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The Radiographic and Ultrasonographic Evaluation of Enteric Duplication Cysts

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Abstract. The pattern of diagnostic evaluation of seven children with duplication cysts of the gastrointestinal tract (two with bronchopulmonary foregut malformations and five with mid or hindgut duplications) is presented. Each child had plain radiographs followed by ultrasonic scans and appropriate confirmatory radiographic studies with contrast. Scans showed a well defined cystic mass in each case. Radiographs were able to identify more precisely the site of origin of the duplication cyst.

Key words: Duplication cysts – Pediatric ultrasonography

Among those patients who had had an abdominal mass evaluated by ultrasound in our institution, we counted five who each had a duplication cyst of the intra-abdominal gastrointestinal tract and two who had bronchopulmonary foregut cysts. These seven are the subjects of this report. Children who have a well defined anterior abdominal mass are evaluated with plain radiographs and then ultrasound. If further diagnostic refinement is necessary, radiographic examinations with contrast are done as determined by the scans.

Case Reports (see Table 1)

Discussion

A duplication cyst of the gastrointestinal tract is an embryological mistake that may or may not cause symptoms depending on its location, size and histology. There are some interesting theories to explain its presence in an otherwise healthy patient. Lewis and Thyng [1] and later, Ladd and Gross [2] thought persistent embryonic diverticula became duplication cysts. Edwards [3] thought they were the result of attempted twinning and Hudson [4] considered the possibility that duplication represented phylogenetic reversion, citing the double cecum of birds as an example. The most popular theory, proposed by Bremer in 1944, considers duplications errors of canalization [5]. The solid gastrointestinal tract of the 6 week embryo becomes the hollow tube of the 8 week embryo as multiple vacuoles coalesce [6]. Two channels rather than one channel through some part of this solid cord results in a duplication cyst paralleling the normal lumen.

In the last 10 years there have been two large reviews of patients with enteric duplications. Grossfield et al. presented a clinical series of 23 patients seen over a span of 18 years [7]; Bower et al. reviewed an experience of 78 duplications in 64 patients over a period of 40 years [8]. This latter series included both clinical and autopsy data. Some general statements concerning duplications can be made on the basis of these and other articles. The diagnosis usually is made in the first year of life, often in the first week. Although cysts related to bronchopulmonary foregut malformations are often diagnosed because of respiratory distress, a small number of patients are asymptomatic. Obstruction is the most common symptom of an abdominal duplication cyst and the ileum is the most common site. Tubular colonic duplications are associated with anomalies of the genitourinary tract. Duplication of the stomach usually occurs on the greater curvature; more distal duplication cysts are on the mesenteric side of the intestine.

The five children with abdominal duplication cysts that we studied came to medical attention because of obstruction (three cases), gastrointestinal

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Table 1. Case reports

Case	Patient (Sex/age)	Symptoms & signs on presentation	Ultrasound	Radiology	Surgery	Pathology of duplication
1	M 5 mo	Vomiting	Cystic subhepatic mass (Fig. 1a)	Parapyloric mass on UGIS (Fig. 1b)	Resection of pyloric duplication	Antral gastric mucosa
2	M 4 wk	Vomiting; RUQ mass	Cystic subhepatic mass (Fig. 2a)	Ileocecal mass on SBFT (Fig. 2b)	Resection of ileocecal duplication	Ileal mucosa
3	F 15 d	Asymptomatic; RLQ mass	RLQ cystic mass	Ileocecal mass on BE	Resection of ileocecal duplication	Ileal mucosa
4	M 1 mo	Bleeding per rectum; RUQ mass	Cystic subhepatic mass	Ileocecal mass on SBFT	Resection of ileocecal duplication	Eroded ileal mucosa
5	M 3 wk	Vomiting; LLQ mass	Cystic mass L mid-abdomen (Fig. 3a)	Ileal mass on retrograde SB study (Fig. 3b)	Resection of ileal duplication	Ileal mucosa
6	M 14 yr	Asymptomatic; gastric mass on chest X-Ray (Fig. 4a), prior hx of "bronchogenic cyst" removed at 3 mo. of age	Cystic mass anterior & lateral to aorta (Fig. 4b & 4c)	Paraesophageal mass on barium swallow (Fig. 4d)	Cyst marsupialized to stomach	Bronchopulmonary foregut malformation. Metaplastic squamous, columnar & pseudostratified columnar epithelium
7	M 1 wk	Multiple anomalies (imperforate anus, hypoplastic right heart, tricuspid atresia); asymptomatic mediastinal mass on chest X-Ray	Cystic mass from R chest straddling esophageal hiatus, lying anterior to aorta, posterior to IVC	Tubular mass indenting esophagus on barium swallow	Resection of tubular mass from R chest & which shared common wall with esophagus	Bronchopulmonary foregut malformation. Squamous & respiratory epithelium, pancreatic tissue, cartilage & smooth muscle

