Vernix caseosa peritonitis presenting post partum as acute cholecystitis

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Vernix caseosa, the “cheese-like” white cutaneous material covering the skin of newborns, is a very unusual cause of peritonitis. Only 15 cases of this entity have been described.1–13 Although spillage of amniotic fluid into the peritoneal cavity is almost inevitable after cesarean section, this spillage is usually insignificant and only rarely does it progress to peritonitis.1 The exact mechanism for the development of peritonitis in these few cases is unknown.12

We present a case of peritonitis due to vernix caseosa post partum in a 27-year-old woman whose presentation mimicked acute cholecystitis.

Case report

A 27-year-old woman, gravida 1, para 0, was initially seen by an obstetrician for premature rupture of her membranes. Despite receiving oxytocin intravenously for a full day, her labour did not progress and her cervix remained 8 to 9 cm dilated. After receiving 5 million units of intravenous penicillin G, prophylactically, a low transverse cesarean section was performed and a live male fetus, weighing 3420 g, was delivered whose Apgar scores were 9 and 10 at 1 and 5 minutes, respectively. The fetus was approximately 38 weeks’ gestation. There was no documented leakage of amniotic fluid into the peritoneal cavity. In addition, meconium staining of fluid was not identified. The patient’s postoperative recovery was unremarkable and she was discharged several days later.

Six days post partum, the patient was readmitted, suffering from fever, chills and pain in her right upper quadrant. She noted the discomfort followed a fatty meal. Laboratory investigations demonstrated a marked leukocytosis and both her alkaline phosphatase and γ-glutamyl transpeptidase levels were elevated (223 U/L and 65 U/L respectively). Urine, sputum and blood cultures taken at readmission gave negative results. Ultrasonography of the upper abdomen demonstrated fluid in the area of the gallbladder in keeping with acute cholecystitis (Fig. 1). In addition, 2 bright echogenic areas, as well as “sludge,” were identified within the gallbladder. Both general and hepatobiliary surgical consults concluded with a working diagnosis of acute cholecystitis and the patient was scheduled for a laparoscopic cholecystectomy.

During the procedure, numerous areas of cheesy-white exudate with surrounding erythema were detected on the peritoneum of the right upper and lower quadrants. Several mesenteric deposits were also identified (Fig. 2). Significant bowel–bowel and bowe–anterior abdominal wall adhesions were identified. There was no gross evidence of acute inflammation or thickening of the gallbladder wall. After discussion and an intraoperative obstetric/gynecologic consult, a decision was made to remove the gallbladder. Findings on laparoscopic examination of abdominal and peritoneal contents, performed to rule out other possible causes of peritonitis, were negative. In addition, biopsy of several foci of the peritoneal exudate were taken and sent for routine pathological examination. The appearance of the abdomen was captured on video for future reference.

Microscopic examination of the peritoneal exudates demonstrated inflammatory granulation tissue containing an admixture of neutrophils, lymphocytes, eosinophils and histiocytes aggregated about anucleate squames (i.e., necrotic squamous cells) having wrinkled cell borders and resembling the cellular component of vernix caseosa (Fig. 3). No foreign body giant cell reaction was present, and no histiocytes with intracytoplasmic brown meconium pigment were identi-
fied. The nature of degenerated squamous cells was confirmed by positive immunoperoxidase staining for keratin. Gram’s staining did not demonstrate any bacteria, and lanugo hairs were not identified either microscopically or histochemically. Sections of the gallbladder demonstrated cholesterosis and a mild eosinophilic cholecystitis. No acute cholecystitis or gallstones were identified.

Postoperatively, the patient recovered well and was discharged on postoperative day 5 with a 7-day prescription for cephalexin 500 mg orally every 6 hours and metronidazole 500 mg orally every 12 hours.

Discussion

Vernix caseosa, the “cheese-like” white cutaneous material covering the skin of the newborn, consists of an admixture of sebaceous glandular secretions, lanugo hair and desquamated squamous cells. These squamous cells are derived from fetal periderm. Normal amniotic fluid consequently contains numerous squamous cells. During elective cesarean section, the integrity of the amnion is necessarily disrupted, and its contents may enter the peritoneal cavity. The amount of such peritoneal contamination during the course of cesarean section is usually not recorded. The onset of peritonitis due to vernix caseosa occurring after uncomplicated cesarean section has been attributed to incomplete peritoneal lavage of spilled amniotic fluid. However, both Davis and associates and Schwartz and colleagues have reported that peritonitis due to vernix caseosa can be initiated by antenatal or intrapartum leakage of amniotic fluid. Suggested mechanisms include uterine–tubal reflux and unrecognized uterine perforation.

