

# SF Journal of Radiology and Clinical Diagnosis

## PKB Syndrome – A Case of Appendicaecomucocele

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### Abstract

Mucocele of the appendix is a descriptive term that refers to dilation of the appendiceal lumen as a result of mucin accumulation. Majority of these cases present with mucocele involving whole length of the appendix. On extensive literature search, case of mucocele confined only to the proximal part of appendix with caecum involvement has not been reported. In our article, we present a unique case of mucocele of appendix involving part of caecum and only proximal appendix. This case of mucocele of appendix is unique from previously mentioned cases in available literature in location of involvement. Hence we would like to propose a new nomenclature: "Appendicaecomucocele" for mucocele of appendix involving only proximal part and part of caecum, its imaging finding as PK Sign and whole syndrome as PKB Syndrome after the name of the authors i.e., Patel, Kachewar and Bhandari.

**Keywords:** Appendix; Mucocele; Appendicaecomucocele; PK Sign; PKB Syndrome

### Case Report

A 60 year old male presented with pain in the lower abdomen on right side since last 15 days. No history of Fever or any other complaints. On examination slight tenderness noted over the right lower quadrant. There was no history of any other gastrointestinal, urinary symptoms. There was no significant past history.

### Imaging findings

On Ultrasonography retrocaecal appendix was visualized. AP Diameter of appendix measured 1 cm with a dilated fluid filled proximal portion extending into the caecum.

On CT scan, fluid filled dilated proximal appendix/encysted collection was seen at the junction of appendix and caecum, extending retrocaecally adhering to the inferior margin of liver.

On MR (Figure1), T1-weighted axial MRI shows a well-circumscribed, low signal intensity mass in proximal appendix involving caecum, T2-weighted coronal MRI showing a high signal intensity paracaecal tubular mass with low signal intensity thickened wall superiorly (Figure 1).

The appendix resembles to an inverted Letter 'P' in coronal section (Figure 2), where stalk of the letter is formed by the distal appendix and the head is formed by the cystic collection in the caecum and proximal appendix.

Patient was operated and surgical resection of appendix and involved part of caecum was performed.

On histopathological examination (Figure 3) appendix measured 3.5x1 cm, was congested and tense. On opening the pouch like structure at the junction of appendix and caecum, mucinous material was noted. Microscopically (Figure 4). The proximal part of the appendix showed flattened basal epithelial cells with mucinophages. Muscularis mucosa appeared infiltrated by mucinous material. Sections from Distal end of appendix did not show any pathology.

Therefore confirming the radiological the diagnosis of mucocele of appendicular and caecal origin.

### Discussion

Mucocele of the appendix is a descriptive term that refers to dilation of the appendiceal lumen as a result of mucin accumulation and is based on the gross or macroscopic appearance of the appendix. Mucocele usually involves whole of the appendix. Mucocele formation is most commonly caused by epithelial proliferation, either benign or malignant. Much less frequently, inflammatory or obstructive causes, to include appendicitis and obstruction by a fecolith or appendicolith, are the

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Received Date: 28 Feb 2021

Accepted Date: 05 May 2021

Published Date: 10 May 2021

**Citation:** Patel DK, Kachewar GK, Bhandari V. PKB Syndrome – A Case of Appendicaecomucocele. *SF J Radiol Clin Diagn.* 2021; 3(1): 1006.

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**Figure 1:** T2-weighted coronal MRI showing a high signal intensity paracaecal tubular mass.



**Figure 2:** T2-weighted coronal MRI showing a high signal intensity paracaecal tubular mass (Arrow- inverted P appearance =PK sign).

cause of mucocele formation [1].

The most common clinical manifestation is a palpable mass in the lower right quadrant of the abdomen; the patient can present either with or without pain. If a super infection is present, mucinous cystadenoma may be clinically indistinguishable from nontumoral acute appendicitis [2]. Some patients are asymptomatic; in these cases, mucoceles are found incidentally when imaging is performed for other reasons or during a laparotomy or laparoscopy [3].

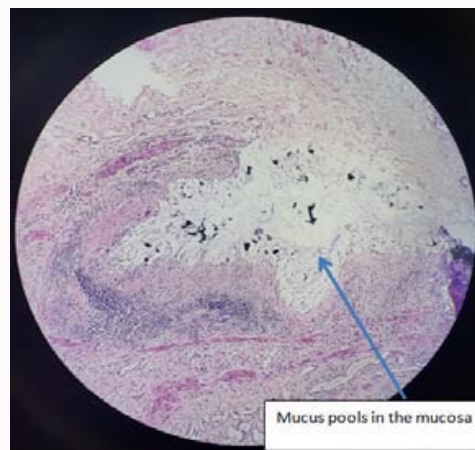
Majority of the appendiceal mucocele cases reported in literature described mucocele involving whole length of appendix. There are few case reports of mucocele of appendix sparing the base [4].

Our case of mucocele was unique in that only proximal part of appendix with part of caecum was involved with distal portion showing no abnormal features. To the best of our knowledge, no particular name is given to this type mucocele, hence the authors would like to propose a new term : ‘Appendiceaecomucocele’.

The Coronal section of CT & MRI shows inverted letter ‘P’ appearance as described above. Authors would like to name this appearance on Coronal CT/MRI scan as ‘PK Sign’ and the whole syndrome of Right lower abdominal pain and tenderness, with PK



**Figure 3:** Gross Specimen showing pouch like structure at the junction of appendix and caecum measuring 3.5x1 cm.



**Figure 4:** Microscopically-The proximal part of the appendix showed flattened basal epithelial cells with mucinophages. Muscularis mucosa appeared infiltrated by mucinous material. Sections from Distal end of appendix did not show any pathology.

Sign on coronal Section of CT/MRI due to appendiceaecomucocele as ‘PKB Syndrome’ after the name of the authors i.e., Patel, Kachewar and Bhandari. Such naming is permissible and is in better tradition of scientific nomenclature [5].

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