnosed as acute appendicitis were included in this study, and in 19 of them rare presentations were seen intraoperatively. All these patients had complete history and underwent thorough clinical, radiological and haematological investigations.

OBSERVATION AND RESULTS

The age range of our patients was 20-60 years. Out of 19, 15 were males and 4 females. Five of the male patients were operated for inguinal hernia and Amyand’s hernia was incidentally found; one male patient had torsion of appendix; two men had mucocele. Two female patients in this series had mucocele appendix. All patients underwent laparoscopic/open appendectomy (Table 1).

Torsion of appendix

The patient admitted in emergency with abdominal pain along with off and on intestinal obstruction. Ultrasonography of the abdomen revealed dilated intestinal loops and inflamed appendix with small amount of collection in the right iliac fossa. In view of obstruction, surgery planned. The midline incision made and a band was overlying the small bowel loops originated from the tip of the appendix. The tip of the appendix was inflamed and formed a mass. This appendicular band was only causing obstruction by twisting to the ileal loops. The band measuring 8 cm in length was coiled around the small bowel loops and colour came to normal, so the resection of the intestine was not performed. Appendectomy was done with preservation of the bowel and gross resected specimen showed long appendix along with an inflamed mass at the tip (Figure 1).

Amyand’s hernia

Four cases were admitted with uncomplicated inguinal hernia and were planned for surgery. After identification of the spermatic cord, sac separated and inside the sac hard structure felt. The sac opened and to our surprise we found appendix lying in the hernial sac. The appendix was non-inflamed, the tip was held and its base took out only through the inguinal hernia incision site. There was no inflammation or perforation of the appendix.
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<tr>
<td>1</td>
<td>30 yrs / male</td>
<td>Presented with pain right lower quadrant, nausea, anorexia with neutrophilic and fever since 2 days</td>
<td>Torsion of appendix</td>
<td>Open appendectomy</td>
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<td>2</td>
<td>42 yrs/ male</td>
<td>Swelling on right sided inguinal region</td>
<td>Inguinal hernia right sided, Amyand’s hernia</td>
<td>Hernioplasty with appendectomy (as there was no infection)</td>
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<td>3</td>
<td>38 yrs/male</td>
<td>Swelling on right sided inguinal region</td>
<td>Inguinal hernia right sided, Amyand’s hernia</td>
<td>Hernioplasty with appendectomy (as there was no infection)</td>
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<td>4</td>
<td>60 yrs/male</td>
<td>Swelling on right sided inguinal region</td>
<td>Inguinal hernia right sided, Amyand’s hernia</td>
<td>Hernioplasty with appendectomy (as there was no infection)</td>
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<td>5</td>
<td>20 yrs/ male</td>
<td>Swelling on right sided inguinal region</td>
<td>Inguinal hernia right sided, Amyand’s hernia</td>
<td>Hernioplasty with appendectomy (as there was no infection)</td>
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<td>6</td>
<td>57 yrs/male</td>
<td>Presented with pain right lower quadrant with fever, nausea and anorexia for 3 days; tenderness was present in RLQ with rebound also; usg revealed dilated luminal diameter of appendix with periappendiceal fluid. Chest x-ray revealed bronchitis.</td>
<td>Mucocele appendix</td>
<td>Open appendectomy with local washing was done.</td>
</tr>
<tr>
<td>7</td>
<td>46 yrs/female</td>
<td>Diabetic female presented with pain migrating from umbilical region to RLQ of abdomen. Neutrophilic with pus cells in urine routine examination was present. Usg revealed a positive target sign with dilated appendicular lumen with perforation at tip.</td>
<td>Mucocele appendix</td>
<td>Open appendectomy</td>
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<td>8</td>
<td>23 yrs/female</td>
<td>Unmarried female came to emergency with recurrent attacks of pain lower abdomen in past one year; usg revealed features of acute appendicitis with haematuria.</td>
<td>Mucocele appendix</td>
<td>Open appendectomy</td>
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<td>9</td>
<td>35 yrs/ male</td>
<td>Presented with pain abdomen, mild fever and nausea for 3 days. There was rebound tenderness in RLQ with usg revealed dilated appendicular lumen and peri appendiceal fluid. CT scan was done</td>
<td>Mucocele appendix</td>
<td>Open appendectomy</td>
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<td>10</td>
<td>45 yrs / male</td>
<td>Presented with dull ache pain (recurrent attacks) since last 3 years. Usg of the abdomen was normal. Patient was a chronic smoker and was having bad chest.</td>
<td>Appendicular neuralgia</td>
<td>Lap appendectomy</td>
</tr>
<tr>
<td>11</td>
<td>30 yrs / male</td>
<td>Presented with pain RLQ recurrent attacks since 1 year presented with dull pain umbilical region with nausea, vomiting and fever.</td>
<td>Appendicular neuralgia</td>
<td>Lap appendectomy</td>
</tr>
<tr>
<td>12</td>
<td>34 yrs / male</td>
<td>Presented with pain RLQ with nausea; patient had tenderness in RLQ with ultrasound showing appendicitis with appendix perforated in middle.</td>
<td>Appendicolith</td>
<td>Lap appendectomy</td>
</tr>
<tr>
<td>13</td>
<td>28 yrs / male</td>
<td>Patient had high grade fever with localised peritonitis features in RLQ had a history of 3-4 days on us; there was ruptured appendix with periappendiceal collection.</td>
<td>Appendicolith</td>
<td>Lap appendectomy. A small calculus/appendicolith was stuck at base of appendix and appendix was ruptured at tip.</td>
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<td>14</td>
<td>29 yrs/male</td>
<td>Presented with constant dull ache pain in RLQ with anorexia and malaise since 3 days received analgesia at home. We revealed features of acute appendicitis.</td>
<td>Appendicolith</td>
<td>Lap appendectomy</td>
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**TABLE 1.** Detailed presentation and operative findings
So, we performed appendectomy, followed by meshplasty (Figure 2). In the postoperative period, patients were given third generation antibiotics along with metrogyl for five days, in view of appendectomy and hemiplasty. They were successfully discharged on the 6th day after surgery without any complications such as infection, hematoma or any discharge from the wound site. Patients were followed up for 6 months and there was no recurrence.

