Mucinous cystadenoma of the appendix with raised serum carcinoembryonic antigen concentration: clinical and pathological features

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Abstract

A case of mucinous cystadenoma mimicking ovarian cancer is reported. Serum carcinoembryonic antigen (CEA) concentration was raised, and computed tomography of the abdomen and pelvis demonstrated a long oval shaped cystic mass measuring 9 cm in length on the right anterior side of the uterus. Because of possible right ovarian cancer, laparotomy was performed and the mass was found to be a mucinous cystadenoma of the appendix. This case indicates that mucinous cystadenoma of the appendix may show an unusual presentation including its location as well as the high serum CEA, mimicking ovarian cancer. Therefore, gynaecologists as well as gastroenterologists should consider its possibility as a differential diagnosis of the right adnexal mass in a patient without previous appendectomy.

Keywords: mucinous cystadenoma; mucocoele; appendix; carcinoembryonic antigen; ovarian cancer

An enlarged appendix with luminal dilatation by mucus has generally been called mucocoele. Higa et al., in 1973, investigated cases of mucocoele of the appendix and they classified their lesions into three groups: mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma. These investigators also recommended avoiding the term mucocoele. However, mucocoele is still used for gross description, rather than histological diagnosis. In addition, the term retention mucocoele, also called simple mucocoele, is still applied to a pathological description of the mucinous dilatation of the appendiceal lumen resulting from any cause other than epithelial proliferation. On the other hand, when an appendiceal adenoma secretes large amounts of mucus resulting in a clinically palpable cystic lesion, the term cystadenoma of the appendix preferably is used.

We report a case of mucinous cystadenoma of the appendix accompanied by raised serum carcinoembryonic antigen (CEA) concentrations. The case was unique because of the location of the tumour, which led to an initial diagnosis of ovarian cancer.

Case report

A 75 year old Japanese woman (gravida 6, para 6) underwent a medical check up that identified raised serum CEA. Two months later, she visited a women’s clinic and was referred to our hospital with a putative diagnosis of right ovarian cancer. Laboratory data including CA125 and CA19-9 were all within normal limits, except for a raised value for CEA (17.7 ng/ml; normal range 0–2.5). While there was no mass palpable on the abdomen, physical examination of the pelvis revealed a sausage shaped mass in the right adnexal region measuring 8 cm in length. There was no lymphadenopathy and cytological examination of the cervix and vagina showed no malignancy. Computed tomography (CT) and ultrasonography of the abdomen and pelvis demonstrated an elliptical cystic mass measuring 9 cm in length on the right anterior side of the uterus (fig 1). Barium enema examination showed no abnormalities within the colorectum.

Laparotomy was performed because of possible right ovarian cancer. During the operation, no ascites was noted. Operative findings
confirmed that the mass was located in the right anterior side of the uterus, and that it originated from the appendix, rather than from the ovary. Because serosal invasion and regional lymph node enlargement were not evident, appendectomy was performed.

Immediately after the operation serum CEA concentrations became normal. The patient has been well for 10 years since the surgery without any symptoms or recurrence.

The resected appendix was diffusely enlarged, measuring 8 cm in length and 3 cm in maximal diameter. On sectioning, the lesion was cystic and contained large amounts of mucus. Microscopically, the cyst wall was lined by columnar mucinous epithelium with tall, crowded, and basally located nuclei (fig 2), which was immunohistochemically positive for CEA. There was no evidence of malignant change in 5 mm thick serial sections from the resected specimens.

Discussion

Carr et al. recently classified non-carcinoid epithelial tumours of the appendix into the following five groups: simple mucocele, hyperplastic polyp, adenoma, mucinous tumour of undetermined malignant potential, and adenocarcinoma. In that classification, the application of the term mucocele is limited to inflammatory or obstructive lesions showing neither mucosal hyperplasia nor neoplasia. The term mucocele is, however, still confusing because it is frequently applied by clinicians to mucinous tumours of the appendix, and pathologists use the terms retention mucocele or simple mucocele. In this report we specified the lesion as mucinous cystadenoma, according to the classification by Carr et al.

The tumour in the present case was characterised by raised serum CEA concentrations and an unusual location. These features were initially suggestive of ovarian cancer. While patients with mucinous cystadenocarcinoma, a rare malignant counterpart of cystadenoma, have been known to manifest high serum CEA values, there has been no reported case of mucinous cystadenoma of the appendix with raised serum CEA. This rare association seems to be partly explained as serum CEA has not been measured routinely preoperatively in cases of mucinous cystadenoma of the appendix.

A correct preoperative diagnosis of cystic mass of the appendix can readily be made with CT and ultrasound. However, these procedures are not perfect, because there are cases of variable location and appearance as demonstrated in our case. Such an unusual location of mucocele of the appendix, which was initially diagnosed as twisted ovarian cyst, has been described.

In conclusion, we present an uncommon case of mucinous cystadenoma of the appendix that mimicked ovarian cancer. Our case suggests that appendiceal tumour should be considered as a differential diagnosis for the right adnexal mass of subjects without previous appendectomy. Because serum CEA possibly increases in subjects with the tumour, pathologists should examine the whole resected specimens for this type of tumour to rule out completely the coexisting carcinoma.