

## MUCINOUS CYSTADENOMA OF THE APPENDIX ASSOCIATED WITH MUSCULAR AND NEUROMATOUS HYPERPLASIA: REPORT OF A CASE

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### ABSTRACT

Mucinous cystadenoma of the appendix is a type of appendiceal mucocele and a rather rare condition usually found incidentally in the course of other abdominal surgery. A previous evaluation of 73 appendiceal mucoceles showed that this disease was often associated with adenocarcinoma and other epithelial atypia. This observation suggested that patients with mucinous cystadenoma of the appendix also have some underlying disorders. However, non-epithelial changes associated with appendiceal mucocele have not been reported so far. In this study, we presented for the first time a mucinous cystadenoma of the appendix associated with muscular and neuromatous hyperplasia.

Key Words: Mucinous cystadenoma, Muscular and neuromatous hyperplasia, Appendix

### INTRODUCTION

Mucinous cystadenoma of the appendix is a type of appendiceal mucocele and a quite rare condition that is incidentally discovered in the course of other abdominal surgery. It presents no symptoms despite cystic dilatation of the appendix lumen with stasis of mucus.<sup>1,2)</sup> As a result of a latency of its detection, a rupture of mucinous cystadenoma may occur, resulting in pseudomyxoma peritonei, a condition involving the spread of adenoma cells throughout the peritoneal cavity in the form of multiple mucinous deposits.<sup>3,4)</sup>

The previous evaluation of 73 appendiceal mucoceles showed that this disease was often associated with adenocarcinoma and other epithelial atypia.<sup>5)</sup> This observation suggested that patients with mucinous cystadenoma of the appendix harbor some disorders. However, non-epithelial changes associated with appendiceal mucocele have not been reported so far. In this study, we presented mucinous cystadenoma of the appendix associated with muscular and neuromatous hyperplasia for the first time.

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## CASE REPORT

A 33-year-old woman was admitted to our hospital on April 12, 2006 with a right ovarian cyst. A laparoscopic right ovarian cystectomy performed on April 13 unexpectedly showed a swollen appendix. Although we tried, we could not remove the appendix by laparoscopic surgery at that time, because it was firmly imbedded in the retro-peritoneum.

To evaluate the appendiceal mass on May 11, we first performed abdominal CT scanning that revealed a cystic mass (8 cm × 3 cm in diameter) in the right lower abdominal cavity (Fig. 1a). Although we could not make a definitive diagnosis that the mass was an appendix, we thought that it might be an appendiceal mucocele since it matched the appearance of the mass as described in the laparoscopy. We next performed a total colonoscopy on May 15, 2006. However, no symptom was observed at the appendiceal base, suggesting that the mucocele remained within the appendix. To further confirm that the cystic mass was an appendiceal mucocele, we performed an abdominal MRI on May 27 when the same cystic mass was observed (Fig. 1b).

An appendectomy was performed on June 22. Laparotomy showed the swelled appendix was imbedded in the retro-peritoneum. We carefully separated the swollen appendix from the retro-peritoneum and mesoappendix and successfully removed it without a rupture (Fig. 2a, b). The patient was discharged from our hospital in good general condition on June 29.

The surgically resected appendix was examined by an expert pathologist (H. Shibata). The appendiceal mucocele was determined to be a mucinous cystadenoma enclosing the accumulated mucus (Fig. 3a). The cystadenoma itself was also surrounded by muscular and neuromatous hyperplasia (Fig. 3b).

## DISCUSSION

Appendiceal mucocele is a descriptive term that implies a dilated appendiceal lumen caused by the accumulation of mucus.<sup>6,7)</sup> Although a proper preoperative diagnosis is recommended, fine needle biopsy should not be performed because of the risk of pseudomyxoma dissemination. For the same reason, though the diagnostic therapy should be surgical, a laparoscopic approach is not always advised.<sup>8)</sup> Appendectomy by laparotomy is advised for focal or diffuse mucosal hyperplasia and cystadenoma when the appendiceal base is intact. Cecal resection is performed for a cystadenoma with a large base, with a right colectomy recommended for a cystadenocarcinoma. In cases of disseminated pseudomyxoma peritonei, an ultrasonic surgical aspirator can be used.<sup>9,10)</sup>

