PERFORATED PEPTIC ULCER IN A MONGOLOID INFANT ASSOCIATED WITH CONGENITAL DIAPHRAGMATIC HERNIA

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Peptic ulcers are not common in childhood. According to Hutchins, quoted by Margolis, Valdes-Dapena, and Boles (1949) only 243 cases had been recorded in the literature up to 1944, and 77 of these had perforated. Margolis described perforation of a duodenal ulcer in an infant only 1 month old. In a further search of the literature during the past 10 years we have traced 13 reports of peptic ulcers occurring in children below the age of 7 years; seven were situated in the duodenum and, of these, four had perforated; the remainder were gastric and all had perforated.

Lee and Wells (1923) describe an example of hour-glass stomach in a 13-day-old infant suffering from pyloric obstruction. At operation a chronic ulcer in which a previous perforation had been sealed, was found and later confirmed by necropsy and histological section. The authors, who reported their case under the title *Perforation in utero of a Gastric Ulcer*, considered that the ulcer had been present from before birth, and symptoms and signs became manifest after taking food. Strayer (1949) described a baby, 24 hours old, with 'coffee ground' vomiting, in whom immediate operation revealed perforation of a duodenal ulcer. This patient died on the seventh day, and at necropsy bilateral hydronephrosis was also found.

Margolis points out that peptic ulceration in infants is usually acute, terminating suddenly with haemorrhage or perforation. Premonitory symptoms, such as anorexia, abdominal pain, haematemesis, or melaena may be present over a period of weeks or months. Pyloric obstruction is seen occasionally and a mistaken diagnosis of congenital hypertrophic pyloric stenosis may be made.

Various aetiological factors have been incriminated from time to time. It has been suggested

that asphyxia, resulting from prolonged and difficult labour, may produce a circulatory disturbance with devitalization and consequent ulceration of the gastric mucosa; and the absence of inflammatory reaction is thought to favour this view. Gastric irritants, infection, and malnutrition have all been invoked as possible causative factors, and Berglund (1929) reported six cases of meningitis associated with peptic ulcer. Finally, clinical and experimental observations have long associated peptic ulceration with lesions of the hypothalamus. Mossberger (1947) described a minute tumour which he found immediately anterior to the mamillary bodies in a 5-day-old infant which died of perforated duodenal ulcer. The tumour arose, apparently, from the floor of the fourth ventricle and in structure it resembled pineal gland. The acute ulcer associated with severe burns in children is well recognized, and has been ascribed either to the initial shock or to absorption of toxins from the infected area of skin.

In the case now presented there did not appear to be any particular aetiological factor to account for the ulceration and perforation of the duodenum. No lesion was found in the central nervous system, and the fact that the patient was a mongoloid defective is unlikely to have had any bearing on the problem. He was known to have a severe and overwhelming respiratory infection and the diaphragmatic hernia was probably an incidental finding.

Case History

B.D., a boy aged 4 years and 4 months, was the thirteenth in a family of 14 children who were all alive and well, the eldest being 28 and the youngest 3 years of age. Birth was at full term, the delivery was normal, and the birth weight 8 lb. 12 oz.; he was breast-fed for the first two months and given dried milk feeds with adequate additional vitamins subsequently. Progress

was slow: he sat up at 12 months, stood at 18 months, walked at 2 years, and was able to feed himself at 2½ years. His vocabulary was limited to single words.

Past Illnesses. During early infancy he suffered from repeated attacks of bronchitis and conjunctivitis and was admitted to hospital for short periods on three separate occasions with burns of the left arm, cellulitis of the left leg, and suspected poisoning.

Present Illness. On the present occasion he was first admitted to another children's hospital on September 7, 1950, with a two days' history of diarrhoea and vomiting, and was transferred on September 9, 1950, to an isolation cubicle at Alder Hey Children's Hospital.

Clinical Examination. On admission he was an undernourished child, weighing 23½ lb., and he had the typical appearance of mongoloid deficiency. The temperature was 101° F., pulse rate 140 per minute, respiration rate 36 per minute, and there were signs of extensive bronchopneumonia in both lungs. The heart sounds were obscured, and no abnormalities were found on examination of the ears, throat, abdomen, or central nervous system.

Progress. Treatment was at once begun with sulphonamides and penicillin. Oral feeding was given for 24 hours, but vomiting occurred and necessitated intravenous fluid replacement with an infusion of half-strength plasma with Hartmann's solution, followed by Darrow's solution, half-strength saline, and 5% dextrose—a total of 2,500 ml. in 36 hours. Oral feeding was resumed with difficulty and supplemented with oesophageal tube feeds. The stools were relaxed but not more than two or three were passed each day. No pathogenic organisms were isolated from the faeces.

