MUCOCELE OF THE APPENDIX

BY

J. H. JOHNSTON, M.B., F.R.C.S., F.R.C.S.I.

Senior Surgical Registrar, Liverpool Royal Infirmary

Mucocele of the appendix is an uncommon condition. Woodruff and McDonald (1940) reported its incidence as 0.3% of appendicectomies in the Mayo Clinic over a period of 24 years. The disease was described by Rokitsansky in 1842 and by Virchow in 1863. Werth in 1884 first described pseudomyxoma peritonei associated with cystadenoma of the ovary, and in 1901 Fraenkel recorded the same condition in a case of appendix mucocele.

Case 1

A man aged 76 was admitted to hospital in October, 1952, complaining that for 24 hours the rupture in his right groin had been painful and irreducible and that he had been vomiting. There were the typical signs of a strangulated right inguinal hernia, but rectal examination revealed the presence of a large firm swelling in the rectovesical pouch. At operation the hernial sac was found to contain strangulated but viable small intestine. The bowel was reduced and the abdomen explored by a finger inserted through the neck of the sac. A firm smooth mass, which was palpable, extended from the right iliac fossa into the pelvis. The peritoneal cavity was opened through a transverse incision above the inguinal canal, disclosing the mass, which proved to be a large mucocele of the appendix. It was delivered with some difficulty, as it was adherent to its surroundings, and was removed. The hernia was repaired and the incision closed. On subsequent questioning the patient denied any history of previous abdominal pain or dyspepsia. He collapsed and died suddenly of a coronary thrombosis on the fourth day after operation.

The appendix (see illustration) measured 6½ in. (15.8 cm.) in length and 7½ in. (19 cm.) in its greatest circumference. It contained a colourless jelly-like substance, and the lumen

Photograph of the appendix in Case 1.

was completely occluded at the base by a short fibrous stricture. Histological examination showed that the wall was only 1/16 in. (1.6 mm.) thick. The serous coat was normal, but the muscle was entirely replaced by hyaline connective tissue with many cholesterol clefts. The lining epithelium consisted of a single layer of tall columnar cells secreting mucus.

Case 2

A woman aged 38 was admitted to hospital in June, 1951, with a history of central abdominal pain and vomiting of four days' duration and more recent pain in the right iliac fossa. The temperature was 100.2° F. (37.9° C.) and the pulse rate 90 a minute. There were tenderness and muscle guarding in the right iliac fossa and hypogastrium; rectal examination revealed tenderness of the pelvic peritoneum but no palpable tumour. The pre-operative diagnosis was appendicitis with perforation and peritonitis. At operation thin pus was found in the peritoneal cavity. The appendix was much enlarged and was surrounded by mucoid material,

which was also present in the pelvis. The left ovary contained a small cyst. Appendicectomy was performed and the abdomen drained. Recovery was uneventful. Section of the specimen showed that a pre-existing mucocele of the appendix had become acutely inflamed and had ruptured.

In September, 1952, 15 months after appendicectomy, the patient was readmitted complaining of swelling of the abdomen which had been progressing for nine months. On examination there was a central cystic tumour in the hypogastrium extending from the pubis to the umbilicus. Pelvic examination revealed that the uterus was of normal size; the large cyst was palpable through the left anterior fornix, and a smaller cyst was found in the right appendage. At operation the peritoneal cavity contained large quantities of mucoid substance which was exuding from a perforation of a multilocular cyst of the left ovary. The right ovary contained an endometrial cyst. Bilateral salpingo-oophorectomy was carried out. Histological examination showed that the large ovarian cyst was a pseudomucinous cystadenoma. Subsequently radiotherapy was applied to the abdomen.

Discussion

The essential factors in the development of appendix mucocele are obstruction of the lumen and sterility of its contents. In most cases the obstructing agent is a fibrous stricture at the base of the appendix which completely occludes the lumen and is the result of former appendicitis. On occasion the lumen is narrowed but not totally obliterated, since a mucocele may fill with barium in the course of an opaque-meal examination (Vorhaus, 1930). Wilson (1950) estimated the degree of occlusion necessary for the formation of appendiceal diverticula and found that an internal circumference of 4 mm. was the critical level at which obstruction and retention of secretions occurred. Wangenstein *et al.* (1937) recorded a resistance to the flow into the caecum of fluids injected into an appendicostomy and suggested the possibility of functional obstruction due to a sphincter mechanism at the appendix base. Carcinoid tumour (Topping, 1937), carcinoma of the caecum (Muir, 1931), and endometriosis (Shemilt, 1949) are rare causes of the condition.

