Anatomical course of an oesophago-gastro-duodeno-jejunal duplication

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An electrician's mate aged 18 years was referred to the writer by a chest physician in June 1964 with the history that on mass radiography in July 1962 shadows were seen projecting a little from the right side of the mediastinum both above and below the level of the hilum (Fig. 1).

He had had no symptoms apart from a trivial cough since the age of 6 years. There were a few

slightly pigmented spots on the trunk diagnosed as 'oil folliculitis' due to his occupation.

He was admitted for investigation to a thoracic unit where a thoracotomy was performed by another surgeon. A large multi-cystic oesophageal duplication was found. This was mobilized, but since the proximal end disappeared into the neck and the distal end extended down into the abdo-

FIG. 1. *The duplication projects from the right side of the mediastinum.*

FIG. 2. Associated congenital abnormalities are commonly found with duplications. In this case hemi-vertebrae in the lower cervical spine were present.

men, and since the patient was symptom free, its removal was abandoned.

The physician who had originally referred him for surgery felt unhappy that matters should have been left alone because of the possibility of later complications by ulceration, bleeding, obstruction or perforation. He sought another opinion and referred him to the writer at the London Chest Hospital.

There were no additional pre-operative findings except that radiographs of the spine revealed the presence of congenital hemi-vertebrae in the lower cervical spine (Fig. 2).

A myelogram was not done.

OPERATION

With the patient in the semi-left-lateral position, the right side of the chest was re-opened through the eighth interspace. Dense pulmonary and mediastinal adhesions following the first thoracotomy were freed. With a tube in the normal oesophagus, the duplication lying parallel to the latter on the right side was freed from the neck to the diaphragm. It exhibited three sacculations and was completely covered by vigorously peristalting, well developed plain muscle.

An additional opening was made through the fifth interspace. The topmost sacculation consisted of a fibrotic cyst, 1 in. in length, attached to the cricoid cartilage from which it was freed. It did not communicate with the pharynx nor with the second cyst. This was 2 in. in diameter and in length and extended from

the neck of the first rib to join with the third and largest cyst by a fibrous neck behind the azygos vein.

The third cyst was 4 in. in diameter and could be shown to communicate with a cystic extension within the abdomen. Unlike the other cysts, it could be reduced in size on compression. A narrow tubular extension from it could be seen entering the normal oesophageal hiatus. The two upper cysts received their arterial blood supply from the neck and hilum respectively, but the lower cyst was supplied by a large artery arising in the abdomen and lying between the normal oesophagus and the duplication and supplying both. Figure 3 illustrates how this artery arose from the abdominal aorta immediately above the origin of the coeliac axis artery. It created a hazard in the subsequent part of the operation.

The abdomen was then opened through a right paramedian incision extended to divide the costal margin. The normal oesophagus was exposed and displaced forwards and to the left. This was not easy as it was tethered by the large artery referred to.

The now thin tubular extension of the saccular thoracic duplication was seen to be firmly adherent posteriorly and also to be anchored by the artery. It was expected that the duplication would emerge like the oesophagus between the crura into the abdomen, but this was not the case. Instead, after withdrawing the freed thoracic part through the hiatus, it was seen to disappear behind the right limb of the right crus, still fixed firmly posteriorly in a sessile manner.

At this stage it was found that, 4 in. distal to the beginning of the jejunum, there was a large cystic duplication of the latter which did not communicate with the jejunal lumen. Pressure on this distended the lower thoracic cyst, and there was no doubt that they shared a common cavity, although an immediate search did not reveal the course taken by the tract.

The abdomino-thoracic artery was ligated and divided on the aorta.

The right crus was cut across and the duplication was freed at this point, but it continued as a loop to the right, passing behind the upper border of the head of the pancreas and the portal vein, describing an arc parallel with the first, second, and third parts of the duodenum. Opposite the second part of the duodenum it bulged into another sacculus 2 in. long, and throughout its course it was grossly adherent posteriorly. These facts were only ascertained after mobilizing the duodenum and head of the pancreas from the right and reflecting these structures to the left.

The duplication was threaded out to the right of the duodenum, but it extended once more to the left behind the superior mesenteric artery and vein to enter the root of the mesentery immediately to the right of the beginning of the jejunum, where it was represented as a tube $\frac{1}{4}$ in. in diameter.

By making a tunnel, the whole structure was brought out below the transverse meso-colon.

By this time the operation had taken six hours.

