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A Rare Case of Appendiceal Mucocele with Myxoglobulosis

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ABSTRACT

A 63-year-old man was admitted with an abdominal mass in the right lower quadrant. The tumor was diagnosed as mucocele of the appendix after admission and surgically removed. The appendix containing gelatinous mucus with numerous round bodies was distended to the size of an egg. Therefore, a diagnosis of myxoglobulosis was postoperatively made. This is a rare disease and the diagnosis has been preoperatively made in none of patients in Japan. The present case could present an echographic pattern characteristic of this cyst. Studies on all available echograms from previous patients could provide information necessary for preoperative diagnosis of myxoglobulosis.

Myxoglobulosis has been considered to be a variant of mucocele of the appendix. It is called a "frog egg mucocele" or a "jelly belly mucocele" based on its characteristic appearance. In Europa and America, the first autopsy case was reported by Latham et al[7] in 1897 and a total of 45 cases were reported as of 1985. In Japan, 53 cases, including the first case reported by Sato[11] in 1910 and the present case, were reported. According to Dannreuther[4], mucoceles were found in 8 (0.095%) of 8457 excised specimens of the appendix. The incidence of myxoglobulosis in mucoceles varies widely between 0.35% to 8% reported by Millikan et al[8] and Felson et al[5], respectively. These rates indicate that there is one case of myxoglobulosis among about 13,000 to 300,000 patients who undergo appendectomy. Although essentially no case of myxoglobulosis has been diagnosed before operation, the echographic study on myxoglobulosis in the present case has shown that this lesion can be identified before operation. The following is an outline of the case.

CASE REPORT

The patient was a 63-year-old male Japanese office worker. Neither his family history nor previous illness was remarkable. On September 25, 1985, he was admitted to Hiroshima University Hospital for medical examination for persistent anorexia from the summer. His

Table 1

RBC	445 × 10^4	Hbs −Ag (−)
		Lues (−)
Hb	14.0 g/dl	CRP (−)
WBC	4800	AFP < 5 ng/ml
Plt	218000	CEA 1.0 ng/ml
		CA19-9 < 6 U/ml
TP	6.9 g/dl	
A/G	3.8	
TB	0.5 mg/dl	S Amylase 129 U
GOT	16 U/liter	T cholesterol 214 mg/dl
GOT	13 U/liter	Trigliceride 291 mg/dl
LDH	279 U/liter	
ChE	322 U/liter	Urine protein (−)
γ-GTP	22 U/liter	Urine sugar (−)
TTT	1 U	Stool occult blood (−)
2nTT	5 U	Stool parasite eggs (−)
S Amylase	129 U	

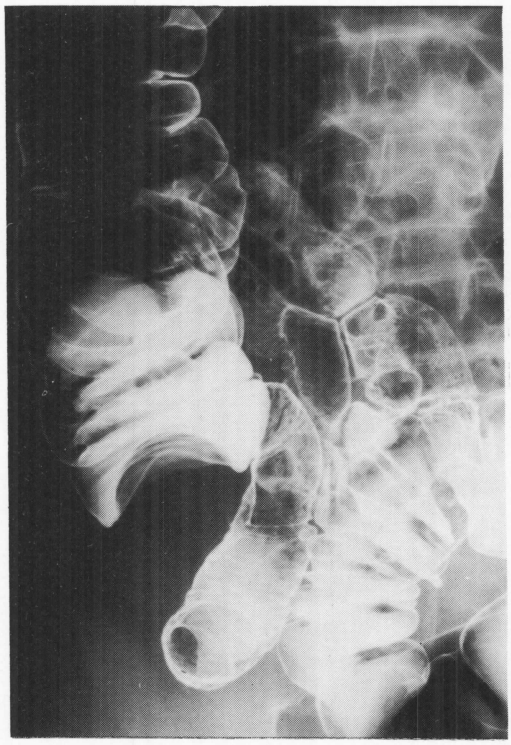

Fig. 1. Radiogram by barium-enema demonstrating replacement of the cecum with a mucocele of the appendix. The appendix is not visualized.

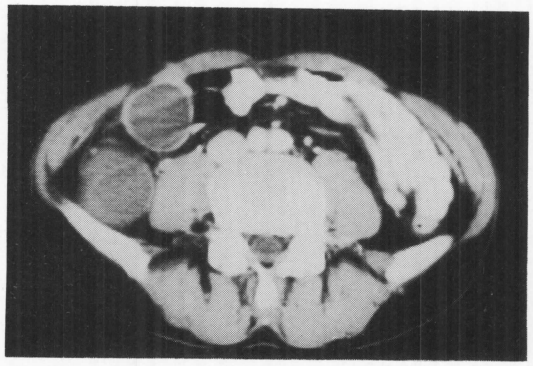

Fig. 2. CT scan demonstrating two cysts in the ileocecal region. The two are connected in a U-shape.

