

Thoracic, thoraco-abdominal, and abdominal duplication

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Abdominal enteric duplication is a relatively common congenital abnormality which is well annotated in the literature. Thoracic accessory enteric formations are less common, and thoraco-abdominal duplications are even more rare. The following case illustrates all forms of enteric duplications as described by Smith (1960), and no similar case can be found in the literature.

CASE REPORT

Christine G. aged 8 years was admitted to this unit for elective thoracotomy for a bi-lobed right-sided posterior mediastinal lesion.

At the age of 7 months she had been admitted to another hospital with profuse melaena. On 1 May 1956 laparotomy revealed a small bowel duplication 50 cm. in length, terminating distally in what appeared to be a Meckel's diverticulum. At the base of the diverticulum an ulcer was found. Conservative resection was undertaken owing to the poor condition of the infant. Further melaena necessitated a second laparotomy on 16 May, and the remaining part of the duplication and adjacent small bowel was resected (Fig. 1).

Histology revealed the mucosa around the ulcer to be intestinal in type but that of the remaining duplication resembled gastric mucosa with oxyntic cells. Post-operative respiratory difficulties necessitated the taking of a chest radiograph which showed a bi-lobed lesion in the right hemithorax. At laparotomy it had been noted that the right kidney appeared enlarged, and a subsequent intravenous pyelogram demonstrated wide separation of the calyces of the right kidney suggestive of polycystic disease. The left kidney was normal (Fig. 2).

The child had no chest symptoms but it was decided to explore and, if possible, remove the intrathoracic mass.

On admission to this unit in 1963 examination of the child's chest revealed dullness to percussion over the base of the right lung posteriorly with diminished air entry in this area. Otherwise the respiratory system was clinically normal. On examination of the cardiovascular system the apex beat of the heart was found to be 0.5 in. (12.7 mm.) lateral to the left mid-clavicular line in the fifth intercostal space. The

cardiovascular system was otherwise normal. In the abdomen a rounded, smooth, soft mass was palpable in the right hypochondrium. The mass was dull to percussion, and the liver could not be identified separately from the mass. There was no clinically detectable spinal deformity. A radiograph of the chest revealed two smoothly rounded opacities lying posteriorly in the right hemithorax. The lower opacity showed a small plaque of calcification (Fig. 3).

Radiographs of the spine revealed no gross abnormality but some scalloping of the anterior surface of the second dorsal vertebra; some loss of disc space between the fifth and sixth dorsal vertebrae could be demonstrated on the lateral projection films.

On 29 October 1963 a right thoracotomy was made in the face-down position through the bed of the sixth rib. A lobulated cystic structure was present in the posterior mediastinum which extended down to the right diaphragm. Aspiration proved that these were two cysts. The upper cyst was found to be firmly adherent to the sixth dorsal vertebra. No oesophageal or tracheal attachment of either cyst could be demonstrated. The inferior cyst was opened, and probing

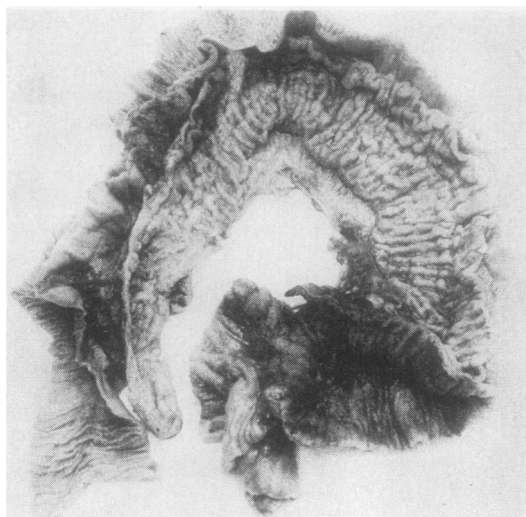


FIG. 1. Resected ileum with associated duplication.

