

An unusual cause of ascites

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CASE REPORT

A 72-year-old woman with an extensive medical history, including cervical carcinoma, rheumatoid arthritis, hypertension and COPD, was seen at the internal medicine outpatient clinic because of ascites. The day of the scheduled ascites puncture, she presented herself to the Emergency Room with nausea, vomiting and diarrhoea. Ascitic fluid was obtained in which malignant cells were found. Histologically these were consistent with ovarian carcinoma. However, a tumour of the gastrointestinal system could not be excluded. We performed a computed tomography (CT) scan of the abdomen (*figure 1*) and thorax, which showed no signs of a gynaecological tumour or primary tumour of the gastrointestinal system. A vast amount of ascites and an omental cake were reported by the radiologist. We consulted a gynaecologist, who could not find a gynaecological tumour using transvaginal ultrasound.

We then tried to obtain histological material from the omental cake. Unfortunately the ultrasound-guided

Figure 1. Abdominal CT-slides showing an abdominal cake and ascites

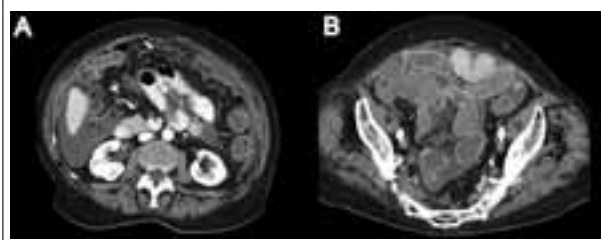
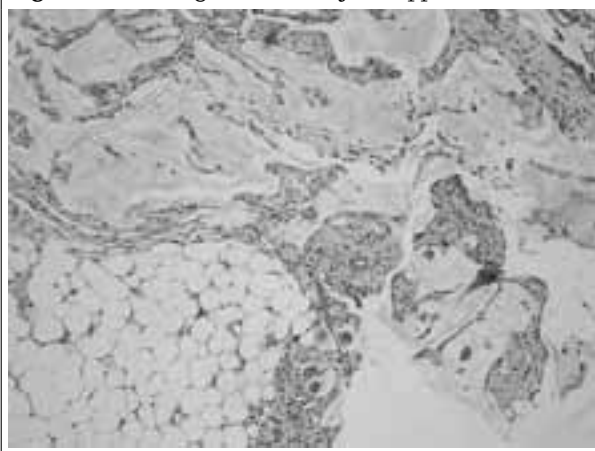


Figure 2. Histologic material of the appendiceal mass



puncture failed because the lesion could not be recognised. In consultation with the gynaecologist it was decided to perform a laparoscopic exploration of the abdominal cavity. During the laparoscopy, no tumours of uterus or adnexes were observed. However, multiple miliary omental lesions and an appendiceal mass were reported. Biopsies were taken (*figure 2* shows the histology of the appendiceal mass).

WHAT IS YOUR DIAGNOSIS?

See page 197 for the answer to this photo quiz.

ANSWER TO PHOTO QUIZ (PAGE 193)

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DIAGNOSIS

Histology shows a mucoid adenocarcinoma with signet cell differentiation. In combination with the clinical findings we made the diagnosis of metastasised mucoid adenocarcinoma of the appendix. Interestingly, the CT scan showed an omental cake (*figure 1A*), but nothing of interest in the appendiceal area (*figure 1B*). This could be due to the fact that the carcinoma has a signet ring differentiation.

Multiple histological types of appendiceal carcinomas are known, of which signet ring differentiation is the smallest group.^{1,2} In signet cells the cell is filled with mucus, leading to an outward position of the nucleus. This type of cancer is exceedingly rare. Dutch numbers are lacking, but extrapolating from US data,³ a case of appendiceal adenocarcinoma with signet ring differentiation is only seen once every five years in the Netherlands.

The condition is usually an unexpected finding during surgery for another indication. However, 80% of cases present with abdominal pain or acute appendicitis.² Other symptoms are a bloated feeling or an abdominal mass. If a physician suspects an appendiceal carcinoma based on the patient's symptoms, regular imaging techniques can be used.

The treatment of choice is a right hemicolectomy.⁴ If the condition of the patient does not allow an operation, or

when (inoperable) metastases are present, the physician should consider chemotherapy. The prognosis of primary carcinoma of the appendix is dependent on the stage of the disease.³ Carcinoma with signet ring differentiation tends to have a poorer prognosis because of very early dissemination to the peritoneum,³ which is probably the reason why it did not show up on the CT scan. Physicians should always keep this type of tumour in mind when malignant cells are found in ascitic fluid without obvious lesions on imaging techniques.

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