Abbreviations: UGIS = upper gastrointestinal series; SBFT = small bowel follow through; SB = small bowel; BE = barium enema; RUQ = right upper quadrant; RLQ = right lower quadrant; LLQ = left lower quadrant; hx = history; IVC = inferior vena cava

bleeding (one case) or incidental abdominal mass (one case). All had a mass when examined carefully.

The ultrasonic examination of a child with an abdominal mass aims to characterize and measure the mass and to identify its site of origin. If a cystic mass is subhepatic, careful scans of the liver and biliary tree can establish whether or not it is related to either [9]. In each patient who had a duplication cyst, examination with ultrasound revealed a unilocular cystic mass which was either spherical or tubular in shape. The greatest diameter of the cysts ranged from 3 to 6 cm. A previous report suggested that an inner echogenic rim, possibly a sign of mucosal lining or secretions might be helpful in diagnosing duplication cysts [10]. None of our five patients demonstrated this sign. None had significant echogenic debris within the cyst – a potential finding if there had been bleeding into the cyst [10, 11]. The debris seen on scans done on Case 1 was not from hemorrhage.

The site of the duplication cyst cannot be predicted from its location on ultrasonic scans. The pyloric duplication and two of the three ileocecal duplications were subhepatic. The presence of an ileocecal duplication probably prevents normal fixation of the cecum in the right lower quadrant. The patient with the more proximal ileal duplication cyst had an evanescent mass on physical examination; the cyst was extremely mobile and moved with the bowel.

The differential diagnosis of an anterior abdominal unilocular cyst includes mesenteric or omental cyst, an exophytic hepatic cyst, pancreatic pseudocyst, and in a girl, ovarian cyst. A choledochal cyst usually can be differentiated because of its intimate relationship to the biliary tree and pancreas. Duplication cysts are more likely to obstruct the bowel than the other cystic masses listed above. Further radiographic evaluation of a suspected duplication cyst includes barium studies of the gastrointestinal tract. These allow accurate localization of the mass. If plain films suggest complete obstruction of the upper gastrointestinal tract (Case 5) it is best to do a barium enema. If the obstruction is not encountered in the colon, retrograde study of the small bowel can follow.

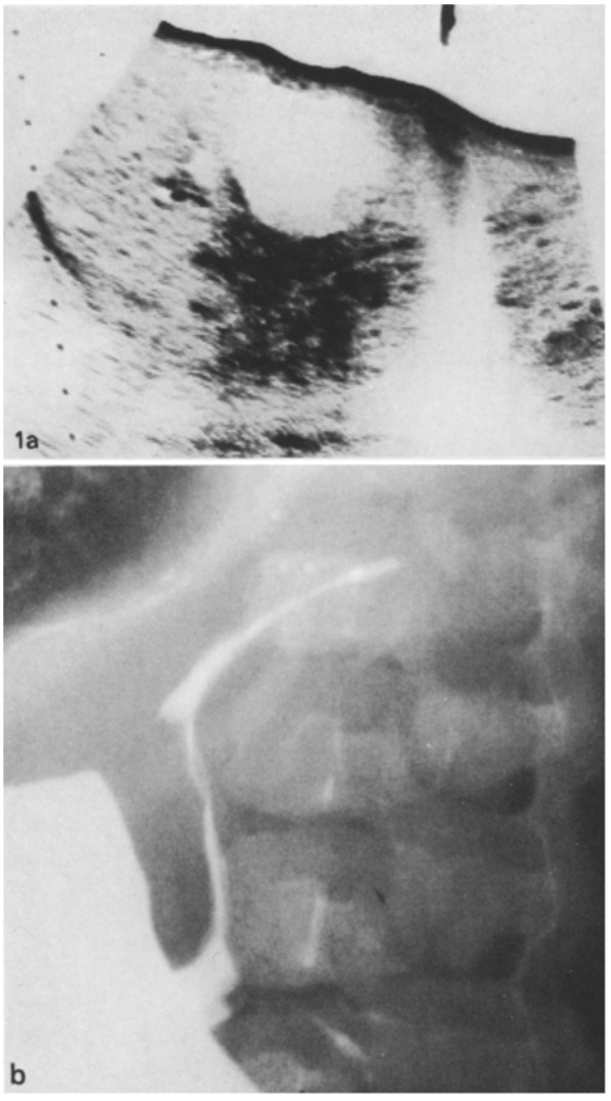

Fig. 1a and b. Case 1: **a** This cystic subhepatic mass was distinct from the biliary tree and gallbladder. Particulate debris is present in the base of the cyst. **b** Spot film of pylorus from upper gastrointestinal series shows an extrinsic mass compressing the normal lumen

