

中文題目：闌尾低度惡性黏液瘤伴隨腹膜偽黏液瘤之內視鏡觀點 – 病例報告

英文題目：Endoscopic viewpoint of low grade appendiceal mucinous neoplasm with pseudomyxoma peritonei: A case report

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## **Introduction:**

Appendiceal mucocele is a rare disease described obstructive luminal dilatation of the appendix caused by accumulation of mucus. Additionally, mucinous appendiceal neoplasms are subclassified as low-grade appendiceal mucinous neoplasms (LAMNs), high-grade appendiceal mucinous neoplasms (HAMNs) and mucinous adenocarcinomas by their histological and invasion grading. Once the appendiceal mucinous neoplasms perforate and spread to the peritoneal cavity, it was regarded as pseudomyxoma peritonei (PMP). Herein, we reported a case of LAMN complicated with PMP presented with atypical endoscopic features.

## **Case presentation:**

A 51-year-old male without systemic disease complained of abdominal fullness for 4 days. He visited local clinic where ascites was found by ultrasound then was transferred to our hospital. Abdominal CT scan revealed multiloculated massive ascites and appendiceal tumor with pseudomyxoma peritonei was suspected. (Fig. 1) Lab data showed elevated CA19-9 (392.1 U/mL) and CEA (9.0 ng/mL) level. Colonoscopy illustrated wide appendiceal opening with mucus efflux. We probed the depth of the appendix by biopsy forceps but there was no definite mass touched. (Fig. 2) The patient underwent laparoscopic right hemicolectomy and much ascites as whitish and translucent jelly like material were observed over all peritoneum, mainly over appendix, ascending colon and causing severe adhesion over upper abdomen. (Fig. 3) (Fig. 4) The pathology disclosed LAMN with pseudomyxoma peritonei with no evidence of invasion over appendix, ascending colon, ileum and peritoneum. Afterward, the patient went to another hospital in Kaohsiung for cytoreductive surgery (CRS) and heated intraperitoneal chemotherapy (HIPEC). Due to EGFR presented overexpression (3+) and RAS wild type, the patient subsequently received cetuximab plus FOLFOX.

## **Discussion:**

Appendiceal mucinous lesions may be found incidentally during radiologic or colonoscopy examination. In ordinary, the endoscopic features of appendiceal mucocele presented as a round, balloon-shaped protruding mass from the appendiceal orifice, along with moving in and out with respiration. The appendiceal orifice could be seen at the center of the cecal mound, as known as “Volcano sign”. When encountered an extrinsic or submucosal lesion over the appendiceal orifice or cecum under colonoscopy, we could poke the lesion with biopsy forceps, the appendiceal mucocele may characterized as firm or soft lesion with cushion sign. In general, mucosal biopsy should be avoided because the overlying mucosa is frequently normal and for fear of rupture with peritoneal spreading.

In our case, the patient was a case of LAMN with PMP. The colonoscopy showed no typical protruding mass nor volcano sign over cecum but wide appendiceal opening with mucus efflux. We compared with the

CT scan, no obviously appendiceal mass but a dilated, elongated shape appendix was found, compatible with endoscopic finding of no definite mass touched by forceps probing and specimen after right hemicolectomy.

**Conclusion:**

The mucinous appendiceal neoplasms may be incidental found on radiographic images or colonoscopy. We highlight the endoscopic viewpoint of diagnosis of mucinous appendiceal neoplasms. The typical features presented as a round, protruding mass from the appendiceal orifice, along with moving in and out with respiration. Once rupture of mucinous appendiceal neoplasms or presented as pseudomyxoma peritonei, typical features may not be seen under endoscopy. Otherwise, wide opening of appendiceal orifice with mucus efflux may be one of the possible endoscopic feature.