**Mucocele appendix**

Four cases were diagnosed as mucocele appendix and one patient presented with pain abdomen predominantly in the right iliac region. There was no history of weight loss but fever was present off/on. In two cases, a mass was felt in the right iliac fossa for which differential diagnosis was kept as appendicular lump or lymphoma. In other two cases, no mass was felt, except for tenderness in the right iliac area. Abdominal ultrasonography suspected the presence of a mass. On CECT scan a mass was present arising from the appendix diagnosed as mucocele appendix most probably benign (Figure 3). Open surgery planned to avoid the spillage of the cells. On surgery, there was a large whitish mass found in the right iliac area with small nodules, which was involving the appendix (Figure 4a). Base was clear, so appendectomy done (Figure 4b). On histopathology, low-grade appendiceal mucinous neoplasm was diagnosed (Figure 5). At one year follow-up, the patient was well and had no recurrence.

**Appendicular neuralgia**

Three cases presented with abdominal pain along managed with off and on medication but not relieved. Ultrasonography of the abdomen was normal, so we proceeded with the computed tomography (CT) which was also normal, and even magnetic resonance imaging (MRI) was done to rule out the cause of abdominal pain, but MRI was normal too. However, the patient was having abdominal pain and the lower limb flexion test was positive. We planned for diagnostic laparoscopy and to our surprise, the tip of the appendix was lying on the psoas muscle, which explained the cause of his pain. Laparoscopy appendectomy was done and the patient had no longer abdominal pain after surgery.

**Appendicolith**

Seven cases were diagnosed as appendicolith and mostly presented with abdominal pain and fever. Ultrasonography revealed an inflamed appendix and in two cases, appendicolith was seen (Fi-
FIGURE 4. a) Operative section revealed whitish colour lump as mucocele appendix; b) gross specimen revealed mucous in the appendix (base is hold by artery)

FIGURE 5. a) High power view showing the mucinous lining of the appendix; b) section showing flattened mucosa of the appendix which at places is lined by mucinous epithelium (H and E X 40X)

FIGURE 5. a) Ultrasonography revealed appendicolith in the appendix; b) the operative area showed laparoscopic appendectomy and the cut area revealed appendicolith; c) and d) operative specimen of appendectomy and appendicolith held with forceps

FIGURE 7. a) White arrow showed lith lying outside from the perforated appendix; b) white arrow showed appendicolith and black arrow showing artery passed through the perforated lumen; c) black arrow showed perforation in appendix and white arrow showed dilated appendix

gure 6a). Laparoscopic appendectomy was done in two cases (Figure 6b). On surgery, the appendicolith with 7–8 cm in size and perforation was present in the middle of the appendix. In three cases, laparoscopic appendectomy was performed to avoid large incisions (Figure 6c, d). In one case, appendicolith with 2x3 cm in size was firm in consistency, with inflamed appendix, and it was lying outside of the appendix through the perforation site of the appendix (Figure 7a). Surgery was successfully done and the patient felt well, without any pain or fever (Figure 7b). A drain in the form of Ryle’s tube was put in and removed in all patients on the 3rd or 4th postoperative day. All patients had uneventful postoperative recovery.