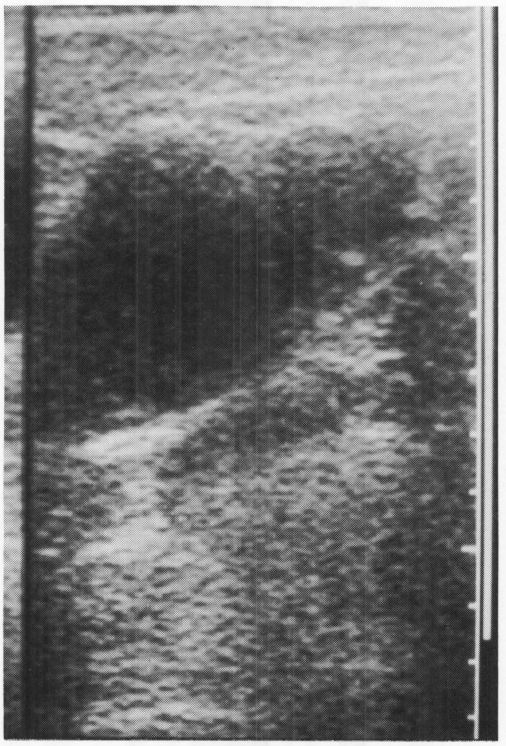

Fig. 3. Echographic appearance of a cyst close to the cecum. A pale uneven shadow like a honeycomb is seen in the cystic cavity. The pattern was swinging.

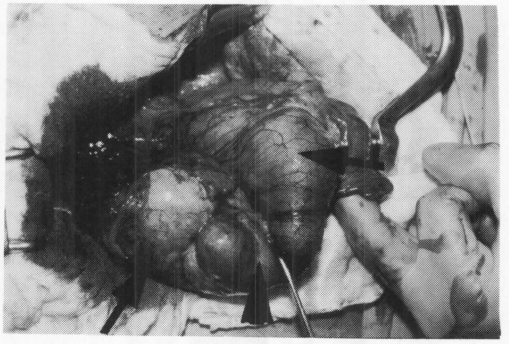

Fig. 4. A view through the surgical opening demonstrating the distal end of the ileum.
↑ : mucocele ▲ : constriction

laboratory data on admission are summarized in Table 1. After admission, an egg-sized tumor in the lower quadrant was pointed out and a diagnosis of mucocele of the appendix was made after a barium-enema examination (Fig. 1), CT (Fig. 2), and ultrasonic echography (Fig. 3). Although he was afebrile throughout the course and did not complain of abdominal pain, he underwent surgery on October 15 because possibility of mucinous carcinoma of the appendix could not be excluded.

A right pararectal incision was made for the laboratomy. There was neither ascites nor

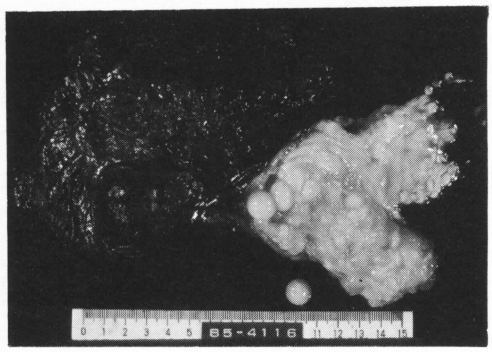

Fig. 5. Contents of a mucocele. Round bodies in the proximal cyst were mostly bigger than those in the distal one.

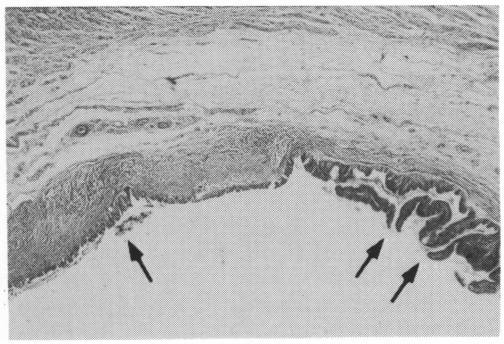

Fig. 6. Microscopic view of the cystic wall. ↑ : A wall lacking mucosal layer comprises granulation tissue containing inflammatory cells and cells induced by foreign bodies. Calcification are seen in some parts. ↑↑ : Tall columnar cells containing mucus form stratified papillary projections. There is little evidence of atypia. No invasion upon the wall is seen.

