

Pseudomyxoma peritonei

A report on two cases at Ibn Seena Teaching Hospital /Mosul

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ABSTRACT

We report here two unusual cases of a rare tumour "pseudomyxoma peritonei".

1. Case one: A 55-year-old female patient, with history of carcinoma of left breast presented with ascites which was proved to be due to pseudomyxoma peritonei rather than to metastases from breast cancer.

2. Case two: An 18-year-old male patient presented with huge abdominal distention in whom a diagnosis of pseudomyxoma peritonei was confirmed; an unusual age and sex for this rare tumour.

الخلاصة

تم تسجيل حالتين من مرض نادر الحدوث (ورم مخاطي كاذب في التجويف البريتوني) في مستشفى ابن سينا التعليمي في الموصل. الحالة الأولى لامرأة عمرها 55 سنة مصابة بورم سرطاني في الثدي الأيسر، وتعاني من استسقاء البطن ثبت أنه ناتج عن ورم مخاطي كاذب في التجويف البريتوني وليست حصيلة لورم ثانوي في البريتون. الحالة الثانية لمريض ذكر عمره 18 سنة، كان يعاني من انتفاخ البطن اثبتت إصابته بالمرض المذكور

Pseudomyxoma peritonei is a rare condition characterized by mucinous ascites ("jelly belly") and multifocal mucinous tumours on the peritoneal surfaces and omenta⁽¹⁾. It is twice more common in females than in males and is found in 2/10,000 laparotomies⁽²⁾.

The term *pseudomyxoma peritonei* was first used by Werth⁽³⁾ in 1884 to describe massive intraperitoneal accumulation of gelatinous pseudomucin ascribed to the perforation of ovarian pseudomucinous cystomas. Based on recent studies reevaluating this condition, tumours previously designated pseudomyxoma peritonei, can now be viewed as two pathologically and prognostically distinct disease processes⁽⁴⁻⁶⁾.

Pseudomyxoma peritonei is a frequently misdiagnosed condition. It has diverse and slightly confusing presentations, including abdominal distension, ovarian tumors, or appendicitis-like syndromes. Less frequent

symptoms include nausea, vomiting, fatigue, and urinary symptoms⁽⁷⁾.

Here we present two different and unexpected cases of such a disease, with a literature review of some important aspects.

Case one

A 55-year-old woman with a recent history of abdominal distension and abnormal chest x-ray shadow (ill defined opacity of the left lower zone) was referred to Ibn Seena Teaching hospital on 18th of January 2005, for further evaluation. The patient is a known case of left sided breast cancer for which a radical mastectomy was done followed by chemotherapy and radiotherapy 3 years ago. Four months prior to her presentation, she started to have mild cough and abdominal distention. She is a widow and has never been pregnant during her twenty five years of marriage.

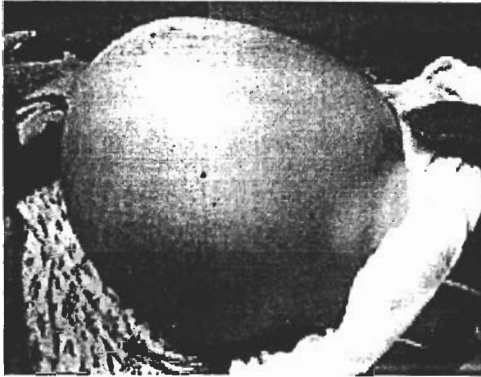


Figure (1): Photograph of the abdomen of patient(1).

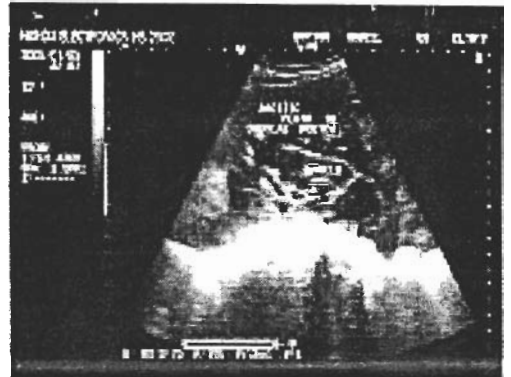


Figure (2): Ultrasound of the abdomen.

On chest examination, there was an old scar of left sided mastectomy; otherwise examination of the chest was unremarkable. Abdominal examination showed distention (figure 1) with ascites and a suspicious mass in the right iliac fossa was felt with difficulty by dipping method only. Her haemoglobin: 100g/L, PCV: 0.40, WBC: $5.8 \times 10^9/L$, ESR: 67mm/ first hr. Blood urea: 5.8mmol/l, s. creatinine: 84 μ mol/l, s. sodium: 140mmol/l, s. potassium: 4.4mmol/l. Urine examination: normal. Ultrasound of the abdomen: Marked multiloculated ascites difficult to spread, mostly in the lower abdomen (figure 2).