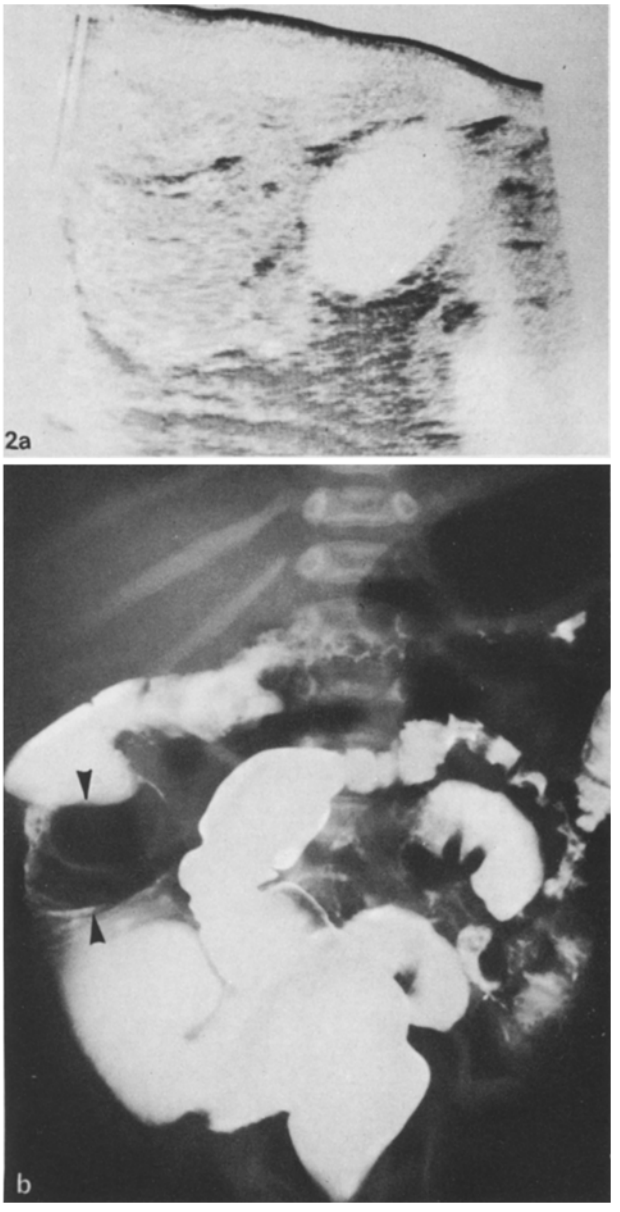

Fig. 2a and b. Case 2: **a** This cystic subhepatic mass is almost identical to that in Case 1 (Fig. 1a). **b** Radiograph taken 24 hours following upper gastrointestinal series shows an extrinsic mass associated with the ileocecal valve in the right upper quadrant. (arrows)

If complete obstruction is not present, an upper gastrointestinal series with follow through until the mass itself is identified is the appropriate examination (Cases 1 & 2).

Bronchopulmonary foregut malformations occur between the 5th and 7th weeks of embryologic life and are more complicated than duplications of the mid and hindgut. They probably result from the genesis of a supernumerary lung bud that migrates along with the growing esophagus [12]. Pulmonary sequestrations – intralobar and extralobar –, esophageal or gastric diverticula, bronchogenic cysts and esophageal duplications are probably all variants of this embryologic aberration.

Enteric cysts also may occur in association with anomalies of the spinal cord and meninges. Their

pathogenesis appears to be different from that of enteric cysts which are part of bronchopulmonary foregut malformations. In early intrauterine life, the embryo has a neurenteric canal which connects ectoderm (amnion) to endoderm (yolk sac) by passing through the dorsal neural folds [6]. Persistence of this or an accessory neurenteric canal could give rise to a series of anomalies including diastematomyelia, hemivertebrae and enteric cysts [13]. Cases 6 and 7 in our series fit best into the group of bronchopulmonary foregut malformations by virtue of their location, pathology and lack of associated vertebral anomalies. The ultrasonographic findings in a patient with neurenteric cyst would, no doubt, be similar.

