Intra-peritoneal rupture or perforation into adjacent abdominal viscera is a rare complication of benign cystic teratoma (dermoid cyst). We report a 48-year-old woman in whom an ovarian dermoid cyst perforated into a loop of the small bowel. The patient had co-existing typhoid fever, which probably predisposed to this complication. She recovered on antibiotic therapy and after surgical excision.

Dermoid cyst is the most common benign neoplasm of the ovary. They may present with acute abdominal pain because of torsion, infection or rupture. Spontaneous rupture of the cyst is extremely rare because of its thick wall, and is reported in 0.3%-0.7% of cases. The cyst can rupture into the peritoneal cavity or rarely into a hollow abdominal organ. The reported sites are urinary bladder, small bowel, rectum, sigmoid colon and vagina.

A 48-year-old postmenopausal woman presented with continuous fever and dull pain in the lower abdomen for 10 days. On examination she was febrile and had mild pallor. Abdominal examination revealed a 20 cm x 15 cm, firm, non-tender, well-defined mass in the lower abdomen. On pelvic examination, the mass was felt through all vaginal fornices.

Investigations: hemoglobin 7 g/dL, leukocytosis with relative neutropenia. Widal test was positive (titer 1:360) and blood culture grew Salmonella typhi. Serum CA-125 level was 130 IU/mL (normal level <35). Plain radiograph, ultrasonography and CT scan suggested a dermoid cyst of the ovary. The patient was started on intravenous ciprofloxacin for typhoid fever.

Three days later she had sudden-onset diarrhea with passage of mucoid and cheesy stools. Vital signs were normal. Abdominal examination revealed marked reduction in the size of the mass without signs of peritonitis. Exploratory laparotomy showed a 10 cm x 8 cm right ovarian cystic mass adherent to the small intestine and omentum. A communication was detected between the tumor and the ileum. A tuft of hair and cheesy material was seen in the lumen of the ileum. The uterus and left ovary were normal. The fistulous opening in the ileum was excised and the defect was closed. Rest of the small bowel was normal. Total abdominal hysterectomy and bilateral salpingo-oophorectomy was performed. She had uneventful postoperative course. The patient was continued on ciprofloxacin for 14 days.

Grossly the tumor contained sebaceous material, hair and a tooth. Microscopic examination confirmed the diagnosis of benign cystic teratoma. The resected margins of the bowel showed inflammation.

Spontaneous rupture of a dermoid cyst, usually seen with large tumors, is now less frequently reported due to greater use of elective surgery for small lesions. Possible predisposing factors include 1) adhesion of the tumor to an adjacent structure resulting in ischemia and necrosis of the cyst wall, 2) infection of the cyst, 3) trauma of labor and, rarely, 4) malignant change in a dermoid cyst. In our case, salmonella infection resulted in small intestinal inflammation that caused adhesion of the dermoid cyst to the bowel wall, eventually leading to fistula formation. This was fortunate, since it prevented contamination of the peritoneal cavity by contents of the cyst and intestines. Acute peritonitis due to intraperitoneal rupture of dermoid has been reported.

Sudden reduction in the size of intra-abdominal masses should prompt clinicians to suspect rupture and perform early exploratory laparotomy.

References

Correspondence to: Dr Maheshwari. Fax: (22) 2414 6937. E-mail: maheshwariamita@yahoo.com

Figure: CT scan of abdomen showing dermoid cyst with fat-fluid level and calcifications; cyst is adherent to loop of intestine.
Triple A syndrome (Allgrove syndrome) is an autosomal recessive disorder consisting of achalasias, alacrima and Addison insufficiency. We report an 11-year-old girl with predominant symptom of achalasia who was diagnosed as Triple A syndrome almost 3 years after initial presentation. [Indian J Gastroenterol 2005;24:217-218]

Triple A syndrome (Allgrove syndrome) is an autosomal recessive disorder consisting of achalasia, alacrima and Addison insufficiency due to mutation in AAAS gene on chromosome 12q13. In view of variable order of presentation and marked phenotypic variation, this association is often diagnosed late, sometimes even in adulthood.