MRI: Report confirmed the presence of ascites with no mention about the mass lesion or the distribution of the ascites.

Ascitic fluid aspiration was performed by using a wide bore needle. The fluid was very difficult to aspirate, gelatinous in consistency and yellow in colour, mucoid-sticky; protein:

40g/L, cell count: 80/cu mm (mostly neutrophils), Bacteria-free and negative for AFB. Cytology: negative for malignant cells. She was then referred to the surgeon with a provisional diagnosis of pseudomyxoma peritonei.

Surgery: Exploratory laparotomy, using right paramedian incision was performed. The surgeon reported multilocular cystic lesions with thick huge mucoid ascites mostly due to ovarian or appendicular tumor. The ascitic fluid was partially removed with surgical debulking and sent for histopathology, which stated: fibrous tissues containing multiple cystic areas lined by mature mucinous epithelium; containing mucoid material (figure 3).

Conclusion: Metastatic adenocarcinoma from the ovaries or appendix (pseudomyxoma peritonei). The patient is still alive till the moment of writing (May 2006).

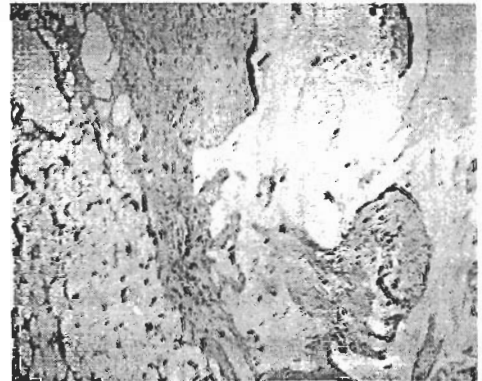
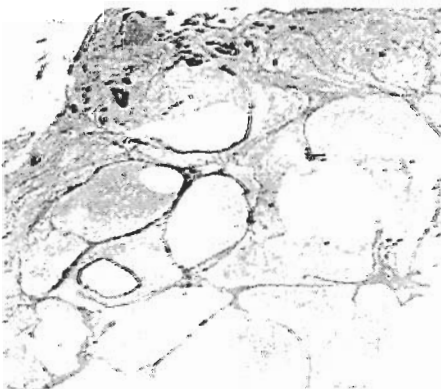


Figure (3): Case (1): Microphotographs showing multiple cysts lined by mucinous epithelium.



Figures (4 and 5): Showing a hugely distended abdomen.

Case two

An 18-year-old male patient was referred to outpatient clinic at Ibn Seena Teaching Hospital, Mosul, on Wednesday 9th of March 2005. He described his condition as increasing abdominal distension that started gradually three months ago, but became remarkable during the last 20-25 days to a degree that brought attention of his parents and his teachers at school. The patient also noted increasing tiredness, fatigue and decline in his usual activities during the last month, but his appetite remained good.

On examination: The patient looked ill, pale, and slightly distressed. He was afebrile, not jaundiced, and not cyanosed. BP was 100/50 mmHg and pulse rate 95/minute, regular. His JVP was not elevated. There were no stigmata of chronic liver disease. Despite generalized wasting, the abdomen was hugely distended with dullness all over. Liver, spleen as well as kidneys couldn't be palpated (figures 4 and 5).

Examination of other systems including the heart and chest was unremarkable.

He was admitted to medical ward for investigation with a provisional diagnosis of huge ascites.

Investigations: Hb:105g/L, WBC count 5.4x 10⁹/L. N: 52%, L: 31, M: 2%, B and E: 0%. RBCs morphology: normocytic, normochromic, ESR:95mm/1 h, s. urea: 3.8 mmol/L, creatinine: 85 µmol/L, blood sugar: 4.8mmol/L. Serum electrolytes (sodium, potassium, calcium, and phosphate) were within normal range. s. albumin: 35g/L; urine analysis was negative for albumin, and casts. Chest X-ray showed normal lung fields and no cardiomegaly; ECG was normal. Ultrasonography of the abdomen reported evidence of encysted fluid mainly at pelvic area, no organomegaly was seen, and absence of free fluid in abdomen.

A diagnostic fluid aspiration using a wide bore needle, guided by ultrasonography, failed to obtain a suitable specimen for testing.

GIT barium series showed thickened, dilated bowel loops mainly at terminal ileum and caecum with encysted fluid at pelvic area, suggestive of intestinal tuberculosis; still a common disease in our locality.

A CT scan examination of abdomen with contrast again was not conclusive. Upper and lower G. I. endoscopy examinations were normal.

