

# Appendiceal pseudomyxoma peritonei in a pregnant woman

Abu Z. Manan, MRCS, FCPS, Mazhar H. Raja, MBBS, FRCSEd, Robert Namukangula, MD, FRCOG, Ashraf A. Benayan, MD, FRCSC.

## ABSTRACT

تظهر حالة واحدة من 1000 حالة سرطانات خلال فترة الحمل. وتحدث الأورام الطلائية في حوالي 1% من جميع أورام القولون والمستقيم وحدوثها أثناء الحمل نادر للغاية. نحن هنا بصدد نشر تقرير عن حالة غير عادية لورم مخاطي صفاقي كاذب (PMP) في امرأة حامل تبلغ من العمر 41 عام. أجريت لها عملية قيصرية وأظهر البطن ورم الزائدة المخاطي مع حجم ضخم لورم مخاطي صفاقي كاذب (PMP). في هذه الحالة يوضح طبيعة ظهور المرض وحدوثه في امرأة حامل دون أي مظاهر سريرية خلال فترة ما قبل الولادة. كما تم توثيق مراجعة الأدب هذه. يكتشف الورم المخاطي الصفاقي الكاذب PMP عادة خلال عملية جراحية لعلاج أمراض أخرى، ويطلب رفع مؤشرات الشك للتشخيص. تعطي طرق التشخيص مثل الموجات فوق الصوتية، والأشعة المقطعية CT، والرنين المغناطيسي MRI بعض الأدلة للورم المخاطي الصفاقي الكاذب PMP، ولكن التشخيص النهائي يتم عن طريق المنظار، أو فتح البطن الاستكشافي فقط.

One in 1000 cancers occur during pregnancy. Epithelial tumors of the appendix occur in approximately 1% of all colorectal tumors, and the occurrence of this neoplasm during pregnancy is extremely rare. We report an unusual case of pseudomyxoma peritonei (PMP) in a 41-year-old full-term pregnant woman. Cesarean section was carried out, and the abdomen was found to contain an appendiceal mucinous tumor with large volume PMP. This case illustrates the incidental nature of the disease and its occurrence in a pregnant woman with no clinical features during the antenatal period. A review of recent literature is also documented. Pseudomyxoma peritonei is commonly discovered during surgery for other conditions, and a high index of suspicion is required to make a diagnosis. Diagnostic modalities such as ultrasound, CT, and MRI can provide some evidence for PMP, but the

definitive diagnosis is only made laparoscopically or by exploratory laparotomy.

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*From the Departments of Surgery (Manan, Raja), Obstetrics/ Gynecology (Namukangula), and ENT (Benayan), Armed Forces Hospital, King Abdul-Aziz Naval Base, Al-Jubail, Kingdom of Saudi Arabia.*

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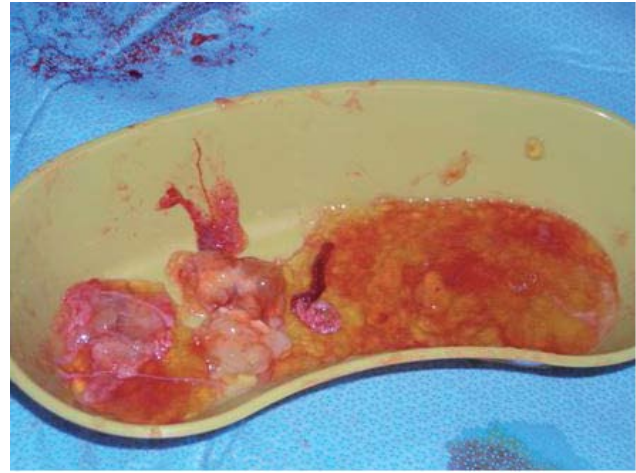
*Address correspondence and reprint request to: Dr. Abu Z. Manan, Department of Surgery, Armed Forces Hospital, King Abdul-Aziz Naval Base, PO Box 413, Al-Jubail 31951, Kingdom of Saudi Arabia. Tel. +966 551768630. E-mail: abuzarkh@yahoo.com*

Epithelial tumors of the appendix account for approximately 1% of all colorectal cancers.<sup>1</sup> This tumor has a wide range of presentations ranging from incidental findings at routine appendectomy, to ruptured high-grade appendiceal malignancy with huge quantities of pseudomyxoma peritonei (PMP).<sup>2</sup> The diagnosis of appendiceal neoplasm in pregnancy is a difficult one, and only a few cases have been reported, which were incidental findings during surgery for other conditions. The definitive diagnosis can only be made by exploratory laparotomy, although diagnostic modalities like ultrasound, CT, and MRI do support the diagnosis of PMP. We present this case of PMP due to the incidental nature of the disease, and its occurrence in a full-term pregnant woman with no signs and symptoms related to the disease in the antenatal period, diagnosed only when the abdomen was opened for cesarean section. A review of the recent literature is also documented.