Esophageal cysts often contain heterotopic mucosa whereas the more distal duplication cyst tends to have the same type of mucosa as the paralleling bowel. The foregut cyst may become apparent by bleeding because the heterotopic mucosa may ulcerate.

Respiratory distress is the usual presenting sign of a thoracic duplication cyst. The two patients that we studied who had an esophagogastric duplication as part of a bronchopulmonary foregut malformation were both asymptomatic. They came to medical attention because of the presence of a mediastinal or gastric mass on radiographs of the chest. They each had a cystic mass straddling the esophageal hiatus on ultrasound examination. The major differential diagnosis in this type of duplication is pancreatic pseudocyst and clinical information is helpful in eliminating this possibility. Barium swallow usually completes the evaluation. It defines the relationship between duplication cyst and normal esophagus or stomach prior to surgery.

Conclusion

The child with an intestinal duplication may present with obstruction of the gastrointestinal tract, gastrointestinal bleeding or be asymptomatic. Careful clinical examination will detect a mass in most cases. The mass is evaluated further by plain radiographs of the abdomen or chest and by ultrasound. The appropriate studies with contrast localize the cyst more precisely prior to surgery.

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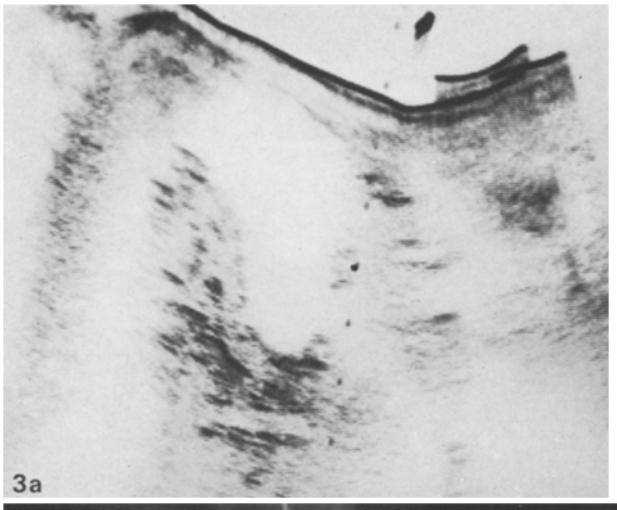

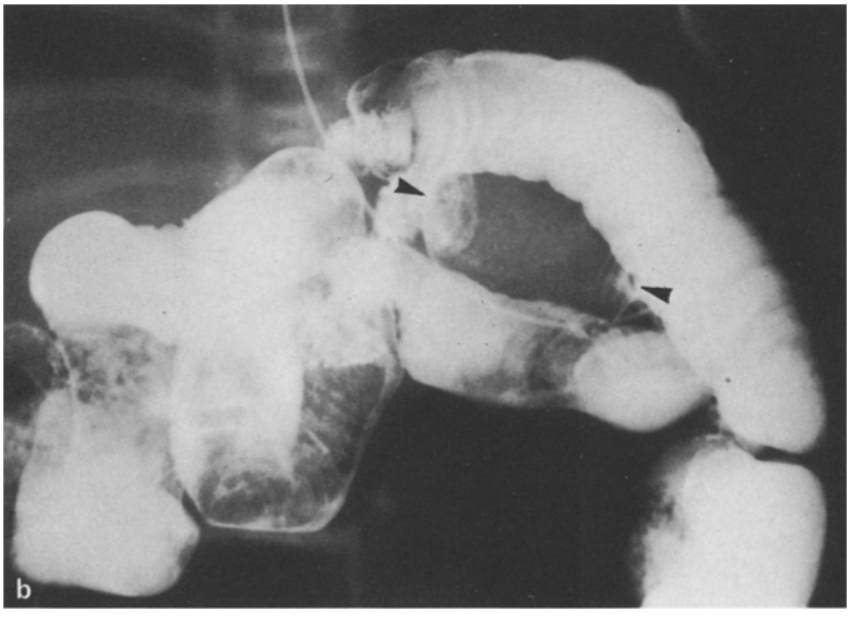

Fig. 3 a and b. Case 5: **a** This decubitus scan over the left flank shows a tubular cystic mass. **b** On retrograde study of the small bowel, a mass (arrows), causing compression of adjacent ileum, was encountered in the upper abdomen