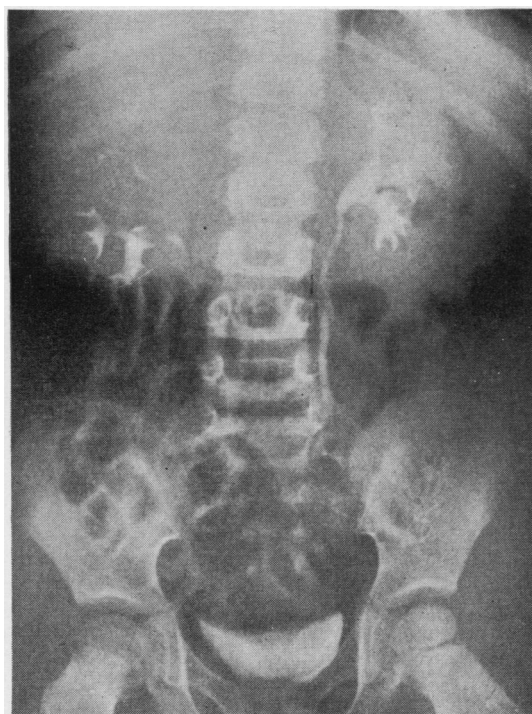


FIG. 2. *Intravenous pyelogram showing distortion of the calyceal pattern of the right kidney.*

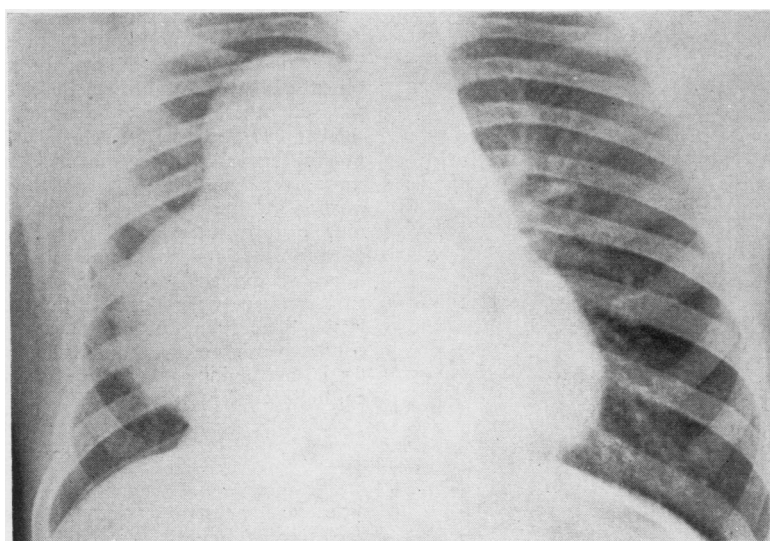


FIG. 3. *Chest radiograph showing bilocular opacity in right hemithorax.*

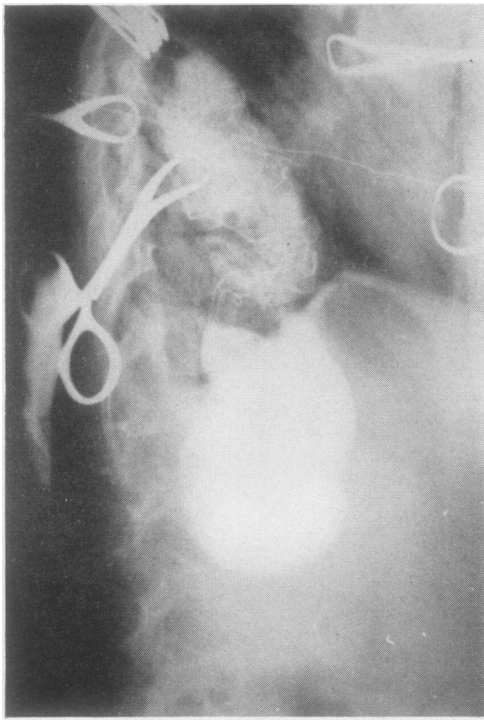


FIG. 4. Lateral radiograph, taken at operation, demonstrating extent of abdominal portion of thoraco-abdominal cyst.