The appendix contains a jelly-like material, pseudomucin, which is a glucoprotein distinguishable from mucin by reduction tests and by staining with mucicarmine. Its presence may be due to abnormal secretion by the mucosa or, since all gradations of staining and reduction reactions occur, the change from mucin to pseudomucin may be a gradual extracellular one. The content of a mucocele is usually sterile on culture. Wells (1936) produced the condition in rabbits by ligation of the appendix base, leaving the blood supply intact. In the experiments of Cheng (1949) and Grodinsky and Rubnitz (1941), however, simple ligation resulted in acute appendicitis, and development of mucocele required preliminary sterilization of the lumen by irrigation with saline, penicillin, or sulphaguanidine solutions. In long-standing cases the pseudomucin becomes inspissated, and calcification may lead to the formation of a large appendiceal calculus (Bunch, 1945).

In experimental mucoceles the muscular coat is hypertrophied in the early stages, but in the fully developed condition the muscle is thin and almost, or entirely, replaced by fibrous tissue, which may become hyalinized and eventually calcified. Diverticulum formation is common and is probably an early occurrence, as Wilson (1950) has shown that an actively contractile muscularis is essential to raise the intra-appendicular pressure and that passive distension of the appendix is ineffective.

Pseudomyxoma peritonei, which also follows rupture of a pseudomucinous cyst of the ovary, is the most important complication of appendix mucocele and is characterized by the presence of masses of colloid in the peritoneal cavity either lying free or enclosed in membranous adhesions. This material is identical with the contents of the mucocele and may be localized to the region of the appendix or widespread. The initial cause of the condition is perfora-

tion of the appendix, usually through a diverticulum, but the large quantities of pseudomucin which may be found have led to speculation concerning its origin. There are three possibilities.

1. The serosal cells may be stimulated by the presence of colloid to secrete an identical substance. This seems unlikely. Trotter (1910) stated that the peritoneal lining cells could assume columnar for as but believed that their reaction was of the nature of a foreign-body peritonitis and limited to the formation of enclosing adhesions.

2. The peritoneal pseudomucin may result from its continued discharge from a perforation of the mucocele, but if this were so the complication would be halted by removal of the appendix. It may, however, recur after appendectomy and eventually lead to the death of the patient.

3. The most probable explanation, in both appendiceal and ovarian types, is that mucus-secreting cells escape into the peritoneal cavity and, becoming implanted there, continue to secrete. Such cellular elements are difficult to find histologically in the peritoneal colloid, but the hypothesis is supported by the experiments of Grodinsky and Rubnitz (1941), who injected the contents of mucoceles into the peritoneal cavities of other animals and produced progressive pseudomyxoma peritonei, while if the contents were first passed through a Seitz filter to remove the cells the condition did not develop.

Woodruff and McDonald (1940), in a study of 146 cases, divided appendix mucocele into two groups. In the first the mucosa consists of a single layer of columnar cells. Rupture of such a mucocele leads to a localized type of pseudomyxoma peritonei which does not recur after appendectomy. In the second, smaller group the mucosa is hyperplastic owing to the presence of papillae covered with large columnar cells containing hyperchromatic nuclei. This is regarded as a grade 1 adenocarcinoma, and perforation causes dissemination of malignant cells and a diffuse, progressive peritoneal lesion.

Pseudomyxoma peritonei of appendiceal or ovarian origin differs, therefore, only in degree of malignancy from the coelomic spread of colloid carcinoma of the alimentary tract.

The occurrence of mucocele of the appendix and pseudomucinous ovarian cyst in the same individual has been recorded many times. Bailey (1916) and Ries (1924) described cases with both lesions present simultaneously and discharging colloid into the peritoneal cavity. In Eden's (1912) patient an appendix mucocele was found two years after operation for a ruptured pseudomucinous cyst. The double condition is usually complicated by pseudomyxoma peritonei, and it is likely that one lesion results from transcoelomic implantation of mucus-secreting cells released by perforation of the other, although the reason for selective involvement of these viscera alone is not clear. Willis (1952) considers that the probable sequence is rupture of a primary ovarian cyst which causes colloid and cellular spread in the subserous tissues and thus involves the appendix leading to mucocele. In Case 2, however, the perforated appendix preceded the large ovarian cyst, although a small cyst was noted at the first operation. Hentz (1932) recorded a case with both lesions but without peritoneal colloid, and suggested that cells passed from the ovary to the appendix by lymphatic channels.

Other complications of appendix mucocele are acute inflammation (Case 2), intussusception (Ward-McQuaid, 1949), volvulus (Jones and Carmody, 1936), and occurrence in a hernial sac (Juvara and Borcescu, 1948).

