PSEUDOMYXOMA PERITONEI OF APPENDICEAL ORIGIN
SURVEY OF THE LITERATURE AND REPORT OF A CASE

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The finding of gelatinous material in the peritoneal cavity was first described by Werth (1) in 1884. He named the condition "pseudomyxoma peritonei," ascribing its origin to the rupture of a pseudomucinous cyst of the ovary, with resultant implantation of the cyst contents upon peritoneal surfaces. In 1901 Fraenkel (2) first reported the finding at post-mortem examination of a pseudomucinous cyst of appendiceal origin in a man seventy-eight years of age, and postulated an inflammatory-excretory theory as accounting for its origin. Since that time, Fraenkel's views have gained wide acceptance.

Reported cases of pseudomyxoma peritonei of appendiceal origin are rare. In 1912 Kaufmann (3) collected 10 cases from the literature, omitting Goette's (4) 2 cases reported in 1903. In 1927 Ritter (5) showed that 40 cases had been recorded in the literature. In 1932, Kemkes (6) was able to find approximately 80 cases. Our survey of the literature leads us to believe that only 90 authentic cases are on record. Only one such case is to be found in the records of the Charity Hospital of Louisiana since 1909, and we are reporting it in some detail because of certain features of interest.

CASE REPORT

A colored woman, forty-seven years of age, was operated upon for umbilical hernia. The hernial sac contained a tag of omentum and gelatinous material. The abdominal cavity was entirely filled with a jelly-like material, the appendix and cecum being so involved that operative interference was deemed inadvisable. Death occurred forty days later.

At autopsy the abdomen was found to contain a large amount of translucent, jelly-like material of fairly firm consistency, with a small amount of free fluid. The omentum and the surfaces of the gut (Fig. 1) were studded with masses of this material, enmeshed and held together by strands of light colored fibrous tissue. The appendix and the lower part of the cecum formed a conglomerate mass which on section was found to consist of a wall of fibrous tissue enclosing a cavity which contained purulent fluid and a large quantity of gelatinous material. The appendix could not be located. A large quantity of the gelatinous material had pushed the diaphragm upward so that it reached as high as the third rib. Masses of gelatinous tissue were attached to the liver and spleen. The gut was partially obstructed for a distance of about two and a half feet proximal to the ileocecal valve, at which point the index finger could barely be admitted. The uterus measured $4 \times 3 \times 1.5$ cm. and a small fibroid tumor was found in the fundus. Large masses of the gelatinous material were attached to the broad ligament. The ovaries were yellowish white in color, measured $1 \times 1 \times 0.5$ cm., and on section were found to consist of fibrous tissue in which several small cysts containing a clear yellowish fluid were embedded.
Microscopic Findings (Hematoxylin-eosin Stain): The gelatinous material shows a faintly bluish staining homogeneous tissue traversed by strands of fibrous tissue. Isolated cell nests are present, some of the individual cells being cubical, others columnar; many are arranged in the form of acini, suggesting a glandular origin. Some irregularly occurring flattened cells arranged in a single layer are suggestive of endothelial origin. Foreign-body giant cells may be seen here and there (Fig. 2). Inflammatory changes are observed in the parenchyma of the liver (Fig. 3) and spleen (Fig. 4), with numerous lymphocytes and plasma cells collected immediately beneath their capsules, especially at the sites of the depressions caused by attachment and growth of myxomatous masses.

Fig. 1. Retouched photograph of a portion of the small intestine, showing myxomatous tissue attached to the mesentery.

The lining of the wall of the cavity involving the appendix and cecum consists of necrotic tissue infiltrated with polymorphonuclear leukocytes, the whole surrounded by fibrous tissue, moderately infiltrated with lymphocytes and plasma cells. Collections of myxomatous material can be seen occurring irregularly in the cavity wall.

Pathological Diagnosis: Pseudomyxoma peritonei of appendiceal origin.

Discussion

Pseudomyxoma peritonei is not a disease in itself. It may eventuate from such conditions as ovarian cystadenomata, omphalomesenteric cysts, intestinal diverticula, mucocoeles of the appendix, and retroperitoneal cystadenomata. It may even occur in the course of a recognized malignancy, as in McCrae and Coplin's (7) case, in which it was a sequel to carcinoma of the gallbladder. Some observers note that the material obtained from ovarian cysts gives the reaction for pseudo-mucin, that from appendiceal gelatinous cysts the reaction for mucin
This differentiation, if valid, could be considered of much diagnostic value in cases of multiple cyst formation in which the exact site of origin is uncertain. Dodge (8), in an analysis of 142 cases of cysts of the appendix, reported that in only 6 of these were the cyst contents examined. In 3 cases the reaction was that of mucin, in 2, that of pseudomucin, and in one that of colloid. Phemister (9) also reported a pseudomucin reaction with material obtained from an appendiceal gelatinous cyst. We may therefore conclude from existing data that the reaction of the cyst contents is of no diagnostic importance.

