Mucocele of the Appendix

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MUCOCELE OF the appendix is that condition where the appendix contains a jelly-like material, pseudo-mucin (Johnston, 1954). It was first described by Rokitansky in 1842. This was followed by the description by Virchow in 1863 of a case. It is a relatively uncommon condition and Woodruff and McDonald (1940) reported that it occurred in 0.3 per cent of appendicectomies over a period of 24 years at the Mayo Clinic in the United States of America. Wesser and Edelman (1961) calculated that there have been more than 600 cases of mucocele of the appendix reported in the literature. Its incidence varies from 0.1 to 0.3 per cent of all appendicectomies (D'Annreuther, 1936; Mayo and Fauster, 1932; Weaver, 1937).

Case Reports

Case 1

A 33 year old Malay man was admitted to the District Hospital, Telok Anson, Perak, complaining of vomiting and constipation for 3 days. He had noticed a swelling in the abdomen for the last 2 years and this followed an attack of acute pain in the right iliac fossa. On examination he was looking ill and had a pulse of 90 per minute, blood pressure 140/70 mm.. Hg. His temperature was 100 degrees Farenheit. The abdomen was distended and the bowel sounds markedly hyperactive. There was a tense tender swelling 2 inches to the right of and below the umbilicus. A diagnosis of bowel obstruction due to a strangulated Spigelian hernia was made. Four hours after admission, the patient was operated upon by the author. It was found that the swelling was due to a mucocele of the appendix. The fundus of the appendix had become attached to the abdominal wall and had ruptured so that a mucoid substance lay immediately beneath the skin. The appendix was fifteen centimetres long and five centimetres wide and was very thin-walled. Inside the appendix was a colourless mucinous material. A fine probe could not be passed from the lumen of the appendix into the lumen of the caecum. The other intra-abdominal organs were normal. Appendicectomy was done and the patient made an uneventful recovery post-operatively. The histological study of the appendix wall revealed features of chronic inflammation.

Case 2

A 28 year old Malay man was admitted to the General Hospital, Kuala Lumpur on 26.11.72 for an interval appendicectomy. He had previously suffered from an appendicular abscess for which drainage was performed on 24.10.72. Appendicectomy was done on 27.11.72 and at operation a mucocele of the appendix was found. There were plentiful adhesions around and involving the appendix.

Discussion

Animal experiments, mainly in rabbits, have shown that three factors are involved in the development of a mucocele of the appendix, namely, obstruction of the lumen of the appendix, continued secretory activity of the appendiceal mucosa and sterility of the contents of the appendix (Cheng, 1949; Grodinsky and Rubnitz, 1941). The usual cause of obstruction is inflammatory stricture (Chan, 1965). Other rarer causes include neoplasms of the appendix, faecal concretion, multiple polyposis,

natural diverticula of the appendix, implants of endometriosis with stricture and neoplasms of the caecum (Hilsabeck et al, 1952; Muir, 1931; Shemilt, 1949; Topping, 1937). Wangensteen (1937) suggested the possibility of functional obstruction due to a sphincter mechanism at the appendix base since he found resistance to the flow into the caecum of fluids injected into an appendicostomy.

Clinically, mucocele of the appendix can present in four ways (Wesser and Edelman, 1961). Firstly, most cases present as acute appendicitis. Secondly, many cases are associated with chronic abdominal pain and a palpable mass. In the third group, there are no symptoms and the condition is discovered on routine physical examination or at autopsy. Finally, there is a fourth group of cases which present with bizarre or complicated courses, such as "appendiceal abscess", mucous fistula after surgery, right iliac fossa pain with intestinal obstruction or diarrhoea due to pseudomyxoma peritonei. The author's first case belongs to this fourth group. Other complications include volvulus of the appendix (Chan, 1965), diverticulosis of the appendix and ileo-caecal intussusception (Adelman and Teplich, 1955; Ward-McQuaid, 1949; Watkins, 1963).

