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Prospective Study of Incidence of Appendiceal Mucocele in Cases of Acute Appendicitis: At a Teaching Institute

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Abstract

Purpose of this study is to know incidence of Mucocele of appendix amongst cases of acute apppendicits operated at an Institution. Mucocele of Appendix occurs when there is abnormal accumulation of mucus within the appendix leading to dilation of appendiceal lumen by mucus. The disease is considered as a rare lesion of the appendix, which is found in 0.2 to 0.3 % of the appendectomies. The course and prognosis of mucocele related to the histological subtypes. Preoperative diagnosis is difficult due to non-specific clinical manifestation and 50% are discovered incidentally at surgery. Patient may present with pain in lower right quadrant of abdomen and surgeon may mistake it for acute appendicitis. Treatment is always surgery and depends on the integrity and size of the appendix base and on the histological type of the disease.

Keywords: Mucocele; Appendix; Pseudomyxoma Peritonei; Sonography.

Introduction

Acute appendicitis is the most common cause of acute abdomen requiring surgical intervention [1-2]. Although typical, uncomplicated cases of acute appendicitis are easy to diagnose and treat, diagnosis of atypical appendicitis is a difficult task and remains a clinical challenge that may test the diagnostic skills of even the most experienced surgeons.

Typical uncomplicated cases of acute appendicitis are easy to diagnose and treat. Typical cases present classically with para-umbilical pain (visceral pain) migrating to the right lower quadrant of the abdomen (RLQ). Pain usually is associated with nausea, vomiting and low-grade fever.

Localized irritation and inflammation of the peritoneum results in pain with cough (Dunphy's sign), tenderness and muscle guarding on palpation in the RLQ over McBurney's point and rebound tenderness elicited by deep palpation with quick release (Blumberg sign). Unfortunately, 20-33% of the patients suspected of having acute appendicitis present with atypical findings [3-5].

One in 15 persons develops appendicitis during his or her life time [6]. The disease occurs at all ages but is most frequent in the 2nd and 3rd decades of life. The people of the developed countries being educated are aware of the consequences of the appendicitis; hence attend clinics/hospitals immediately as soon as they develop abdominal pain. But the situation is different in our areas; people are poor and illiterate, they present late. Therefore, complication rate may be higher and understanding the complications of acute appendicitis, will help to manage these patients properly and decrease the morbidity and mortality. The complications of appendicitis include; appendicular perforation, appendicular mass, appendicular abscess, gangrene of the appendix, portal pyemia leading to liver abscess and intestinal obstruction. Once appendicitis progresses to complications, the morbidity increases and in some cases it may prove fatal.

Mucocele of appendix is an uncommon disorder

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which is often asymptomatic and it has 0.2% to 0.4% prevalence among appendectomies [7-10]. The term mucocele is widely used in diagnosing both benign and malignant lesions, but specific criteria are being proposed for definitive diagnosis and surgical management of appendiceal mucocele [11]. There are 4 histological types of appendiceal mucocele: retention cyst, mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma [12,13]. Mucocele is a generic descriptive term for cystic dilatation of the appendix lumen [13]. Mucinous cystadenoma and cystadenocarcinoma comprise most of the case [14]. The neoplastic mucoceole and retention cysts are clinically and radiologically difficult to differentiate, but malignant mucoceles have additional findings [14] .If not treated properly or perforation of appendix may lead to extravasation of its contents (epithelial cells) into peritoneal cavity may lead to pseudomyxoma peritonei which has very poor prognosis and has a high mortality [15]. Mucocele of the appendix was first described by Rokitansky in 1842, [16], and formally named by Feren in 1876.The clinical presentation of a mucocele is usually non-specific and up to fifty percent are discovered incidentally at surgery [17]. This is characterized by the dilation of the organ lumen with mucus accumulation, being more frequent among individuals aged 50 years or more. Some articles confirm its prevalence among women [18,19], others demonstrates a higher incidence among women [14,20].

The purpose of this study is to know the frequency of incidence of mucocele, its symptoms and management in cases of acute appendicitis. Though a lot of international literature is available on this topic but local research is inadequate. Hence there is need to study the incidence of mucocele of appendicitis in cases of acute appendicitis managed at PIMS, teaching hospital, Jalandhar. Study will tell us frequency of this disease and different treatment options.

Patient presenting with pain in right iliac fossa may be mistaken for acute appendicitis, one of the most common surgical diseases [9,15,21,22]. It is important to differentiate between these two pathologies before surgery.