An 11-year-old girl presented with dysphagia for 3 years, which was intermittent and associated with vomiting off and on. Dysphagia was not related to type of food and vomiting was esophageal in nature. There was generalized hyperpigmentation noticed in the past year. The parents also felt that she was not thriving well in recent years. She was born of nonconsanguinous marriage. Her parents and two younger siblings were asymptomatic.

Barium swallow and upper GI endoscopy had been done 3 years ago but no reports were available. She had been started on nitrates but had no improvement in dysphagia.

On examination her weight was 22 Kg, height 135 cm, BP 90/70 mmHg with no postural drop. She had generalized hyperpigmentation and right-sided pes cavus. Barium swallow and upper GI endoscopy had been done 3 years ago but no reports were available. She had been started on nitrates but had no improvement in dysphagia.

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Barium swallow showed narrowing of the esophagus at its lower end with trickle of contrast through the gastroesophageal junction (bird-beak appearance), proximal hold-up of contrast, and tertiary contractions, suggestive of achalasia cardia. Upper GI endoscopy was also suggestive of achalasia.

Basal (8 AM) cortisol was 70 microgram/mL (normal 193-770), Serum electrolytes were normal. Ultrasonography did not show the adrenal glands. Ophthalmological tear break-up test and Schirmer test suggested alacrima.

The patient underwent fluoroscopy-guided pneumatic esophageal dilatation. She was also started on cortisol supplement and artificial tear drops, and has been advised to increase the steroid dose during stressful events. She is symptomatically much better for the past 3 months.

Triple A syndrome is rare; there are only 23 cases and 8 of its variant previously reported in literature.

Alacrima is usually the earliest manifestation. The patient often reports to a specialist with one complaint, and the association may remain unsuspected, leaving the patient prone to complications like Addisonian crisis, recurrent aspiration, and failure to thrive. All children presenting with one of the three symptoms should be screened for triple A syndrome. Since inheritance and gene for the association is known, early diagnosis can allow genetic counseling with triple A syndrome, as in our case, has been reported earlier.

References

Correspondence to: Dr Bharadia, S R Kalla Memorial Gastro and General Hospital, 78 Dhuleshwar Garden, Behind HSBC Bank, Sardar Patel Marg, C Scheme, Jaipur, Rajasthan. Fax: (141) 511 2043. E-mail: lalitbhharadia@rediffmail.com

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Strangulated femoral hernia presenting as parietal wall emphysema

Sanjay Marwah, N Marwah, Dalbir Singh Sandhu, R K Karwasra

Departments of Surgery and Pathology, Post Graduate Institute of Medical Sciences, Rohtak (Haryana) 124 001

Surgical emphysema of abdominal and thoracic wall, along with features of intestinal obstruction, has not been reported as a complication of strangulated femoral hernia. We report a 31-year-old woman with such a presentation. [Indian J Gastroenterol 2005;24:218-219]

The diagnosis of strangulated femoral hernia is clinically challenging. We report a patient with strangulated hernia presenting with parietal emphysema.

A 31-year-old woman was admitted in hemodynamic shock, with history of pain in abdomen, vomiting and distension of the abdomen since 10 days. She also complained of progressively increasing swelling in the right lower abdomen for three days. On examination, the abdomen was distended and tender; bowel sounds were absent. The abdominal wall in the right iliac fossa was edematous, red, fluctuant and crepitant. The edema and crepitation extended from the groins to the axilla; this finding was confirmed on X-ray (Fig). There was no clinical evidence of groin hernia.

A clinical diagnosis of parietal wall necrotizing fasciitis with reflex paralytic ileus was made and the patient was taken up for drainage and debridement after resuscitation. A vertical incision was made in the right iliac fossa and feculent fluid with foul-smelling gas was drained from...