Although the exact cause of the subsequent development of peritonitis is not known, a number of theories have been suggested. Both meconium and keratinized squamous cells have demonstrated the capacity to induce a brisk inflammatory response, collectively or individually, most probably by mechanical irritation: the former in the setting of neonatal meconium peritonitis secondary to intestinal perforation and the latter in the context of ruptured keratinous cysts, dermoid cysts and keratinizing neoplasms. Interestingly, meconium is composed largely of swallowed amniotic fluid (which contains anucleate squamous cells and lanugo hair derived from vernix caseosa). Furthermore, it is recognized that direct spillage of meconium into the peritoneal cavity can incite a granulomatous peritonitis.

FIG. 2. Top: laparoscopic view of cheese-like white and yellow patches (arrows) on the parietal peritoneum of the right upper quadrant. Bottom: laparoscopic view of these patches (arrows) with surrounding erythema on the large-bowel mesentery.

FIG. 3. Anucleate squamous cells surrounded by numerous histiocytes and neutrophils. No giant cells are present (hematoxylin–eosin stain; original magnification ×500).
The concentration of vernix caseosa in amniotic fluid may have pathogenetic significance. Partial loss of amniotic fluid may be followed by increased concentrations of vernix in the remaining fluid. Difficult labours may be associated with higher concentrations of vernix due to excessive manipulation.\(^\text{10}\) Subsequent leakage on rupture of amniotic membranes may increase the likelihood of peritonitis.

Clinical diagnosis of peritonitis due to vernix caseosa should be considered in all patients presenting with an acute abdomen after elective or emergency cesarean section. Although the other causes of peritonitis such as appendicitis, cholecystitis and a perforated viscus are still more common than vernix caseosa peritonitis, surgeons should be aware of this rare but interesting condition.

Certain gross appearances are characteristic of peritonitis due to vernix caseosa: cheese-like white and yellow patches on the peritoneum and serosal surfaces of many visceral organs, such as the appendix and omentum, are suggestive. Extensive adhesions involving the uterus are characteristic. The absence of inflamed organs with the presence of white patches on the serosal surfaces is the sine qua non of vernix caseosa peritonitis.\(^\text{11}\)

Histologically, the cheesy white peritoneal exudate is of a fibrinous nature, containing epithelial squames, lanugo and, probably, sebum. The characteristic desquamated squamous cells appear singly or in clumps and are polygonal or ovoid in shape. The nuclei are absent, but nuclear remnants are frequently present. I increased acid-phosphatase enzyme activity has been reported to be characteristic.\(^\text{12}\) Cases of peritonitis due to vernix caseosa occurring shortly after delivery have neutrophils as the predominant primary reactant. After several days, both an acute and foreign-body inflammatory reaction are usually evident. Cases occurring at a longer interval after delivery have predominantly granulomatous foreign-body reactions.\(^\text{13}\)

Although Tawfik and colleagues\(^\text{14}\) suggested evaluation by preoperative computed tomography and fine-needle aspiration in suspected cases of peritonitis due to vernix caseosa, this may not always be practical and many patients may undergo surgical exploration. Intraoperatively, when this condition is suspected, biopsy of the white serosal patches is the most definitive method of confirming a pathological diagnosis. In addition to the biopsy, the usual protocol for surgical peritonitis should be pursued; there should be complete exploration to rule out the more common forms of peritonitis. Samples of peritoneal fluid should be taken for culture and the peritoneal cavity should be irrigated in the usual manner. Although a cholecystectomy was performed in this case, Mahmoud and colleagues\(^\text{15}\) did not recommend removal of any noninflamed visceral organ, even when it is covered with the characteristic white or yellow patches. Indeed, if the pathologist, consulted intraoperatively, can recognize the process, unnecessary removal of organs may be prevented. In a study of 8 cases, George and associates\(^\text{16}\) reported that 2 patients underwent unnecessary partial colectomies and 1 underwent a total hysterectomy with bilateral salpingo-oophorectomy.

Postoperative treatment with antibiotics should be continued and later adjusted according to the culture results. Although the use of steroids postoperatively has been reported to have a dramatic effect, these should be used carefully and only after the diagnosis has been confirmed pathologically; the possibility of residual abscesses should also be excluded. In their report of 2 cases, Mahmoud and colleagues\(^\text{17}\) found a notable response when steroids were used for 1 to 2 weeks, then tapered off gradually over 2 more weeks.

Conclusions

Vernix caseosa peritonitis is an infrequent complication of cesarean section and appears to be poorly recognized by clinicians. Surgeons should be aware of this uncommon cause of peritonitis, especially when consulted concerning a woman with acute abdomen or peritonitis post partum. Prompt recognition by surgeons, gynecologists and pathologists may help prevent unnecessary clinical work-up and surgery.

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References