Material & Methods

The study included 160 patients of acute appendicitis undergone open and laparoscopic appendectomy at Punjab Institute of Medical sciences [PIMS] Jalandhar. All the cases were admitted through emergency. Of these, 160 patients were enrolled the study includes 96 patients males and 64 females with mean age of 32 years (range 6yrs-70yrs). 108 patients had undergone open appendectomy and 52 were subjected to laparoscopic appendectomy and all the specimens were sent for histopathology. In all the cases patient was diagnosed clinically and were subjected to routine investigations including CBC, BT, CT, Blood sugar, RFT's, LFT's, Viral markers, Urine examination, ECG and all the patients were advised USG abdomen. Study includes only operated cases and not those who were managed conservatively. Alvarado scoring system was used to make diagnosis of acute appendicitis.

Alvarado Scoring System

Symptoms	Score
Migratory right iliac fossa pain	1
Nausea/Vomiting	1
Anorexia	1
Signs	
Tenderness in right iliac fossa	2
Rebound tenderness in right iliac fossa	1
Elevated temperature	1
Laboratory findings	
Leucocytosis	2
Shift to the left of neutrophils	1
Total	10

The score is based on three symptoms, three signs and one investigation as shown in Table 1. The classic Alvarado Score included left shift of neutrophil maturation (score 1) yielding a total score of 10, Patients with a score of 1-4 are considered unlikely to have acute appendicitis, those with a score of 5-6 have a possible diagnosis of acute appendicitis, not convincing enough to have urgent surgery, and those with score of 7-9 are regarded as probably having acute appendicitis.

Application of Alvarado scoring system in diagnosis of acute appendicitis can provide a high degree of positive predictive value and thus diagnostic accuracy. The positive predictive value of Alvarado score is reported as high as 85.3%, 87.5% and 87.4% in many studies [23-25].

On USG and clinically not a single case was diagnosed as Mucocele of appendix in 160 cases.

Results

It was observed in our studies that preoperative diagnosis is difficult and incidence of this rare disease which is found in 0.2-0.4% of cases. Once preoperative

or peroperative mucocele of appendix is suspected all the precautions to prevent dissemination or rupture of appendix was taken. Treatment is always surgery and depends on the integrity and size of appendix base and histological type of the disease.

Fig. 1: Peroperative Mucocele of appendix

Fig. 2: Yellowish gel like material: to be evacuated out of abdomen (Prevent spillage)

Discussion

Acute appendicitis is a clinical entity which needs surgical treatment in shortest possible time after the attack, if ignored it may get complicated and increase the morbidity and may prove fatal [26]. Mucocele of appendix is a condition where patient can land into dreaded complication of pseudomexoma peritonei if not diagnosed preoperatively as well peroperatively .During four years study from 01-06-2013 to 01-04-2017, 160 cases of acute appendicitis operated,were examined and treated. Not a single case was diagnosed preoperatively. In one case on USG was diagnosed as cystic mass in right iliac fossa was suspected to be ovarian cyst.

In one case cystic dilatation of appendix was 6cm on per operative and diagnosis was confirmed histopathlogically.

Incidence of mucocele of appendix is as low as 0.2%-0.4% as per literature and in our study only 2 cases were diagnosed after surgery as mucocele of appendix. Both the cases were operated by open surgery.

Accumulation of mucous secretion is slow and gradual, with no signs of infection inside the organ leading to dilatation of appendicular lumen. It results from the lumen obstruction in the appendix, which is secondary to epithelial proliferation, either benign or malignant of the appendix mucosa, or of lesions in the caecum, adjacent to the appendiceal ostium. Much less frequently, inflammatory or obstructive causes, to include appendicitis and obstruction by a fecolith or appendicolith, are the cause of mucocele formation. The differential diagnosis of mucocele of the appendix includes mesenteric cyst, duplication cyst, right ovarian cyst and hydrosalpinx. Intra- luminal bubble of gas, or an air-fluid level with in mucocele suggest the presence of superinfection, which can occur in both benign and malignant mucoceles. Mucocele of appendix is a descriptive term for an appendix distended by mucus, secondary to mucinous cystadenoma (63%), mucosal hyperplasia (25%), mucinous cystadenocarcinoma (11%) and retention cyst, [27].

The epithelial lining of the appendix consist of abundant exocrine goblet cells, and thus most tumor types seen in appendiceal samples are mucus producing [28]. Excessive production of mucus by adenomatous tumors lead to formation of a mucocele and is usually caused by the entrapment of mucus and characterized by invasion of mucus into the appendiceal wall [29].

The carcinoembryonic antigen (CEA) level at preoperative may suggest malignancy in the appendix or in the colon [30,31].

Imaging techniques Ultrasonography, Computed tomography (CT), and less often MRI have been described for preoperative diagnosis. On USG typical cystic mass with variable internal echogenicity . The presence of an "onion skin sign" (Sonographic layering within a cystic mass) is considered a highly suggestive feature [32]. Appendix diameter 15mm or more in USG examination has been determined as

the threshold for appendiceal mucocele diagnosis with a sensitivity of 83% and a specificity of 92% [33].

