MEDICAL MEMORANDA

Pseudomyxoma Peritonei: An Unusual Case

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Pseudomyxoma peritonei is a rare condition which occurs predominantly in middle-aged women. Most cases are associated with ruptured ovarian pseudomucinous cysts and some with ruptured mucocoele of the appendix. Many individual cases have been reported, and in some both ovaries and appendix have been involved, though it is uncertain whether both have been primarily diseased or if one is secondary to the other.

Case Report

A 52-year-old woman was well until January 1969, when she developed a painless swelling in the right groin. This was diagnosed as a reducible inguinal hernia and she was admitted to hospital in May for herniorrhaphy. There were no other abnormalities on physical examination, the haemoglobin and W.B.C. were normal, and the E.S.R. was 25 mm in one hour (Westergren). At operation the hernial sac was found to contain a jelly-like substance coming from the peritoneal cavity. The sac was excised and the hernia repaired. Histologically the wall of the sac contained some atypical epithelial cells, suggesting the possibility of mucoid carcinoma, and the contents appeared to be pseudomyxomatous material.

Laparotomy in June showed pseudomyxoma peritonei confined to the pelvis and lower abdomen. The appendix contained a ruptured mucocoele but the ovaries were macroscopically normal. Appendectomy was performed and the mucin was removed as completely as possible. Histologically the appendix contained a ruptured mucocoele and there was pseudomyxomatous material on its surface.

She remained well until December 1971, when she developed painless abdominal distension, anorexia, heartburn, and mild dyspnoea of effort. The abdomen was soft, distended, and dull to percussion, though shifting dullness was not demonstrable. There were no palpable masses and bowel sounds and rectal examination were normal. The blood count was normal but the E.S.R. had risen to 70 mm in one hour. Barium-meal examination showed that the bowel was normal but displaced by something filling most of the peritoneal cavity.

At laparotomy in January 1972 about 6.5 litres of tenacious lime-coloured jelly were removed from the peritoneal cavity. There was a large multiloculated right ovarian cyst partly ruptured and containing a similar jelly. The left ovary, Fallopian tubes, and uterus appeared normal, though there were multiple jelly-containing cysts adherent to the greater omentum and surface of the liver and spleen. Most of the jelly was removed and bilateral salpingo-oophorectomy was performed together with excision of the greater omentum.

Histologically the right ovary contained a multiloculated pseudomucinous cystadenoma and the omentum was covered with yellowish mucoid nodules. Both showed tall columnar secretory

epithelium filled with mucinous material lining the cystic areas. The left ovary and Fallopian tubes were normal.

Comment

There have been several case reports of an association between mucocoele of the appendix, pseudomyxoma peritonei, and cysts of the ovary. However, in most of these cases all three entities were present at the initial laparotomy, and because of the extent of the disease and the fragility of the lesions it is difficult to be certain of the true pathology. It is possible to explain most of the reported cases in the following ways. Ruptured mucocoele of the appendix may lead to pseudomyxoma peritonei, with secondary deposits on the ovary (for example, the cases reported by Byron *et al.* 1966); ruptured pseudomucinous cysts of the ovary may lead to pseudomyxoma peritonei, with secondary cystic deposits adherent to the caecum and appendix. Myer (1907) reported a case with almost total destruction of the caecum by cystic infiltration, and Smith (1908) reported a case with involvement of the appendix tip. Both patients had primary disease of the ovary; in ruptured pseudomucinous cysts of the ovary with pseudomyxoma peritonei the pressure of the mucin and cystic deposits may occlude the lumen of the appendix and, in predisposed individuals, lead to true mucocoeles. This was suggested by Willis (1952).

In the present case the appendix and ovary were affected at different times, two and a half years apart, though both were associated with pseudomyxoma peritonei.

The presence of the two primary conditions in this patient was probably coincidental. However, the fact that both are associated with pseudomyxoma peritonei indicates a little more than coincidence, since not all cases of ruptured mucocoele of the appendix or ruptured pseudomucinous cysts of the ovary lead to pseudomyxoma peritonei.

It may well be that in predisposed individuals liberation of mucin, whether from ruptured ovarian cysts or ruptured mucocoele of the appendix or any other site, will initiate a "foreign body" response by the peritoneum (Carleton, 1955). Thus removal of all the mucin and potential sites should improve the prognosis.

It is interesting to speculate whether pseudomyxoma peritonei would have recurred in this patient if the ovaries had been removed at the first laparotomy. Little, Halliday, and Glenn (1969) suggested that the appendix and both ovaries, whether diseased or not, should be removed in a patient with pseudomyxoma peritonei. This case supports this view.

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