Fraenkel (2) and others, in discussing the etiology of pseudomyxoma peritonei of appendiceal origin, stress the significance of obstruction to the appendiceal lumen. The most important factor in the production of this obstruction, they believe, is a previous inflammatory condition. Such inflammation, if at all extensive, leads to destruction of the epithelial lining, with resultant extensive fibrosis and obliteration of the lumen. The inflammatory condition must therefore involve only a small portion of the appendix, preferably the base. Subsequent contraction of the scar tissue produces a stricture, with
obliteration of the lumen and inability of the appendix to empty itself. The glands in the appendix continue to secrete, the organ gradually becoming distended with mucus. The mucinous cyst thus formed may attain an enormous size (10), the resulting pressure causing marked distention and thinning of its walls. The accumulated mucus in such a cavity becomes changed into a jelly-like material, especially through an absorptive process. Rupture or perforation of the cyst may follow, with extrusion of its mucous contents into the peritoneal cavity or between pericecal adhesions.

Such a sequence of events requires a long period during which no symptoms referable to the appendix may be observed. Naeslund (12) ligated the base of the appendix in rabbits and produced a mucocele which was identical histologically with that found in man. That occlusion of the lumen may be produced by causes other than inflammatory lesions, is exemplified by Hudacsek’s (13) case, in which a patient with a femoral hernia, and no symptoms referable to the appendix, was found at operation to have widespread pseudomyxoma peritonei. The hernial sac contained both the cecum and the appendix. Hudacsek believed that there existed obstruction to the appendix due to the formation of periappendical adhesions with kinking and mechanical obliteration of its lumen.
Diverticula of the appendix are not uncommon. MacCarthy and McGrath (14) found 17 in 5,000 appendices examined. Gardham, Choyce, and Randall (15) suggest that obliteration of the proximal portion of the diverticulum may result in cyst formation with pseudomyxoma peritonei a possible outcome.

The myxomatous masses extruded from supposed cysts may be found in various places in the abdomen. They may be localized in the pelvis or distributed generally throughout the abdomen, forming huge aggregates resulting in intestinal obstruction. Monod and Vuillième (16) consider that following rupture of an appendiceal cyst, continued secretion of mucus through the opening into the peritoneal cavity occurs, with engrafting of the mucinous material upon the peritoneum. In their case, the appendix was distended with mucinous material and a microscopic opening was present near the tip, through which, as they believed, mucus was extruded. These writers add that microscopic examination of the myxomatous material showed no cells capable of secretion. Lejars (cited by Monod and Vuillième) doubts that such large quantities of mucinous material as have been found can be extruded through small openings. Moreover, he points out that many of the glands of the appendiceal mucosa under conditions of obstruction become atrophied and are devoid of secretory power. The remaining glands, while showing hypertrophy, are capable of pro-
ducing large quantities of mucus. Microscopic examination of the mucinous masses found in his case disclosed collections of epithelial cells which were of the cylindric and goblet types. Lejars' conclusion is that pseudomyxoma peritonei is caused by seeding of the peritoneum with mucosal cells, with subsequent proliferation of these cells and formation of mucus, the growth being a true adenocystoma. With this opinion, we are in sympathy.

Trotter (17) agrees with this view. The cubical and columnar cells found in the myxoma he believes are derived from three sources, the epithelial lining of the appendix, the endothelial cells of the peritoneum, and phagocytes which have wandered in to engulf the foreign material. The peritoneal changes represent an attempt on the part of the peritoneum to wall off or absorb the material.

Hansmann and Budd (18), in a recent paper, reviewed the recorded cases of retroperitoneal tumors and added 17 cases of their own. Included among their cases were several in which the histology was similar to that of pseudomucinous cystadenoma.

COMMENT

In the case here reported, the fact that the myxomatous material was present in the cavity and in the wall of the appendiceal abscess, and nowhere else, proves, we believe, that the growth was of appendiceal origin.

The course of events might be explained as follows: Myxomatous changes had taken place in the appendix with resulting dissemination throughout the abdominal cavity. Additional infection occurred, with destruction of the organ and formation of a localized abscess. The cells found in the myxoma originated from the appendix, as evidenced by their predominating alveolar arrangement. The areas of invagination or depressions in the surfaces of the liver and spleen caused by attached myxomatous masses lead to the conclusion that myxomatous masses of this type have growth and reproductive power, and must therefore be classed as true adenocystomata.

In our case, the condition was evidently of long standing, for symptoms of chronic obstruction were present at the time of examination. This supports the view that the growth is of a comparatively benign nature, associated mortality usually being due to obstruction of the intestinal tract.

SUMMARY

A case of pseudomyxoma peritonei of appendiceal origin is presented, together with a brief survey of the literature.

The opinion is expressed that most such cases are adenocystomata.
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References