Pseudomyxoma peritonei is the most important complication of mucocele of the appendix and is characterised by the presence of masses of colloid in the peritoneal cavity. The colloid lies either free or enclosed in membranous adhesions. It is identical in composition with the contents of the mucocele. It may be widespread in the peritoneal cavity or localised to the region of the appendix. It appears that the initial cause of pseudomyxoma peritonei is perforation of an appendix distended with a mucocele, usually through a diverticulum of the organ. However, it is not certain how the large quantities of pseudomucin which are present in pseudomyxoma peritonei arise. Three theories have been postulated, namely:—

1. The peritoneal pseudomucin results from the continued discharge of this material from a perforation of the mucocele. Against this theory is the fact that pseudomyxoma peritonei may recur after appendicectomy and finally lead to the patient's death (Johnston, 1954).
2. The presence of pseudo-mucin in the peritoneal cavity stimulates the serosal cells of the peritoneum to secrete an identical substance. Trotter (1910) believed that the cells lining the peritoneum could become columnar by metaplasia but he thought that their reaction was that of a foreign-body peritonitis and limited to the formation of enclosing adhesions.
3. The most likely theory is that mucus-secreting cells escape into the peritoneal cavity through the perforation in the appendix. Upon becoming implanted on the peritoneal surface, these cells continue to secrete pseudo-mucin. Although such cells are difficult to demonstrate histologically in the peritoneal pseudo-mucin, the theory is supported by the work of Grodinsky and Rubnitz (1941). They injected the contents of mucoceles into the peritoneal cavities of animals and produced progressive pseudomyxoma peritonei. However, if the contents of the mucocele were first passed through a Seitz filter to remove the pseudo-mucin secreting cells, pseudomyxoma peritonei did not develop.

Woodruff and McDonald (1940) studied 146 cases of mucoceles of the appendix and divided them into two groups. In the smaller group, the mucosa of the appendix is hyperplastic due to papillae covered with large columnar cells containing hyperchromatic nuclei. This is regarded as a Grade 1 adeno-carcinoma and perforation of the appendix in such cases leads to the dissemination of malignant cells in the peritoneal cavity and a diffuse progressive pseudo-myxoma peritonei. In the larger group of cases, the appendiceal mucosa consists of a single layer of columnar cells. Rupture of such a mucocele leads to the localised variety of pseudo-myxoma peritonei which does not recur after appendicectomy.

Pesudo-myxoma peritonei may also follow rupture of a pseudomucinous cyst of the ovary. In the female patient, there may be present a concomitant pseudo-mucinous cyst of the ovary and for this reason the ovaries should always be examined when operating upon a mucocele of the appendix in a woman. Eden (1912) described a patient in whom an appendix mucocele was found two years after an operation for a ruptured pseudo-mucinous cyst of the ovary. Bailey (1916) and Ries (1924) reported patients with concomitant ovarian pseudo-mucinous cyst and mucocele of the appendix. These patients also developed pseudomyxoma peritonei. These concomitant ovarian and appendiceal lesions are usually complicated by pseudomyxoma peritonei (Johnston, 1954) and it is possible that one lesion results from trans-coelomic implantation of mucus-secreting cells released by perforation of the other lesion, although it is not clear why ovary and appendix are alone selectively involved. Willis (1952) states that the probable sequence of events is rupture of a primary ovarian pseudo-mucinous cyst which causes colloid and cell spread in the subserous tissues and finally involves the appendix leading to a mucocele of the appendix. In the case reported by Hentz

(1932), both the appendix and ovary were involved but there was no pseudo-myxoma peritonei. He suggested that the pseudomucin-producing cells passed from the ovary to the appendix by lymphatic channels.

The material in a mucocele of the appendix is pseudo-mucin which is a jelly-like gluco-protein distinguishable from mucin by reduction tests and by staining with muci-carmine (Johnston, 1954). Its presence may be due to abnormal secretion by the mucosa or, since all grades of staining and reduction reactions occur, the change from mucin to pseudomucin may be a gradual extracellular one. In mucoceles present for a long time, the pseudomucin may become inspissated and calcification of this may lead to the formation of a large stone in the appendix (Bunch, 1945).

The treatment of mucocele of the appendix is appendicectomy. During the operation in a female patient, both ovaries should be inspected for a concomitant pseudomucinous cyst of the ovary and oophorectomy done if necessary. When pseudomyxoma peritonei is present, as much of the pseudomucinous material is removed as is possible. When this complication is present, Byron et al (1966) recommend that the peritoneal cavity be also irrigated with mechlorethamine (Nitrogen mustard) whilst Johnston (1954) advises post-operative radiotherapy to the abdomen. The irradiation destroys the mucus-secreting cells in the peritoneal cavity. It is not advisable to drain the peritoneal cavity as there is then a tendency to secondary infection.

Summary

Two personal cases of mucocele of the appendix seen and treated by the author are presented. The incidence and aetiology of the condition is discussed. The most important complication is pseudomyxoma peritonei. Treatment of the mucocele is by appendicectomy and the pseudomyxoma peritonei is treated by intraperitoneal mechlorethamine, manual evacuation and post-operative radiotherapy. The four types of presentation of appendiceal mucocele are also discussed.

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