**Case Report.** A 41-year-old healthy full-term pregnant woman, gravida 7, para 6, with known history of gestational diabetes mellitus was admitted to the Obstetrics & Gynecology Department for elective lower segment cesarean section given the large size of previous babies. Her antenatal care was unremarkable apart from the gestational diabetes mellitus, which was



**Figure 1** - Appendix with mass removed.



**Figure 2** - Gelatinous material removed from the peritoneal cavity.

controlled on diet. All the previous babies were delivered by spontaneous vaginal delivery with a history of severe shoulder dystocia. The first baby was a stillbirth, and the second was a forceps delivery. The antenatal course was smooth with regular outpatient visits and routine regular ultrasound examinations. In the antenatal period the baby was healthy with normal fetal movements and no signs of distress with expected fetal weight of 3.9 kg. She did not complain of any abdominal pain or loss of weight or appetite, jaundice, hematemesis, or melena. The physical examination, which included abdominal and vaginal examinations, was normal with no evidence of any tenderness, organomegaly, or ascites. All the routine blood investigations, including full blood count and chemistry were normal, except for a few readings of elevated blood sugar levels. She was admitted, and a trial of induction of labor was given, but over the course of a few days she failed to progress. A decision was then made for lower segment cesarean section. She was taken to the operating theater, and the abdomen was opened through Pfannenstiel incision. The whole of the peritoneal cavity was found to be full of gelatinous material. The ovaries were normal size, but stacked to the uterine fundus. The appendix tip was swollen with a cavitated mass. A live baby boy was delivered in good condition. A per-operative surgical consultation was sought, and the surgical team made a per-operative diagnosis of PMP. Appendicectomy was carried out with removal of the mass (Figure 1). The gelatinous material (Figure 2) was scooped out of the peritoneal cavity and all the organs were made free from it. The peritoneal cavity was thoroughly irrigated with copious amounts of saline, and the abdominal cavity was closed in layers. The appendix with mass, and the gelatinous material was sent for histopathology. Her postoperative course was uneventful. Her blood sample was sent for

tumor markers, CA-125 and alfa-fetoproteins. The CA-125 was elevated at 57.3 U/ml (normal = 0-35 U/ml) and alfa-fetoprotein was within normal range. The biopsy result showed well differentiated mucinous cystadenocarcinoma of the appendix. The gelatinous material was PMP. She was subsequently referred to oncology for further management. She was advised regarding the possibility of repeat laparotomy, intra and postoperative heated intraperitoneal chemotherapy in the future, which she said she would consider, but did not follow up. It was later found that she went abroad for further management of the disease.

**Discussion.** Our case of PMP, occurring during pregnancy highlights interesting challenges in the diagnosis and management of cancer in pregnancy. Like other cases, our case shows paucity of evidence in the diagnosis and optimal management.<sup>3</sup> Previously, the etiology of pseudomyxoma was considered to be an appendiceal mucinous adenocarcinoma and not merely obstruction of the appendix lumen with resultant mucocele. Recently, it was agreed that PMP is a condition in which peritoneal mucus is found in the presence of an intraperitoneal adenocarcinoma.<sup>4</sup> In most cases, PMP arises from a mucin producing adenocarcinoma of the appendix and ovary. Some have reported association of this disease with pancreatic, stomach, breast, and bile duct cancers. The PMP occurs in 2 in 10,000 laparotomies. Malignant tumors of the appendix account for 0.2-0.5% of all gastrointestinal tumors. The mean age of patients with PMP of appendiceal origin is 46, as compared to 59 years in those with ovarian primary cancer.<sup>5</sup> The signs and symptoms include abdominal pain, palpable mass, weight loss, and abdominal distention as a late sign. Some degree of anemia may be present. Laboratory tests

are of little value diagnostically, as in our case, she had no clinical features of the disease in the antenatal period and all the routine investigations were normal. The carcinoembryonic antigen (CEA) level is sometimes elevated and may be useful in follow-up evaluations of the patient whose CEA level decreases after resection and tumor debulking. Radiographically, CT and MRI are helpful and yield a diagnosis of PMP. A barium study of the gastrointestinal tract and ultrasound is less helpful but can be used as screening modalities. Most cases are diagnosed surgically as in our case, although this situation is changing. Histologic examination shows most malignancies to be low-grade carcinoma as in our case. This is consistent with the indolent behavior of the disease, lack of distant metastasis, and frequent long-term survival associated with PMP.<sup>6</sup> The extensibility of the surgical approach depends upon which segments, and how much, of the small or large intestine is involved.<sup>2</sup> Operative treatment aims for total removal of the tumor, however, this was achieved in only 20% of the cases in the study by Gough et al.<sup>7</sup> Only 7 cases were reported in a review of the literature of appendiceal mucinous tumors occurring during pregnancy in the absence of PMP syndrome, 6 of them were confined to the appendix, and one non-mucinous appendiceal adenocarcinoma with peritoneal carcinomatosis.<sup>4-10</sup> In one case, diagnosis was made in the third trimester of pregnancy and early delivery was carried out before the definitive management. In one patient, the diagnosis was only made at the time of cesarean section at full term. Four of these patients presented with an acute abdomen and a definitive diagnosis only made at laparotomy. One of these patients elected for therapeutic abortion prior to repeat surgical treatment, one patient had right hemicolectomy at 26 weeks of gestation and delivered at term, the remaining 2 were treated with appendectomy. In the final patient, diagnosis was made in the first trimester when she had spontaneous abortion and underwent surgery 3 months later when imaging studies showed progression of the disease. On the basis of our case and the previous reports, it seems reasonable to carry out diagnostic surgical assessment of a mucinous appendiceal neoplasm during pregnancy, ideally in the second and third trimester. In those patients who present with an acute abdomen from a ruptured mucinous tumor, initial surgical treatment should be total

appendectomy with meso appendectomy, with biopsy of omental and peritoneal tumor deposits. Since these rarely metastasize to the lymph nodes, extensive surgery in the form of right hemicolectomy is not warranted at the time of initial diagnosis as it can pose undue risk for the mother and fetus.<sup>11</sup> Definitive management of low to moderate grade tumors can be delayed until after delivery as cytoreduction and intraperitoneal chemotherapy are contraindicated during gestation.<sup>12</sup> With optimal management of complete cytoreduction and intraperitoneal chemotherapy, patients with PMP from a well-differentiated appendiceal mucinous carcinoma will have a survival rate of 50% at 15 years.

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