Jelly from the belly button: What's the diagnosis?

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A 38-year-old healthy-appearing woman presented with increasing abdominal girth of 6 months' duration and a 6-day history of an umbilical discharge (*Figure 1*).

To find the answer and an overview of the condition, turn to the following page.

Figure 1. A yellow, gooey, gelatinous substance oozing from the patient's umbilicus.

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ANSWER

This patient had “jelly belly” (JB)—synonymous with, and usually named, pseudomyxoma peritonei (PMP). Here, we refer to it as JB/PMP. The substance oozing from the patient’s umbilicus is pathognomonic of mucinous ascites, often described as jelly belly.^{1–7} This unique type of ascites results from diffuse implants of mucinous tumor on peritoneal surfaces and omentum.

ORIGIN

Results of detailed histopathologic analysis,⁸ molecular genetic studies,⁹ and immunohistochemical investigations¹⁰ provide convincing evidence that the mucinous implants arise almost always from epithelial neoplasms of the appendix. Mucus-producing ovarian tumors—once considered to be a primary site of origin of JB/PMP—are now thought to represent secondary involvement.^{1,2,6,9,11} Rarely, JB/PMP has been associated with malignant tumors of the rectum, colon, small intestine, pancreas, stomach, gallbladder, bile ducts, fallopian tubes, urachus, urinary bladder, breast, and lung.¹²

INCIDENCE

Werth introduced the term PMP in 1884,¹³ and by 1949, approximately 500 cases had been identified worldwide.¹⁴ Since then, the number of documented cases has increased substantially. This condition reportedly affects approximately one person per million per year⁴ and is an unexpected finding at roughly 2 out of every 10,000 laparotomies.¹⁵

Individuals with JB/PMP have ranged from 16 years¹⁶ to 83 years,¹¹ with an average age at diagnosis of 58 years.¹⁷ In most reports, women predominate (up to 75% of the cases^{4,5,17}). There is no apparent correlation with race, ethnicity, or comorbid disease.

PRESENTATION

This malady chiefly presents in one of four ways¹⁸: (1) increased abdominal girth—typically painless, slow in progression, and months to years in duration; (2) symptoms and signs suggesting acute appendicitis; (3) ovarian mass; and (4) new-onset inguinal or umbilical hernia, especially in men.

A striking clinical feature in many of these patients is the disparity between their physical abnormalities and their general well-being.¹⁹

Except for the patient shown here, we found no other reported case of JB/PMP in which jelly spontaneously exited the belly *at presentation*. We did, however, find one example of jelly exiting spontaneously from the right flank, but that event took place 91 months *after* the initial diagnosis.²⁰

HISTOPATHOLOGY

In recent years, at least six different histopathologic classifications of JB/PMP have been proposed.²¹ As a result, the matter remains controversial and, to some extent, confusing. This is unfortunate, because adopting a uniform system of reporting would be helpful for therapeutic purposes.

We and many others favor the classification from the World Health Organization (WHO) in concert with the American Joint Committee on Cancer; it is the easiest to understand and use. The WHO considers any neoplastic proliferation confined to the appendiceal mucosa to be an adenoma. If neoplastic epithelium from a mucinous appendiceal tumor extends beyond the appendix, the tumor is a mucinous adenocarcinoma. Such tumors are then subcategorized as low-grade or high-grade mucinous adenocarcinoma. The American Joint Committee on Cancer provides specific staging guidelines for this classification.²²

Three additional points merit emphasis: (1) the histologic findings in a particular case can change from a less aggressive to a more aggressive form upon repeated surgical and intraperitoneal chemotherapy interventions,^{8,23} (2) the pathologic findings and clinical behavior of the tumor do not always correlate closely with one another,^{14,20,24} and (3) the tumor typically coats abdominal organs but rarely invades them or spreads to distant sites.^{14,25}

DIAGNOSIS

Important clinical clues at presentation include painless and progressive abdominal swelling of many months’ duration, abdominal masses of indefinite contour,¹⁴ the general well-being of the patient despite advanced disease, and ascites that is difficult or impossible to aspirate, even when using a large-bore needle.^{17,26}

Ultrasonic findings can suggest the diagnosis when numerous thick-walled, multiseptate, and predominantly fluid-filled masses are present throughout the abdomen without evidence of visceral invasion or metastases. Matted loops of bowel may be seen as well.²⁷

Computed tomography of the abdomen is particularly useful in determining the extent of disease. The findings include ascites with attenuation values greater than that of water, low-attenuation soft-tissue masses, rim-like calcifications, and compression of abdominal viscera without direct invasion. Scalloping of the hepatic margin from extrinsic compression of the liver by fluid-filled ascitic spaces containing gelatinous material may be specific for JB/PMP.²⁶

In almost all cases, confirmation of the diagnosis requires detection at laparotomy¹⁴ or laparoscopy²⁸ of the jelly-like ascites with mucinous peritoneal implants. Endoscopic ultrasound-guided fine-needle aspiration of an intra-abdominal mass has established the diagnosis in a few cases.⁷ As mentioned earlier, paracentesis often fails because the mucinous ascites is too thick to aspirate through a needle. In that circumstance, the pathognomonic jelly can be extracted from the belly by means of a trocar^{24,*}—an approach seldom used any longer.