Laparoscopy of the abdomen was decided as there was no response to a six weeks course of anti-tuberculous treatment. It revealed the presence of large amount of thick, gelatinous, yellowish to brown material in the peritoneal cavity, covering the peritoneum and liver surfaces. The bowel loops were adherent to each other. An ill-defined mass at terminal ileum and appendicular areas was found which was confirmed by exploratory laparotomy later on. Multiple peritoneal, liver and omental biopsies as well as samples of the material were taken for histopathological and cytological examination. The histopathology and cytology reports concluded that: the result of examination of submitted tissues and material is highly suggestive of a rare tumour called "pseudomyxoma peritonei" most probably of appendicular origin.

The patient was discharged home for few days rest, and then referred to surgeon. De-bulking and removal of the excessive material and tumour mass was performed, which actually was very difficult, owing to intense adhesions and fibrosis.

The patient did well with some decrease in the bulk of his abdomen for about seven months after surgery, and then he had recurrent abdominal distention and pain. He

was considered unfit for further surgery and died shortly after that.

Discussion

Pseudomyxoma peritonei is an unusual condition in which there is copious mucinous and recurrent ascites with omental and peritoneal implants⁽⁸⁾. The disease is commonly seen in female patients aged 50 - 70 years. Our first patient is in the same age range (55 year old) female; but the second one was a teenager male. Usually there is a high female/male ratio^(5,9). There has been considerable debate on the true origin of these tumours. It could be a primary appendiceal malignancy with metastases to the ovary, an ovarian primary malignancy with metastases to the appendix, or there could be two independent primary disease processes^(7,10,11). This has considerable significance in the management of the disease. The argument for an appendiceal origin for these tumours was further strengthened by molecular genetic and immunohistochemical studies. Recent molecular evidence supports that mucinous tumours involving the appendix and ovaries in women with PMP are derived from a single site, most likely the appendix⁽¹⁾. Ronnett *et al* also pointed out that women with PMP actually have synchronous appendicular and ovarian tumours; the ovarian tumours were secondary to appendicular tumors⁽¹²⁾.

In our patients the histopathological reports indicate the possibility of ovarian or appendicular origin in case one as both the right ovary and appendix were involved, while appendicular origin was confirmed in case two. In PMP the ascitic fluid is very difficult to aspirate and the fluid is thick gelatinous in consistency, yellow in colour with an operative finding of multilocular cystic lesion with thick huge mucoid ascites⁽¹¹⁾; all these findings were present in our patients and support the diagnosis.

Immunohistochemical studies show that the pattern of cytokeratin expression in mucinous tumors associated with PMP is similar to that of appendiceal mucinous cystadenocarcinomas in the absence of PMP (CK20+ and usually CK7-) and different from that of primary ovarian mucinous tumours of low malignant potential (CK20+ and CK7+)^(12,13). Such tests may support the idea of appendicular or ovarian origin of PMP, but unfortunately these tests are unavailable in our country. Though cytomorphologic features (cytology) of peritoneal washings

material which can categorize cases of PMP as either disseminated peritoneal adenomucinosis or peritoneal mucinous carcinomatosis was performed in our cases but not clearly fixed the above categories. Furthermore, analysis of follow-up data from some studies revealed that cases diagnosed as disseminated peritoneal adenomucinosis had a better prognosis than those diagnosed as peritoneal mucinous carcinomatosis⁽¹⁴⁾.

The first patient is still alive at time of reporting (May 2006), while the second one died seven months after operation. The disease was not suspected initially in both cases. In the first a secondary metastasis to peritoneal cavity originating from primary breast carcinoma was suspected and in the second such a diagnosis was never suspected at admission for reasons already mentioned.

Extensive surgical cytoreduction combined with intraoperative heated intraperitoneal chemotherapy (i.e. administration of mitomycin C) is feasible in patients with PMP and improved long-term survival might be achieved⁽¹⁵⁾. Extensive surgical de-bulking, which is the currently accepted mainstay of treatment for patients with PMP of appendiceal origin⁽¹⁵⁻¹⁷⁾ was performed, but neither patient received any intra-peritoneal cytotoxic drugs. Some studies have demonstrated that mortality and morbidity in adjuvant chemotherapy-treated patients are not statistically different from those of non-adjuvant treated patients⁽¹⁶⁾. There is no currently recognized role for adjuvant chemotherapy in PMP^(16,17). The uses of mucolytic agents to loosen mucinous deposits and phototherapy-mediated dissection at laparotomy have not been shown to provide clinical benefit⁽¹⁸⁾.

Conclusions: Although the possibility of secondary metastases from the breast cancer was suspected in the first case and the disease was not expected in the second case because the age and sex of the patient did not fit with this disease, yet the diagnosis of PMP was made in our patients due to the presence of:

- a-Multiloculated ascites difficult to spread, mostly in the lower abdomen.
- b-The fluid was very difficult to aspirate and it was gelatinous, yellow mucoid and sticky in consistency.
- c-Histopathologically: Fibrous tissues with multiple cystic areas lined by mature mucinous epithelium; containing mucoid material.

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