FIG. 3. Course of the oesophago-gastro-duodeno-jejunal duplication from cricoid to jejunum. Points of special interest are (a) its passage behind the right limb of the diaphragmatic crus; (b) its further passage, closely adherent, behind the portal vein and pancreas within the normal duodenal loop; (c) the blind ends; (d) the arterial supply from the aorta to the central section was shared with the corresponding length of oesophagus.

The final distal jejunal part of the duplication was 5 in. in length, appearing as a large saccular cyst sharing a common wall with the jejunum, being separated only by two layers of mucosa. In these circumstances and because of the length of time the boy had been on the table, the long specimen was cut off with part of the final cyst, and the hole in the latter was sutured (Fig. 4). The anti-mesenteric border of the jejunum was opened, and the greater part of the mucosal barrier was removed to provide free drainage, instead of resecting the length of jejunum. It was noted that the distal cyst received its blood supply from the superior mesenteric artery. There were no pancreatic or hepatic elements attached to the duodenal part of the anomalous gut.

The only other abdominal abnormality was a malrotation of the colon and a narrow mesenteric base. The whole small bowel was rather thick-walled and was in a state of chronic volvulus, having undergone a rotation through 180 degrees. This was corrected but

could not be fixed. The chest and abdomen were closed.

Ten days later the patient developed subacute intestinal obstruction with fluid levels. The abdomen had to be re-opened. The volvulus had not recurred, but an obstructing band was found attached to the colon. This was removed and the patient has remained well and symptom-free since.

MICROSCOPY

UPPERMOST CERVICAL CYST There is irregularly arranged muscle and fibrous tissue. No epithelial lining remains (Fig. 5).

FIG. 4. The resected specimen. The lowermost sacculus of the duplication (dotted line) was not resected but was widely opened into the jejunum.

FIG. 5. Microscopy of upper cyst. There is irregularly arranged muscle and fibrous tissue. No epithelial lining remains.

MIDDLE CYST (upper thoracic) The tube is lined by low columnar epithelium with a few goblet cells. There are mucous glands and a few lymphoid follicles in the lamina propria. The appearances suggest intestinal metaplasia (Fig. 6).

FIG. 6. Microscopy of middle cyst. The tube is lined by low columnar epithelium with a few goblet cells. There are mucous glands and a few lymphoid follicles in the lamina propria. The appearance suggests intestinal metaplasia.

FIG. 7. Microscopy of lower cyst. There is cuboidal or flattened epithelium, and glands and lymph follicles as in the middle cyst. Well-developed columnar epithelium and a few glands are seen.

LOWER CYST (thoracic part) There is cuboidal or flattened epithelium, and glands and lymph follicles as in the middle cyst (abdominal part). Well developed columnar epithelium and a few glands are seen (Fig. 7).

COMMENT

This appears to be the first record of anatomical details of the abdominal part of a thoraco-abdominal duplication.

The mechanism of formation of duplications is still perhaps in doubt, but Bremer's theory (1944), inspired by Johnson (1910), is at present accepted as sound in foundation.

Oesophago-abdominal duplications have been described by Butler and Ende (1950), Gross, Holcomb, and Farber (1952), Gross (1953), Ansell and Edwards (1958), and Borrie (1961), but a search of the literature has not revealed any description

of the course taken by the anomalous gut below the diaphragm.

Of the recorded lesions of this type, all appear to have had distal communication with the stomach, duodenum or jejunum.

In only one other case is there a record of the successful removal of the abdominal as well as the thoracic part, and this was reported by Gross (1953) in a child aged 4 months. No details of course or of blood supply are mentioned.

Gross also succeeded in removing the thoracic parts in two other children aged 4½ years and 3½ months respectively, but was saved the necessity of abdominal exploration because there was the safety factor of gut communication at the distal end in each case.

Borrie met with serious complications in resecting only the thoracic portion in spite of a distal opening into the duodenum.

It is clear that where there is no distal communi-

cation, resection of the thoracic portion alone can lead to fistula and other difficulties; but where the abdominal portion of the duplication can be found (and this may be a difficult task), and is not removed, it should be anastomosed to the intestinal tract.

With reference to mucosae, functional duplications, such as the double oesophagus described by Ansell and Edwards (1958), appear to be lined by mucosa identical with that of the normal companion. Where the duplication is functionless and yet has a distal communication, the mucosal pattern merely resembles that of the gut into which it opens, but is often more rudimentary and degenerate. Where there is no communication, the mucosal lining may resemble that of any part of the intestinal tract or even exhibit ciliated epithelium (Barlow, 1957).

Degenerative changes in cyst linings are common. In gastric-lined cysts, ulceration is prone to occur, and this may result in haemorrhage or perforation. Because of the lymphoid tissue in some cyst walls they may become infected.

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