Case report

Large Asymptomatic Mucocele of The Appendix – An Incidental Finding

ABSTRACT

Mucocele of the appendix is a rare condition. It is characterized by an abnormal dilatation of gland due to accumulation of large quantity of mucus. It may be caused by epithelial proliferation of appendix which a be either benign or malignant. It is often incidentally found on routine appendicectomy specimen or radiological studies done for some other abdominal ailments. However, a correct pre-operative diagnosis and selection of appropriate surgery ranging from simple appendicectomy to right hemicolectomy depending on the size, location and lymph node status is crucial. We present here a case of a young female patient who came for annual routine health checkup and was incidentally diagnosed with mucocele of appendix. Presence of mucocele was confirmed on CT scan and the patient underwent surgery with histopathological examination confirming the simple mucocele of appendix. She recovered well and the postoperative follow-up period was uneventful.

This case report aims to highlight the importance of routine annual health check-up and timely diagnosis of conditions like mucocele of appendix.

Keywords: Mucocele, appendix, mucinous cystadenoma, mucinous cystadenocarcinoma, health checkup

1. INTRODUCTION

Mucocele of the appendix is a rare disease that is characterized by distention of the appendiceal lumen regardless of the process [1]. There is a progressive retrograde dilatation of the vermiform appendix associated with intraluminal accumulation of the mucoid substance. The reported prevalence of mucocele in appendicectomy specimens is 0.2-0.3% [2].

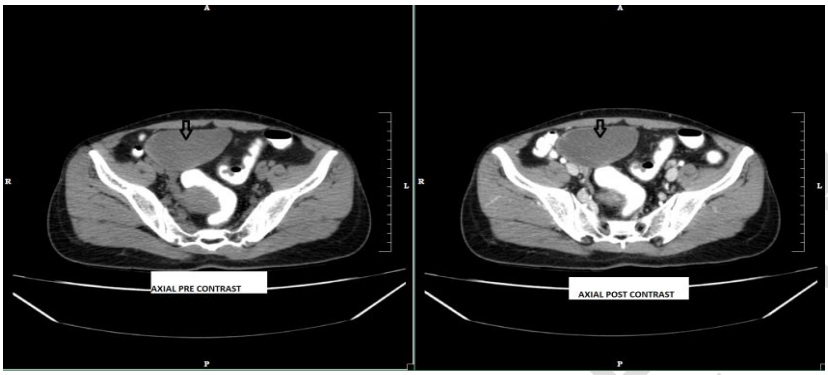
They do not have a specific clinical presentation. They sometimes present with vague lower abdominal pain and might be mistaken for acute appendicitis hence they are commonly detected in the pathological specimen of an appendicectomy. Such appendiceal mucinous lesions are generally detected incidentally while evaluating an unrelated complaint [3-5].

A correct preoperative diagnosis of this condition is crucial for the selection of an adequate surgical method to prevent peritoneal dissemination and to prevent other intraoperative and postoperative complication and to avoid repeat surgery in such patients. One of the chief principles of surgical treatment is that the intact mucoceles do not possess a threat to the patients. A perforated mucocele or an inadequate treatment causes the epithelial cells to escape into the peritoneal cavity developing pseudomyxoma peritonei which is difficult to treat with a high mortality rate [6].

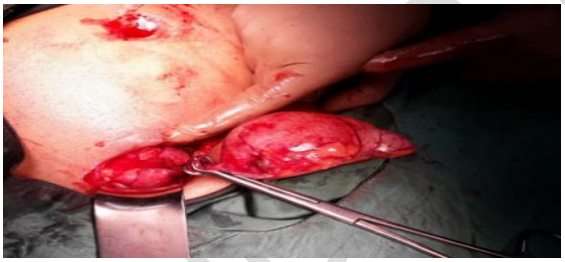
We present a rare such case of mucocele of appendix which was detected incidentally in a young female patient on routine annual health check-up.

2. PRESENTATION OF CASE

A 36-year-old female patient came to the hospital wellness center for a routine annual health check-up. She had no history of abdominal pain, fever, nausea, or vomiting. There were no urinary or bowel complaints. Her ultrasound scan revealed a well-defined cystic lesion of size 9 cm X 5 cm with echogenic material with debris noted in the right iliac fossa suggestive of appendicular mucocele or a mesenteric cyst and hence the patient was referred to the surgery OPD for further evaluation. Her abdominal examination on inspection and palpation was within normal limits. A contrast-enhanced CT scan of the abdomen and pelvis revealed a dilated appendix of size 5 cm filled with a low-density material suggestive of a mucocele of the appendix (Figure 1). After thorough work-up and counselling, the patient was posted for an open appendicectomy. The appendix was large and swollen at its tip and body with an inflamed wall without any evidence of perforation (Figure 2). The appendicular dimensions were 9.1 x 4 x 4 cm approximately. The base appeared normal with approximately 2 cm of length free from any swelling (Figure 3). No collection was seen in the peritoneal cavity. There was no evidence of lymphadenopathy. Conventional appendicectomy was performed as the base of the appendix was free from any pathology and the specimen was delivered out intact without rupture. The histopathological examination confirmed the simple mucocele of the vermiform appendix. The postoperative period was uneventful and the patient was discharged after three days and had been on regular follow-up.