The clinical features of uncomplicated mucocele are vague and symptoms are often absent. There may be a history of recurring pain in the right iliac fossa and a mass may be palpable in the abdomen or the pelvis. Radiology may show calcification of the appendix wall or of its contents, or a soft-tissue shadow displacing the caecum or producing a filling defect may be visible with an opaque meal. Rarely does the mucocele fill with barium.

The treatment is appendectomy; in the female, especially when the mucocele is perforated, the ovaries are examined for concomitant pseudomucinous cyst. Occasionally the dense adhesions of the appendix necessitate resec-

tion of bowel for its removal. When the complication of pseudomyxoma peritonei is present the myxomatous material is removed so far as possible, and post-operative radiotherapy to the abdomen is carried out in an attempt to destroy the disseminated mucus-secreting cells. Drainage of the peritoneal cavity is avoided because of the tendency to secondary infection. A more accurate prognosis may be given in such cases if the appendix mucosa is examined for the presence or absence of papillomatous formations.

Summary

Two cases of mucocele of the appendix are described, one of them associated with pseudomyxoma peritonei and with pseudomucinous cyst of the ovary. The pathogenesis, complications, and clinical features of the condition are discussed.

I am grateful to Mr. W. M. Beattie and Mr. C. H. Walsh for permission to record Case 2; to Dr. J. Carr Brundret for the histological reports; and to Mr. J. Cosbie Ross for advice in the preparation of this paper. I have to thank the central photographic department of the University of Liverpool for the photograph.

REFERENCES

- Bailey, F. W. (1916). *Surg. Gynec. Obstet.*, 23, 219.
Bunch, G. H. (1945). *Ann. Surg.*, 121, 704.
Cheng, K. K. (1949). *J. Path. Bact.*, 61, 217.
Eden, T. W. (1912). *Lancet*, 2, 1498.
Grodinsky, M., and Rubnitz, A. S. (1941). *Surg. Gynec. Obstet.*, 73, 345.
Hentz, V. G. (1932). *Ann. Surg.*, 96, 456.
Jones, T. E., and Carmody, M. G. (1936). *Amer. J. Surg.*, 32, 511.
Juvara, I., and Borcescu, U. (1948). *British Medical Journal*, 1, 931.
Muir, J. B. G. (1931). *Lancet*, 1, 131.
Ries, E. (1924). *Surg. Gynec. Obstet.*, 39, 569.
Shemilt, P. (1949). *Brit. J. Surg.*, 37, 118.
Topping, M. H. (1937). Cited by T. R. Waugh and D. Findlay, *Amer. J. Surg.*, 1937, 37, 518.
Trotter, W. (1910). *British Medical Journal*, 1, 687.
Vorhaus, M. G. (1930). *J. Amer. med. Ass.*, 94, 165.
Wangenstein, O. H., et al. (1937). *Ann. Surg.*, 106, 910.
Ward-McQuaid, J. N. (1949). *Brit. J. Surg.*, 37, 109.
Wells, A. Q. (1936). *Ibid.*, 24, 766.
Willis, R. A. (1952). *The Spread of Tumours in the Human Body*, 2nd ed. Butterworth, London.
Wilson, R. R. (1950). *Brit. J. Surg.*, 38, 65.
Woodruff, R., and McDonald, J. R. (1940). *Surg. Gynec. Obstet.*, 71, 750.

A CASE OF ULCERATING HODGKIN'S DISEASE

BY

T. G. EVANS, F.R.C.S.Ed.

Lately Surgical Registrar, St. Luke's Hospital, Bradford

The ulcerative form of Hodgkin's disease is so rare and so difficult to diagnose that the following case is worthy of record.

Case Report

A woman aged 44 was admitted to St. Luke's Hospital, Bradford, in July, 1948, complaining of a painful swelling on the anterior chest wall, over her sternum, of twelve months' duration. The swelling had gradually become larger, recently fluctuant, and had been aspirated two weeks before admission. She also complained of painful stiffness of the neck for about three years, and aching of both arms for twelve months. The spleen was not palpable and no glands were enlarged. The E.S.R. was 18 mm. in one hour; haemoglobin, 85%; W.B.C., 16,000 per c.mm. (polymorphs 83%, lymphocytes 12%, monocytes 5%); platelets normal. The Wassermann reaction and Kahn test were negative on two occasions. X-ray examination showed erosion of the left sternoclavicular joint and adjacent sternum, with no new bone formation.

The fluctuant swelling was incised, pus and necrotic bone were curetted out, and the wound was closed. The wound healed by first intention. The pus obtained was sterile on culture and no organisms were found in the smear. A provisional diagnosis of a tuberculous lesion was made, and the patient was discharged home four weeks later, to await a bed at a sanatorium.