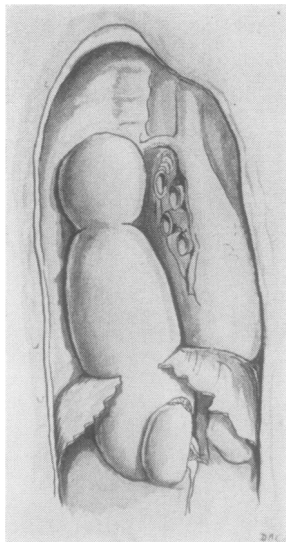


FIG. 5. Diagrammatic representation of position and extent of the thoracic and thoraco-abdominal duplications.

revealed that it extended into the abdomen through the right hemidiaphragm. The posterior margin of the diaphragmatic aperture was the right median arcuate ligament. The size and extent of the abdominal portion of the thoraco-abdominal cyst was demonstrated by radio-opaque packing and radiography (Fig. 4).

As it was impossible to remove the abdominal extension of the lower cyst by the approach in use, a right lumbar incision was made, excising the right twelfth rib. In mobilizing the abdominal portion of the cyst, it was found to be densely adherent to the diaphragm throughout its circumference but was easily freed from the inferior vena cava and vertebral bodies to which it was closely related. The right kidney was of normal size but grossly flattened to a disc over the anterior aspect of the abdominal portion of the cyst. This portion of the cyst was entirely retroperitoneal, and no communication with the stomach, duodenum or intestine could be demonstrated (Fig. 5).

The diaphragm was repaired by stitching the anterior border of the defect to the right median arcuate ligament, and the chest was closed. Post-operative progress was uneventful except that difficulty was experienced in achieving sufficient expansion of the right lung to fill the right hemithorax after excision of the large cysts.

PATHOLOGY The content of both cysts was opalescent fluid containing cholesterol crystals. The upper cyst was about 8 cm. in diameter with a wall approximately 2 mm. in thickness. The thoraco-abdominal cyst was about 20 cm. long, the abdominal portion being about 7 cm. in diameter with a wall approximately 2 mm. thick, and the thoracic portion about 10 cm. in diameter, the wall being slightly thinner.

Histology The thicker parts of the walls of both cysts consisted of three smooth muscle layers. The outer two layers of muscle fibres lay at right angles to each other, the inner layer of the two being the thicker. The third innermost layer, or muscularis mucosae, was of the same orientation as the outermost layer. The thinner parts of the wall in the thoracic portion of the lower cyst showed scattered smooth muscle fibres interspersed with areas of hyaline connective tissue and streaks of calcification. In scattered areas of the walls of both cysts, nerve fibres and ganglion cells were identified in relation to the muscle layers. The lining of both cysts was mostly coarse connective tissue or chronic inflammatory granulation tissue, but definite areas of both cysts were lined with epithelium of gastric type (Fig. 6).

DISCUSSION

The case presented demonstrates the described varieties of accessory enteric formations (Smith, 1960). The abdominal abnormalities initially described are typical of many recorded cases of

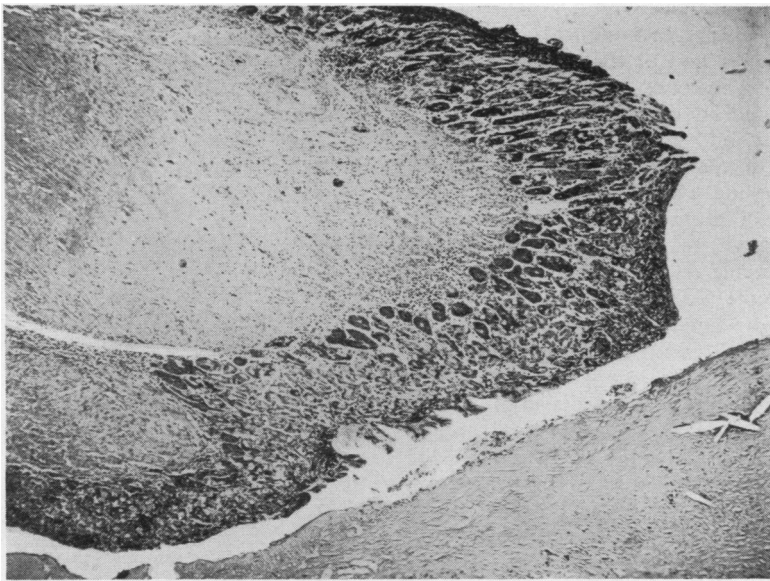


FIG. 6. Photomicrograph showing cyst lining with epithelium of gastric type. H. and E. $\times 45$.