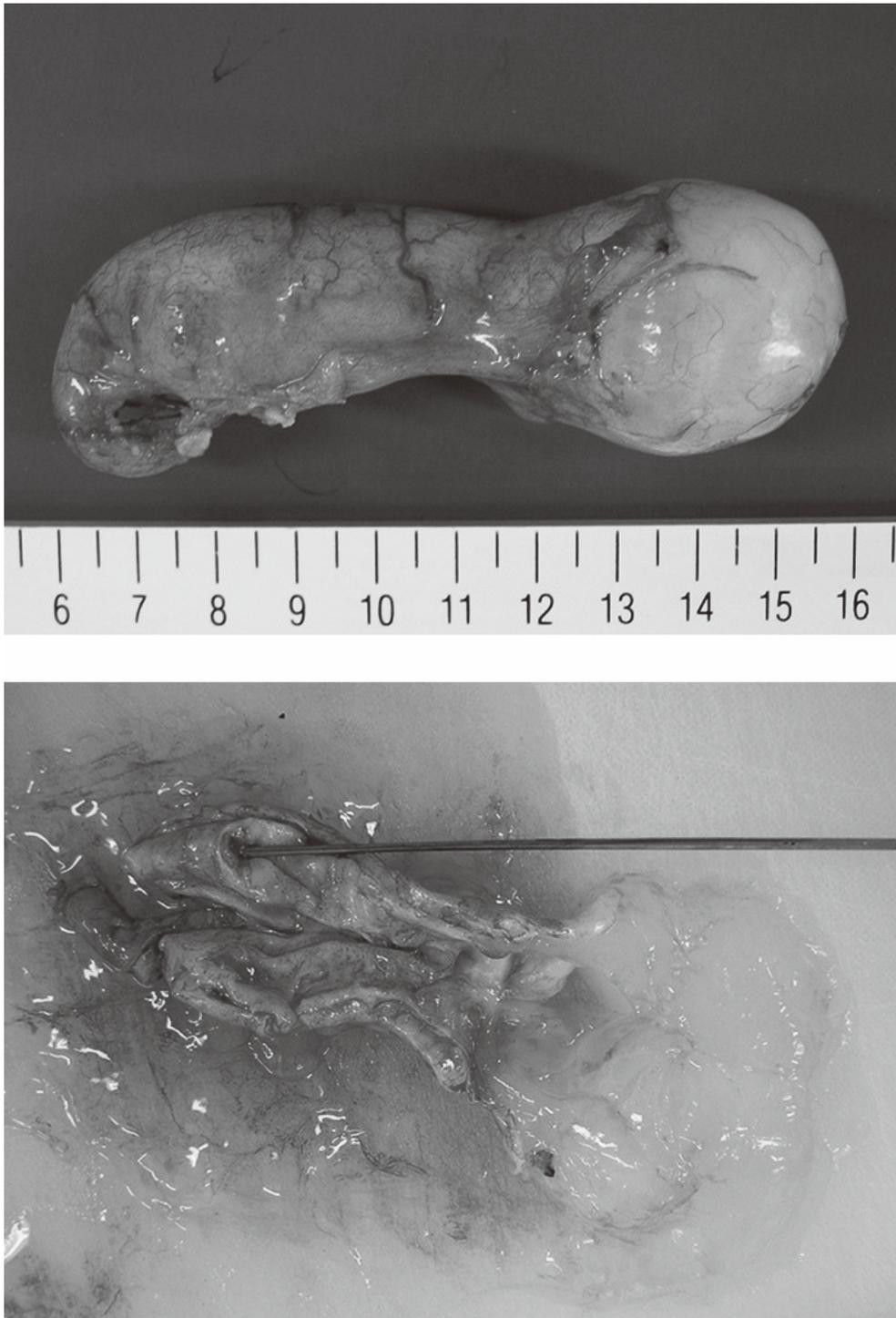
Higa *et al.* evaluated 73 appendiceal lesions fulfilling the criteria of a so-called `mucocele`. They are comprised of the following 3 distinctive clinicopathologic entities: focal or diffuse mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma.<sup>5)</sup> Of the 46 cases with mucinous cystadenoma, 9 were associated with adenocarcinoma of the colon, and 4 with ovarian mucinous cystoma. Among the 18 cases with focal or diffuse mucosal hyperplasia, 5 were associated with adenocarcinoma of the colon, and 1 with ovarian mucinous cystoma. As we report here, an appendiceal mucocele is often associated with other neoplasias, especially adenocarcinomas of the colon and ovaries. Djuranovic *et al.* have also recently reported a mucinous cystadenoma of the appendix associated with adenocarcinoma of the sigmoid colon and hepatocellular carcinoma.<sup>11)</sup> Taken together, an appendiceal mucocele would often be associated with other neoplasms, suggesting that these patients may have an underlying condition such as a genetic disorder.

In this study, we presented for the first time a mucinous cystadenoma of the appendix associ-