DISCUSSION

Mucocele of the appendix was coined for the first time by Karl Freiherr von Rokitansky in 1842. It is the condition of appendix in which it is transformed into a mucus filled sac; its incidence ranges from 0.07% to 0.63% and it affects both genders between the 5th and 7th decades of life (9). Mucocele of the appendix is a rare condition and its pathological classification and management strategy have not been standardized yet. A classification of mucinous appendiceal neoplasia was developed, and it was agreed that “mucinous adenocarcinoma” should be reserved for lesions with infiltrative invasion. The term “low-grade appendiceal mucinous neoplasm” was supported and it was agreed that “cystadenoma” should no longer be recommended. A new term of “high-grade appendiceal mucinous neoplasm” was proposed for lesions without infiltrative invasion but with high-grade cytological atypia. It was agreed that low-grade and high-grade mucinous carcinoma peritonei should be considered synonymous with disseminated peritoneal adenomucinosis and peritoneal mucinous carcinomatosis, respectively (10).
Abdominal ultrasonography (US) and computerized axial tomography scan (CT), respectively suspected the diagnosis in only one case (9). If mucocele is benign, it is clinically dominated by acute or chronic pain in the right lower quadrant (11). In our study, patients had no features of weight loss, except for abdominal pain in the right iliac fossa, or were asymptomatic as in cases of Amyand’s hernia. Differential diagnosis should be established with benign pathologies of the appendix like leiomyoma, neuroma, fibroma and lipoma and other conditions such as mesenteric cysts, hydrosalpinx, carcinoid, lymphoma, intussusception, endometriosis and adenocarcinoma of the appendix (12). Thorough histologic examination is essential and it provides the final diagnosis of the appendiceal disease upon histologic examination of the appendectomy specimen.

Another life threatening complication of the appendicitis is the chronically inflamed appendix acting as a tourniquet around a loop of the terminal ileum (13). The exact cause of torsion is unknown. It may develop as a consequence of sudden rotation of the body, or it may be due to a long pedicle, or the vein may be longer and more likely to twist around its accompanying artery, or it may be caused by excess fat in the pedicles. It can also cause intestinal obstruction by forming a band on the abdominal wall or an adjacent loop of bowel and thus kink the bowel; or a loop of the small bowel may be caught under the adherent band; another possibility is that it may initiate intussusceptions (14). Although CT imaging is a highly effective investigative modality in these cases, operative treatment should not be delayed for a radiological investigation in the presence of abdominal peritonism (13). Intraoperative findings revealed inflamed appendix which was rotated around the ileum and tip was forming a mass.

Amyand’s hernia is defined as the occurrence of the appendix in an inguinal hernial sac and in case of appendicitis; its incidence is only 0.1%. If diagnosis is made before surgery by CT, it is possible to treat Amyand’s hernia laparoscopically (15). They reported three rare cases with different presentations and emphasize that USG and CT plays an important role in diagnosis and management. We came across with five cases diagnosed as Amyand’s hernia, which was an incidental finding in cases where we performed inguinal hernia surgery. If appendix in the hernial sac is not inflamed, then it should be removed and Meshplasty could be done.

Appendicolith is also another rare phenomenon also known as faecolith, coprolith, stercolith, enterolith, or concretion and is composed of firm faeces and some mineral deposits in the lumen of appendix. It contains fats (coprosterols), inorganic salts (calcium phosphate) and organic residue (vegetable fibres) in a proportion of 50%, 25% and 20%, respectively. Other causes of calcific areas of high attenuation in the abdomen include dropped gallstones, calcified epiploic appendagitis, dropped surgical clips, and calcified mesenteric lymph nodes (16). The prevalence of faecoliths in the general population is 3%, and appendicoliths are seen in 10% of cases with acute appendicitis. However, giant appendicoliths (>2 cm) are extremely uncommon and the largest appendicolith found by us was 2.3 cm (2 cm on CT) (17, 18). All reported cases of giant appendicoliths have been managed by extraction, either surgically or endoscopically, mostly due to the presence or perceived risk of appendicitis (19). Singal R et al. (20) reported the case of an inguinal hernial stone termed as herniolithiasis, where a stone was found in the hernial sac. The stone contains calcium (60%) and phosphate (40%) to be the principal constituents without any amounts of oxalate, urate and cholesterol. X-ray of the stone showed radiopaque shadow (21). In our study, we came across three cases diagnosed intra-operatively as appendicitis due to appendicolith. Laparotomy has been the most common approach, but two cases of endoscopic extraction have been described in recent years (16).

Another very rare entity of the appendix was a rare cause of chronic right lower quadrant abdominal pain (RLQAP) (1). The term “neurogenic appendicopathy” has been used for patients operated on for acute appendicitis with their appendices lacking signs of acute inflammation. They studied 40 cases diagnosed with neurogenic appendicopathy out of 121 cases. Appendix specimens were immunohistochemically examined for the expression of S-100, vasoactive intestinal polypeptide (VIP), and substance P. VIP was more strongly expressed in control specimens (p = 0.0211). Substance P was of no diagnostic value. Postoperative pain scores differed significantly between the groups, favouring appendectomy (P = 0.005) (21). They observed that persistent or recurrent lower abdominal pain can be treated by elective appendectomy with significant pain reduction in properly selected cases, despite the lack of abnormal histology in neurogenic appendicopathy presented with chronic right lower
CONCLUSION

We came across with different and rare intraoperative pathological entities in the appendix in patients who presented in emergency with clinical and radiological features of appendicitis and received timely surgical treatment.

“Abdomen is like magic; you don’t know when it may reveal a surprise finding or/how much time it will take to finish; so, always fill your stomach and empty your bladder”. By Rikki Singal.

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REFERENCES