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Case Report

Pathology Section

Unmasking the Uncommon: A Rare Case Report of Submucosal Lipomatosis of the Appendix Mimicking Acute Appendicitis

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ABSTRACT

Acute Appendicitis (AA) is a common medical emergency that presents with pain in the right iliac fossa and can mimic many other non neoplastic entities that clinically present as acute abdomen, such as intestinal obstruction, adhesions and perforation. Intestinal Submucosal Lipomatosis (SML) is a rare condition with a prevalence rate of 0.2%; however, it is even rarer in the appendix. Although SML can follow a varied course, it most commonly leads to AA or more serious complications like intestinal intussusception or massive intra-abdominal bleeding. Since AA can be a sequela of SML, it can obscure the true cause of inflammation on radiologic imaging due to wall thickening. Therefore, appendectomy, along with Histopathological Examination (HPE), is mandatory for diagnosing Appendiceal Submucosal Lipomatosis (ASL) and preventing complications associated with it. SML can be diffuse or focal and is microscopically characterised by the presence of increased mature adipocytes in the submucosal layer of the gastrointestinal tract, without a capsule—unlike lipoma—making this an important distinguishing feature between the two. Present case is of a 22-year-old male who presented to the surgical outpatient department with complaints of recurrent abdominal pain. Clinically, and even on radiology, it mimicked AA due to wall thickening but was later confirmed as SML of the appendix based on the final HPE report. This case highlights that an accurate preoperative diagnosis of SML, based on radiological findings of fat infiltration in the wall, can avoid laparoscopic appendectomy unless there is some associated complication; in some scenarios, it can also prevent potential complications.

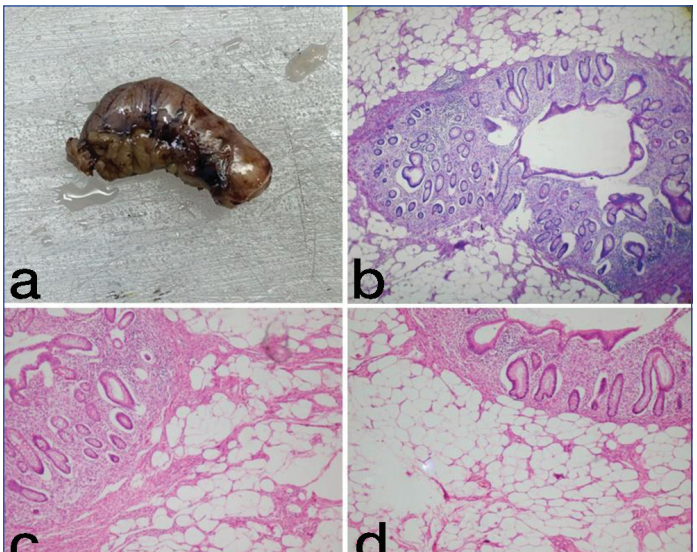
Keywords: Abdominal pain, Appendectomy, Appendiceal submucosal lipomatosis

CASE REPORT

A 22-year-old male presented to the surgical outpatient department with recurrent episodes of abdominal pain for the past two months. There was no history of fever, diarrhoea, nausea, vomiting, or blood in the stools. His bowel habits were normal. On examination, his general physical condition was normal. Localised tenderness was present in the right iliac fossa, with no rebound tenderness noted. There was no past history of any surgeries or co-morbidities. He was advised to undergo an ultrasonography of the abdomen. The sonogram showed evidence of appendicitis, with an appendicular length of 5 cm and a maximum outer diameter of 1.7 cm. A suspicion of perforation near the base, with minimal surrounding collection of 1-2 cc, was noted. There was no significant periappendicular inflammation or enlarged lymph nodes observed. His complete blood count was normal, with a Total Leukocyte Count (TLC) of 4,600/cumm.

He had a similar history two months prior, for which he underwent Contrast-Enhanced Computed Tomography (CECT) of the abdomen. It revealed a heterogeneous ill-defined collection in the right iliac fossa adjacent to the caecum, measuring 58×32 mm. The appendix was contained within this collection and measured 14 mm in caliber, with wall thickening noted. A suggestion of appendicular lump formation was given. He was then treated symptomatically, with a provisional diagnosis of AA, and he was relieved of pain during the first episode. Due to his recurrent episodes, a clinical suspicion of AA with perforation was made in the current episode, leading to Laparoscopic Appendectomy (LA).