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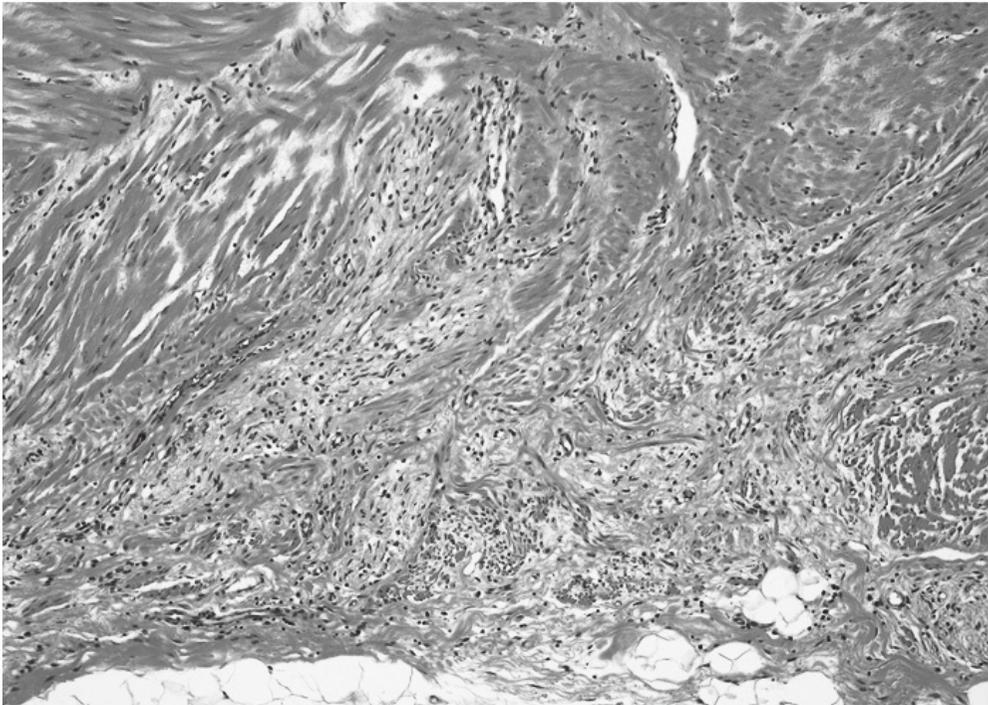


**Fig. 1** **a** CT of sacral space. A cystic mass (8 cm × 3 cm in diameter) was observed in the right lower abdominal cavity. **b** MRI of sacral space. The same cystic mass was observed.



**Fig. 2** **a** The surgically resected appendix. Massive mucus had accumulated. **b** Opened appendix.

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**Fig. 3** a Mucinous cystadenoma of the appendix surrounding accumulated mucus. (Hematoxylin and eosin,  $\times 100$ )  
b Muscular and neuromatous hyperplasia of the appendix. (Hematoxylin and eosin,  $\times 100$ )

ated with muscular and neuromatous hyperplasia. Until now, non-epithelial changes associated with appendiceal mucoceles have not been reported. Further studies such as a genetic examination may reveal the pathogenesis of an appendiceal mucocele.

## REFERENCES

- 1) Minni F, Petrella M, Morganti A, Santini D, Marrano D. Giant mucocele of the appendix: Report of a case. *Dis Colon Rectum*, 2001; 44: 1034–1036.
- 2) Rampone B, Roviello F, Marrelli D, Pinto E. Giant appendiceal mucocele: Report of a case and brief review. *World J Gastroenterol*, 2005; 11: 4761–4763.
- 3) Crawford J. Tumors of the appendix. In *Pathologic basis of disease*, edited by Cotran R, Kumar V, Robbins S. pp. 824–825, 1994, Philadelphia Saunders.
- 4) Landen S, Bertrand C, Maddern GJ, Hreman D, Pourbaix A, de Neve A, Schmitz A. Appendiceal mucoceles and pseudomyxoma peritonei. *Surg Gynecol Obstet*, 1992; 175: 401–404.
- 5) Higa E, Rosai J, Pizzimbono CA, Wise L. Mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma of the appendix. *Cancer*, 1973; 32: 1525–1541.
- 6) Kim SH, Lim HK, Lee WJ, Lim JH, Byun JY. Mucocele of the appendix: ultrasonographic and CT finding. *Abdom Imaging*, 1998; 23: 292–296.
- 7) Krebs TL, Daly BD, Wong-You-Cheong JJ, Grumbach K. General case of the day. Mucinous cystadenocarcinoma of the appendix. *Radiographics*, 1998; 18: 1049–1050.
- 8) Gonzales Moreno S, Shmookler BM, Sugarbaker PH. Appendiceal mucocele. Contraindication to laparoscopic appendectomy. *Surg Endosc*, 1998; 12: 1177–1179.
- 9) Keating JP, Frizelle FA. Use of ultrasonic surgical aspirator in operative cytoreduction of pseudomyxoma peritonei. *Dis Colon Rectum*, 2000; 60: 341–343.
- 10) Zagrodnik DF 2nd, Rose DM. Mucinous cystadenoma of the appendix: diagnosis, surgical management, and follow-up. *Curr Surg*, 2003; 60: 341–343.
- 11) Djuranovic SP, Spuran MM, Kovacevic NV, Ugljesic MB, Kecmanovic DM, Micev MT. Mucinous cystadenoma of the appendix associated with adenocarcinoma of the sigmoid colon and hepatocellular carcinoma of the liver: Report of a case. *World J Gastroenterol*, 2006; 12: 1975–1977.