TREATMENT

Managing this condition has ranged from watchful waiting²⁹ to various types of surgery, chemotherapy,

* Personal experience of HLF with 2 patients: one in 1957 and one in 1963.

Figure 2. Computed tomographic examination of the patient shown in *Figure 1*. **(a)** The midabdominal axial view depicts ascites (20 Hounsfield units), an umbilical hernia, and fluid-filled cystic spaces in the right peritoneal gutter (top arrows) and at the base of the umbilical hernia (bottom arrows). **(b)** The coronal view shows ascites and a fluid-filled cystic space below the cecum (arrows).

radiation, or combinations thereof. For many years,¹ repeated surgical debulking of the tumor was the mainstay of treatment. More recently, however, many authors have recommended a new treatment protocol for patients with peritoneal spread of appendiceal epithelial neoplasms.^{1,3,4,30–33} This protocol—designed to be curative—consists of cytoreductive surgery (CRS) combined with hyperthermic intraperitoneal chemotherapy (HIPEC). In some cases, early postoperative intraperitoneal chemotherapy is dispensed over a 5-day period, with use of a different agent from the one administered during CRS. The CRS entails stripping of the diseased peritoneum and resection of affected visceral organs, aimed at leaving a minimal amount of residual tumor. Such extensive surgery, however, carries substantial risks, similar to those of a Whipple procedure.³⁴ To avoid or reduce these untoward effects and to derive maximal benefit from this approach, careful patient selection with an optimal level of postoperative care is crucial.

Systemic chemotherapy is ordinarily reserved for patients with advanced disease, recurrent disease, or disease that has spread to lymph nodes or distal sites.^{15,17,25,35}

Radiotherapy includes megavoltage to treat the entire abdomen and pelvis,¹⁵ low-dose voltage for symptom control,²⁰ and intraperitoneal radioisotopes in conjunction with debulking surgery.¹⁵

PROGNOSIS

Without treatment, JB/PMP is fatal.^{5,33} With treatment, however, prolonged survival is common (up to 24 years¹⁶), and the potential for cure is real.³¹ In a multi-institutional study of early and long-term outcomes in 2298 patients with JB/PMP who were treated with CRS and HIPEC, the median survival rate was 16.3 years, with a 15-year rate of 59%.³¹ If the tumor is minimally invasive and cytoreduction is complete, the 20-year survival rate is 70%.³⁰ Therefore, factors that favor a good prognosis are appropriate patient selection, low-grade tumor, effective CRS, combined use of HIPEC and early postoperative intraperitoneal chemotherapy, and the overall experience of the personnel caring for the victims of JB/PMP.

CASE SUMMARY

When this patient presented in June 2007, her abdominal computed tomography examination (*Figure 2*) displayed high-density ascites and a fluid-filled cystic space below the cecum. At laparotomy, a yellow, jelly-like material filled her peritoneal cavity and firmly encased the entire abdominal and pelvic contents, making it impossible to identify or biopsy any specific structure.¹⁴ Multiple sections of the peritoneum, umbilicus, and ascitic fluid showed no tumor cells. Nevertheless, with a presumptive diagnosis of mucocele or mucinous adenocarcinoma of the appendix, she responded well to several rounds of oral chemotherapy.

For the next 8 years, her disease remained stable. Then, in 2015, progressive abdominal discomfort prompted laparotomy, which revealed a large cecal mass with prominent contiguous lymph nodes. Microscopic samples of the mass and nodes disclosed mucinous adenocarcinoma. Her condition slowly deteriorated, and she died in November 2016. There was no autopsy.

TAKE-HOME POINTS

- “Jelly” (mucinous ascitic fluid) in the belly is pathognomonic of pseudomyxoma peritonei.
- The cause is almost always a benign or malignant mucinous epithelial tumor of the appendix.
- Characteristically, the tumor coats but does not invade other intra-abdominal organs and rarely spreads beyond the peritoneum.
- In many cases, a striking clinical feature is the disparity between the prominent physical abnormalities and the patient’s general well-being.
- Typically, the clinical course is indolent and, with treatment, survival for 20 years or longer is possible.

ORCID

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