Intraoperatively, the appendix appeared elongated and thickened. His postoperative period was uneventful, and the patient was discharged the next day. The appendectomy specimen was sent for histopathology in the Pathology Department. The appendix was grossly enlarged, measuring 5 cm in length with a maximum outer diameter of 1.5 cm. The external surface was congested [Table/Fig-1a]. Upon cutting, the wall was thickened, measuring 0.5 cm and the lumen appeared narrowed due to fat deposition in the appendicular wall. There was no evidence of perforation, appendicolith, or mass found in the appendix. Histopathological examination of the sections revealed a thickened appendicular wall with large areas of mature adipocytic tissue in the submucosa, causing narrowing of the lumen [Table/Fig-1b-d]. No capsule or atypia was identified. The Peyer's patches were markedly thinned out and absent in places. There was no evidence of inflammation, perforation, or fecoliths observed. Based on the above histomorphological findings, a diagnosis of SML of the appendix was made given the absence of a well-defined capsule, unlike in lipomas. There was no atypia or necrosis present. Since the luminal mucosa was unremarkable and



[Table/Fig-1]: a) Whole gross image of intact appendix with slightly distended luminal diameter); b) Low power view showing circumferential involvement. (H&E, 4x); c) Section shows submucosal infiltration by benign mature adipocytes. Luminal lining epithelium is unremarkable (H&E, 40x); d) There is loss of submucosal lymphoid tissue (H&E, 40x).

there was no acute inflammatory infiltrate in the wall, the possibility of AA was ruled out on microscopy. The patient returned to the surgical outpatient department for postoperative follow-up without any complaints and exhibited proper suture healing. The general physical examination was unremarkable, and the follow-up routine Complete Blood Count (CBC) was within normal limits.

DISCUSSION

The ASL is a rare entity that clinically mimics AA, with patients presenting with pain in the right iliac fossa, altered bowel habits, or bleeding [1,2]. It was first identified by Antoci in 1956. AA is a medical emergency with a risk of perforation and peritonitis, commonly treated with laparoscopic appendectomy [3-5]. ASL is a benign entity characterised by the presence of mature adipose tissue in the submucosa of the appendix, causing luminal narrowing and leading to symptoms that may clinically resemble AA. ASL can be treated symptomatically, and laparoscopic appendectomy is usually not indicated [3,5].

Colonic lipomatosis is rare, with a prevalence rate of 0.2%. The incidence is even rarer in the appendix, with only a few case reports mentioned in the literature. Lipomatosis in the intestinal tract presents with non specific findings, such as abdominal pain, diarrhoea, constipation, ileus and bleeding [4,6,7]. Present case presented with recurrent abdominal pain in the right iliac fossa, raising suspicion of AA. On histopathology, a diagnosis of ASL was made with no evidence of inflammation to suggest AA. ASL should be considered as a possibility when suspecting AA. Clinical findings should be correlated with radiographic and laboratory findings in the absence of classic signs of inflammation indicative of AA to avert unnecessary LA.

AA is a common medical emergency caused by mechanical obstruction to the lumen due to fecaliths, *Enterobius vermicularis* infection, lymphoid hyperplasia, or neoplasia. The obstruction causes stasis and increased intraluminal pressure, resulting in inflammation [3,5]. Lipomatosis, or lipohyperplasia, is defined as increased infiltration of well-differentiated mature adipocytes in the submucosal layer of the intestinal tract, with the absence of a capsule, which differentiates it from a lipoma. Lipomatosis can be a rare cause of obstruction due to the mechanical narrowing of the lumen by fat [3,5,6].

Present case is notable in that the lipomatosis presented clinically like AA. However, there were no signs of appendicitis on HPE, including gross evaluation and microscopy. Radiographic scans showed a thickened appendicular wall with suspicion of an appendicular lump and perforation. Nevertheless, there was no periappendicular inflammation or mesenteric lymphadenopathy noted, which are usually associated with AA. Furthermore, the patient's CBC report did not show any leukocytosis. C-reactive protein (CRP) levels were not measured, although they are typically elevated in cases of AA. Very few cases of ASL have been reported in the literature. In most of the retrieved cases, patients were young adults who presented with abdominal pain. Laboratory findings, including CBC and CRP, were within normal limits. A final diagnosis of submucosal lipomatosis was made in both the cited reports and the present case based on histopathology of the appendectomy specimen. In the study by Sanli et al., the diagnosis was made solely using radiological imaging and laparoscopic appendectomy was not performed [7]. This underscores that early diagnosis can prevent laparoscopic surgery in many cases and reduce surgery-related morbidity [8,9].

Although preoperative radiological diagnosis can be challenging, an adequate study of the relevant parameters can help secure a diagnosis of SML [10-12]. There are no specified guidelines for the treatment of intestinal lipomatosis. Treatment ranges from symptomatic management, including hydration and pain relief, to acute surgical exploration. Laparoscopic appendectomy is an invasive procedure with a complication rate of 8.7-13.3% and associated morbidity [3]. It could be avoided in patients without radiographic or laboratory findings suggestive of AA, provided the clinical and laboratory findings are stable [6,7].

CONCLUSION(S)

Though rare, ASL should be considered in the differential diagnosis of AA. Radiography plays an important role in differentiating these entities. A holistic interpretation of radiographic, clinical and laboratory findings is essential to avert unnecessary laparoscopic appendectomy in cases of ASL.

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