Meckel's diverticulitis and small gut duplication. The case also illustrates one of the complications of the condition, *i.e.*, bleeding. The theories of origin of these abnormalities have been discussed extensively in the literature, *e.g.*, McLetchie, Purves, and Saunders (1954) and Forshall (1961).

The thoracic and thoraco-abdominal cysts in the present case are more unusual. To explain this type of abnormality in association with abdominal duplication, Saunders (1943) postulated endodermal-ectodermal fusion in the foetus. This theory is summarized by Forshall (1961) and the split notocord syndrome is described by McLetchie *et al.* (1954), Fallon, Gordon, and Lendrum (1954), and Bentley and Smith (1960). The frequency of cervical or thoracic vertebral abnormalities is explained and emphasized. In the present case the thoracic cyst was firmly attached to the right lateral aspect of the body of the sixth dorsal vertebra, but no spinal abnormality similar to those seen by others could be demonstrated. Le Roux (1962) included in his series five patients with posterior mediastinal cysts, three of whom had associated vertebral abnormalities. In the remaining two, only vertebral attachment of the cyst could be demonstrated. One of these two cysts was lined by chronic inflammatory tissue and the other by gastric mucosa. The rest were lined by either ciliated columnar or stratified squamous epithelium. Morrison (1958) described six patients with

posterior mediastinal enterogenous cysts, two of whom had no vertebral abnormalities. Three of the six had cysts with mucosal linings resembling gastric mucosa and one of these had associated vertebral abnormality. Neuhauser, Harris, and Berrett (1958) document four people with posterior mediastinal cysts and associated vertebral abnormalities. Only one had a cyst lined with gastric mucosa. Forshall (1961) included in her series 10 examples of posterior mediastinal cysts. One had no vertebral abnormality, and in a second there was no record of spinal abnormality. The lining of both these cysts was enteric in nature. Six of the other cases had elements of gastric mucosa in the cyst linings.

The thoraco-abdominal form of the abnormality is less well documented. Elwood (1959) described one patient who had a thoraco-abdominal cyst and one with a thoracic cyst. In both instances upper vertebral abnormalities were demonstrated. The lining of the thoracic cyst resembled gastric mucosa but that of the thoraco-abdominal cyst resembled jejunum. Forshall (1961) included three examples of thoraco-abdominal duplications. In two, no spinal abnormality was demonstrated. In one of these, the cyst was shown to have a firm attachment to a dorsal vertebral body. In the other there was no vertebral attachment but the lower, abdominal end had a wide muscular attachment to the stomach. Separation presented

no difficulty. The third case had a gross vertebral abnormality and also a communication with the jejunum at the lower end of the duplication. The cyst linings in these instances were either stratified squamous, duodenal or small intestinal epithelium. Gross, Neuhauser, and Longino (1950) presented three patients with giant diverticula of some part of the small intestine extending through the diaphragm forming a thoraco-abdominal mass. All had spinal abnormalities. Van der Hal and Mooy (1950) and McLetchie *et al.* (1954) both described one similar example. All these duplications, with one exception, were lined with gastric mucosa.

No record can be found in the literature of a case comparable with the one presented in this paper, in which a posterior mediastinal enteric cyst coexisted with a thoraco-abdominal enteric cyst. Neither could a record be found in the literature of a comparable thoraco-abdominal cyst in relation to its position and attachments. In our case the only fixation was to the diaphragm, and there were no visceral attachments. Also the position of the abdominal portion of the cyst was retroperitoneal, in close relation to the right side of the vertebral column, and extending to the level of the third lumbar vertebra, causing distortion of the right kidney which lay anterior to the cyst.

Smith (1960) proposed a classification and nomenclature of accessory enteric formations. He suggested the division of these formations into three categories, namely duplications, vitelline remnants, and dorsal enteric remnants. Our case possesses congenital abnormalities of all three categories, that is, ileal duplication, Meckel's diverticulum, and extensive dorsal enteric remnants in the form of thoracic and thoraco-abdominal cysts.

SUMMARY

A case of abdominal duplication with a Meckel's diverticulum and associated posterior mediastinal and thoraco-abdominal cysts is described. The literature of posterior mediastinal enteric cysts and thoraco-abdominal duplications is briefly reviewed and the possible theories of origin are mentioned.

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