(Figure 1: CT scan in axial section with contrast injection, showing a dilated appendix in arrow)



(Figure 2: Mucocele of the appendix as seen during surgery)

(Figure 3: Mucocele of the appendix delivered out intact)

3. DISCUSSION

Mucocele of the appendix is characterized by abnormal accumulation of a huge quantity of mucus in the lumen resulting in dilatation of the gland. The appendix is lined by epithelium which has more goblet cells. Thus, a majority of epithelial tumors start as mucoceles [6]. The incidence of mucocele of the appendix is only 0.2% to 0.7% hence making it a rare entity [7,8]. Most of these cases are identified on histopathological examination of appendicectomy specimens or diagnosed incidentally on medical imaging examinations. Pathologically it is classified into four types: a) simple or retention cyst, b) mucosal hyperplasia, c) mucinous cystadenoma which is a dilated appendix filled with mucin lined by atypical mucinous epithelium having minimal dysplastic features, and d) mucinous cystadenocarcinoma which is defined as mucin filled dilated appendix along with the presence of high-grade cell dysplasia and usually stromal invasion beyond the muscularis mucosa [9,10].

Most commonly the patients are asymptomatic and appendiceal mucocele is an incidental finding during surgical intervention, imaging, or endoscopic procedures. It may also present sometimes as acute appendicitis. Other modes of presentation can be lower abdominal pain, abdominal mass, anemia, and weight loss [11-13]. There is a known association between mucocele of the appendix and synchronous or metachronous colorectal tumors and ovarian mucinous tumors. So, colonoscopy and appropriate radiological in the pre or postoperative period is of paramount importance along with surgical exploration. Ultrasound examination shows a cystic mass with poorly defined wall and variable internal echogenicity depending on the composition of mucocele. It shows a pathognomonic onion-skin appearance. The diameter of the appendix over 15 mm has been implicated as the threshold for diagnosis of appendiceal mucocele. The outer diameter limit for the diagnosis of acute appendicitis has been established as 6 mm. The role of CT scan in diagnosing a mucocele is related to the detection of appendicular lumen greater than 1.3 cm, the presence of cystic dilatation, and mural calcification. CT scan is a cardinal diagnostic tool for mucocele of appendix and it can determine the relation between lesion and the surrounding structures and help to confirm the diagnosis. CT scan is more reliable method of diagnosis as compared to USG and is preferred because of its higher diagnostic capacity for the mucocele [14-16]. The diagnostic finding of mucocele on colonoscopy is the volcano sign in which the appendiceal orifice is in the center of a firm mound covered by normal mucosa or a yellow-colored lipoma like submucosal mass. Colonoscopy is also useful in identifying synchronous GI tumors in patients with abdominal pain [17].

Surgery is the mainstay of treatment. The extent of surgical intervention ranges from appendectomy to right hemicolectomy, which depends on various factors like the size of the tumor, location in the appendix, involvement of the cecum and ileum, if mucocele is perforated or not, involvement of lymph nodes, presence of mucus collection, the safety margin and the final histology [6,13,14]. For the synchronous lesions, the extent of surgery should encompass the colonic pathology, regardless of the character of the mucocele [18]. So, patients may require an appendicectomy or up to a right hemicolectomy including cytoreductive surgery. Atraumatic removal of the appendix plays a crucial role in avoiding rupture and dissemination. Conversion into laparotomy should be considered if the lesion is traumatically grasped or there is an extension of the tumor beyond the appendix or signs of malignancy like peritoneal deposits are seen [19].

If a mucocele is seen during laparoscopy, Dhage-Ivatury and Sugarbaker recommend converting the procedure to a laparotomy. This aids to perform surgery meticulously so the cyst is not ruptured resulting in spillage in peritoneal cavity, and with an open surgery it is feasible to inspect and palpate the areas where mucinous tumors are more common. The

procedure is converted to a hemicolectomy in cases of suspected malignancy. Iatrogenic spillage can give rise to pseudomyxoma peritonei which requires intraperitoneal chemotherapy [6]. Right hemicolectomy and cytoreductive surgery (CRS) are combined with heated intraperitoneal chemotherapy (HIC) or early post-operative intraperitoneal chemotherapy (EPIC) in cases of perforation, appendiceal lymph nodes, or positive margins of excision [20]. A long-term follow-up is suggested for such patients. A correct preoperative diagnosis helps in planning of a careful mobilization and resection to prevent peritoneal spillage and contamination.It is also essential to differentiate between acute appendicitis and mucocele of appendix before surgery and select all necessary surgical approach. It not treated adequately or appropriately the mucocele may progress and the epithelial cells may contaminate the peritoneal cavity resulting in pseudomyxoma peritoni, which has a high mortality [21,22].

In this case, mucocele was an incidental finding on a routine annual health check-up. It was unperforated, located more than 2 cm away from the base of the appendix (negative margins of resection), and no regional lymphadenopathy was found. Hence appendicectomy was performed which was adequate as per the standard algorithm. Histopathological examination confirmed it to be a simple mucocele. The patient had an uneventful follow-up.

4. CONCLUSION

Though mucocele of the appendix is a rare finding, it should be considered as a differential diagnosis of cystic lesions in the right lower quadrant of the abdomen. In our institute, we perform regular preventive health check-ups. In this particular case, we found an asymptomatic cystic mass in the abdomen which was diagnosed as a Mucocele of the appendix and was surgically treated**.** Preventive health check-up plays a very vital role to identify such rare, asymptomatic, and treatable diseases. To conclude, we recommend all tertiary care institutions and health care organizations to take an initiative in conducting frequent preventive health check-ups and thereby help in the early diagnosis and treatment of such rare diseases.

CONSENT

All the authors declare that ‘written informed consent was obtained from the patient (or other approved parties) for publication of this case report and accompanying images’.

ETHICAL APPROVAL

As per international standard guidelines, written ethical approval has been collected and preserved by the author(s).

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