CT is regarded as the most accurate method of diagnostics. CT can be used to discover the signs specific to mucocele with high accuracy: appendix lumen more than 1.3 cm, its cystic dilatation, and wall calcification [9,21,22,34,35,36].

Colonoscopic findings show " Volcano sign", the appendiceal orifice seen in the center of a firm mound covered by normal mucosa or a yellowish, lipoma-like sub mucosal mass [37].

Patient may present with right lower quadrant pain, change in bowel habits, per rectal bleeding or a palpable mass. Approximately 23-50% of patients are asymptomatic, with the lesions being discovered incidentally during surgery, radiological evaluations or endoscopic procedures [38,39,40]. So preoperative diagnosis of appendiceal mucoceles are difficult because of lack of clinical symptomatology.

Surgical excision of mucocele of an appendix can be done either by laparotomy or laparoscopic surgery. Laparoscopic ally, careful handling of the specimen is recommended as spillage of contents can lead to pseudomyxoma peritonei. This can be achieved by atraumatic handling of the appendix and use of endobags for removal of specimens. Conversion to laparotomy should be considered if the lesion is traumatically grasped or if the tumor clearly extends beyond the appendix or if there is evidence of malignancy such as peritoneal deposits, [41]. Laparoscopic approach has an increased risk of rupture and subsequent pseudomyxoma peritonei formation[42,43,44]. Moreno et al [43]suggest conversion to an open appendectomy in case of mucocele when laparoscopic appendectomy is intended. Some authors still recommend a minimally invasive approach in selected patients for this rare entity [45, 46, 47].

Involvement of caecum or adjacent organs is an indication for right hemi-colectomy and thorough exploration of the gastrointestinal tract and ovaries [48]. Complications for appendiceal mucoceles consists of inflammation, invagination, intussesception, obstruction, bleeding or fistula formation [46]. A right hemicolectomy is a frequently performed if a malignant cause is suspected based on imaging or on intraoperative frozen section [49]. Right hemi-colectomy was not performed in these cases because frozen section analysis at the time of surgery showed no malignant characteristics. Since the risk of developing an adenocarcinoma of the colon is six time greater in patients with a mucocele than in the general population, colonic surveillance is warranted in these cases [50]. The most dreaded complication of benign or malignant mucocele is pseudomyxoma peritonei, which is difficult to treat surgically or medically. It has an uncertain prognosis, with a 5-year survival rate between 53% and 75% [51,52].

An algorithm for selection of the type of surgery has been furnished by Dhage-Ivatury and Sugarbaker [53]. It envisages several factors: (a) whether or not a mucocele is perforated; (b) whether the base of appendix (margins of resection) is involved in the process; and (c) whether there are positive lymph nodes of mesoappendix and ileocolic. As a result patients may require different operations: appendectomy to the right colectomy, including cytoreductive surgery, heated intraoperative intraperitoneal chemotherapy, early postoperative intraperitoneal chemotherapy.

In our patient mucocele was not perforated (no discharge into peritoneal cavity), there was no pathological process in the base of appendix (negative margins of resection), and the regional lymph nodes were negative. Therefore only appendectomy was done and there were no late complications.

Complete abdominal exploration during intraoperative is indicated due to the occurrence of mucocele in synchrony with other tumors, like colon and ovaries. This is necessary when emergency surgery is done with no preoperative workup [54].

Conclusion

Though incidence is very low but diagnosis is very important, may be preoperative or peroperative .

USG or CT findings of a cystic mass in right iliac fossa, adherent to the caecum, in a patient who has not undergone appendectomy, are highly suggestive of mucocele of appendix. Presence of mural calcification can be valuable clue to the diagnosis. The presence of low attenuation ascities with peritoneal implants is suggestive of associated pseudomyxoma peritonei.

Mucocele of the appendix can mimic an adenexal cyst and is difficult to diagnose. In a female presenting with right iliac fossa mass and with no clinical indication of any gynecological pathology , an appendiceal origin should be considered in the differential diagnosis.

Appendiceal mucocele presents a challenge to the surgeon who does not know the pathological diagnosis on the operative procedure. Appendectomy alone should be performed only on mucinous lesions

that are confirmed to be non-neoplastic after biopsy. If appendectomy done, precautions should be taken to minimize the risk of seeding the peritoneal cavity with tumorous mucin during manipulation. Appendectomy whether open or laparoscopic does not guarantee the removal of all neoplastic tissue, including extensions into surrounding tissue and lymph nodes in case of malignant pathology [55,56].

Conflict of Interest

There is no conflict of interest.

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