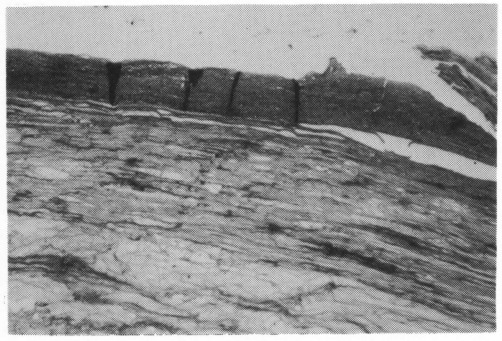

Fig. 7. A microscopic view of the round bodies. PAS stain. A stratified structure like an onion is seen.

mucinous material in the peritoneal cavity. No metastatic nodule was palpable in the liver. The appendix was constricted in the middle forming a U-shape and as large as an egg. The dorsal mass was pressing the cecum anteriorly at an angle (Fig. 4). The dilated appendix was removed with the ileocecal region, although no enlarged lymph node was found. Amorphous or sandy gelatinous substance as shown in Fig. 5 was contained with 23 soft globoid bodies in the cavity. The globoid bodies ranged from 4 mm to 10 mm in diameter and looked like pearls. Most of the globoid bodies which were larger than 5 mm in diameter were found in the cystic cavity near to the cecum. The globoid bodies decreased in size and gelatinous substance, both sandy and amorphous, increased towards the distal end. There was no visible communication between the cyst and the cecum. The cystic wall comprised tissues of two histologic types. One comprised a connective tissue indicating a remarkable inflammatory reaction without the mucous epithelium. The rest of the wall showed benign papillary projections of the mucous epithelium (Fig. 6). The contents of the round bodies were stained by both PAS and alcian blue. A stratified structure like onion was visible by microscopy (Fig. 7).

DISCUSSION

Many reports have dealt with the etiology of this disease. All of them consistently described this disease as a variant of mucocele of the appendix. The requisite conditions for the formation of mucocele of the appendix are as follows. 1) The appendix is obstructed at the basement. 2) The appendicular mucus glands are active. 3) The appendicular cavity contains no fecal matter. 4) The appendicular lumen is sterile. The genesis of the small globoid bodies as found in the present case has long been discussed. According to Cagnetto (1909)[3], minute particle of mucus is first formed in the glands of the appendicular mucosa and the particle grows into small globoid bodies in the cystic cavity. Although the precise mechanism remains to be clarified yet, this speculation might present a right sketch of the genesis. According to Rubnitz and Harmann[10], mucocele and myxoglobulosis were experimentally produced in rabbits readily by ligating the proximal appendix. This

result may support the righteousness of the sketch described above. In Japan, also, Yoshinaga (1941)[12] succeeded in producing the small globoid bodies in the appendix in rabbits. Small globoid bodies were often formed in a few days when appendicostomy was added to the distal end of the appendix ligated at the base. This indicates that concentration of appendicular contents plays an important role in the etiology. Although neither communication between the appendix and cecum nor appendicular perforation was visible in the present case, there should be some concentration of appendicular content.

The series of 53 cases of myxoglobulosis occurring in Japan range from 21 to 83 years in ages of the onset. The cysts arose their 20s in 8, 30s in 10, 40s in 9, 50s in 8, 60s in 12, 70s in 3, and 80s in 1. The distribution was essentially even between ages from 20 to 80 years. This series comprised 44 males, 7 females and 3 of unknown sexes; male patients were predominant. Their preoperative diagnoses were acute or chronic appendicitis in 28, abdominal tumors and invagination in 9 cases each, ileus in 2, and unavailable in 3. Myxoglobulosis was found by chance during operation for other diseases in 3 cases, in which appropriate diagnoses were made postoperatively. As far as we investigated, the preoperative diagnosis of myxoglobulosis was made only in one case by Alcalay et al[1] depending on multiple calcified round bodies in the ileocecal region on plain abdominal X ray photograph. Their calcification, however, rarely occurs. Only 3 cases of calcification in the ileocecal region, besides one described above, have been previously reported[2,5]. Calcification was not found in none of the Japanese cases, possibly because these patients underwent operation under diagnoses of appendicitis without preoperative abdominal examination by X ray.

Preoperative echography was carried out in 3 including the present case in Japan. Kawamoto et al[6] and Ohno et al[9] noticed a cystic pattern encircling a pale, fine honey-comb echographic pattern. A similar honey-comb pattern was also noted in the present case. Moreover, the pattern showed liquidity when observed carefully. The echographic fluidity of fine honey-comb pattern in combination with appropriate clinical symptoms allows us to make a right diagnosis of myxoglobulosis before operation.

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