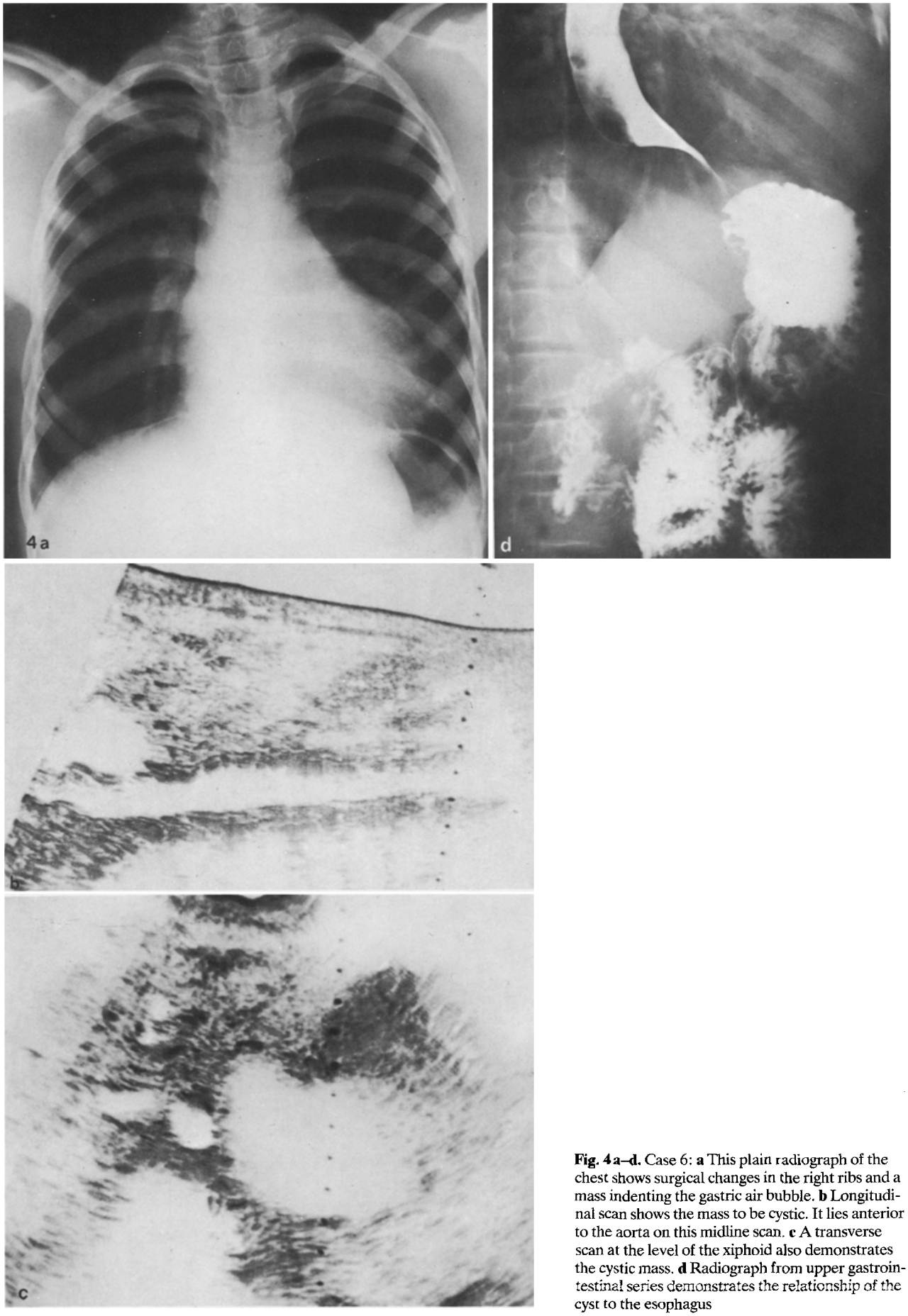

Fig. 4a–d. Case 6: **a** This plain radiograph of the chest shows surgical changes in the right ribs and a mass indenting the gastric air bubble. **b** Longitudinal scan shows the mass to be cystic. It lies anterior to the aorta on this midline scan. **c** A transverse scan at the level of the xiphoid also demonstrates the cystic mass. **d** Radiograph from upper gastrointestinal series demonstrates the relationship of the cyst to the esophagus

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