On September 13, 1950, at 12.10 a.m. about 5 oz. of bright red blood was passed by rectum. At 6.45 a.m. he again passed a small quantity of blood. Later in the day the stools were relaxed and almost black, suggesting that bleeding was still occurring in the upper part of the alimentary tract. On examination the abdomen was moderately distended but there was no muscle guarding and no palpable tumour. During the examination of the chest it was thought that bowel sounds could be heard in the precordial region, but the patient's extreme restlessness and numerous moist sounds made it impossible to be certain of this finding. The following diagnostic possibilities were considered.

GASTRO-ENTERITIS. This was the diagnosis under which the patient had been sent into hospital. The stools, although relaxed, had never been offensive. No pathogenic organisms had been isolated, and the bright red blood passed per rectum was not mixed with the faeces.

BLEEDING MECKEL'S DIVERTICULUM. The bleeding from such a lesion is usually profuse: the fact that the blood was bright red at first was in favour of this diagnosis.

INTUSSUSCEPTION. Against this was the absence of a palpable tumour and of mucus in the stool or on the glove finger when a rectal examination was performed.

DIAPHRAGMATIC HERNIA. Ulceration may occur from pressure of the sharp edge of a hernial orifice on some portion of the alimentary tract. Oesophageal ulceration

is a common complication of certain types of diaphragmatic herniae.

The patient's pulmonary condition was so serious that it clearly overshadowed the melaena and precluded further investigation at that time. The temperature ranged between 98° and 102° F. and the respiratory rate between 30 and 40 per minute. An antero-posterior

FIG. 1a.—Radiograph showing diaphragmatic hernia and gas shadow under the right side of the diaphragm.

FIG. 1b.—Diagram of Fig. 1a.

radiograph of the chest and abdomen was taken on September 13, 1950, but lateral views and radiographs after a barium meal were postponed until his general condition would justify these procedures. The chest radiograph confirmed the diagnosis of a diaphragmatic hernia and showed the presence of a small amount of air

between the right lobe of the liver and the dome of the diaphragm (Figs. 1a and 1b). The appearance of air above the liver strongly suggested the presence of a perforated hollow viscus, but there was no clinical evidence of a perforation. The abdomen was always relaxed and there was no local tenderness.

On September 14, 1950, his condition was further complicated by the development of whooping cough, with which disease he had been in close contact shortly before admission. The physical signs in the chest became more extensive and his condition deteriorated rapidly. The child died on September 26, 1950. A necropsy examination was carried out by Dr. E. G. Hall.

Necropsy Findings. The body was that of a young boy showing the typical features of mongolism. The heart was small but there were no anatomical abnormalities. A little yellow fluid, from which was cultured coagulase positive *Staphylococcus aureus*, was present in both pleural cavities. The lungs showed bilateral basal bronchopneumonia. The lower half of the oesophagus was dilated and the mucosa necrotic. A diaphragmatic hernia was present through a defect in the anterior leaf of the left half of the diaphragm. The hernial sac, approximately 3 in. in diameter and depth, lay immediately behind the sternum and anterior to the heart; its apex was slightly to the left of the mid-line and reached the level of the manubrio-sternal junction. It contained the middle 9 in. of the transverse colon and corresponding portion of a long mesocolon, but no other viscera. The stomach was normal. On the antero-superior surface of the duodenum there was an acute ulcer, 2 mm. in diameter, which had perforated the wall; the rest of the duodenum was normal. The small intestine was rather short and its mesentery oedematous and fibrotic. There was no evidence of general peritonitis but light, filmy adhesions between the duodenum and the under surface of the liver and a few flecks of inspissated pus over the antero-superior surface of the duodenum were seen. The liver was rotated to the right and the left lobe was smaller than normal. No abnormalities were found in the pancreas, spleen, thymus, kidneys, adrenals, or central nervous system.

HISTOLOGICAL SECTION OF THE DUODENUM. The peritoneal epithelium was destroyed around the ulcer and replaced by a relatively acellular, loose, fibrinous granulation tissue containing degenerated polymorphs. Extending into this tissue from the oedematous subserosa were newly-formed capillaries. The subserosa was infiltrated with lymphocytes, plasma cells, and some eosinophils. The muscle layer at the edge of the ulcer tailed into material similar to that on the serous surface; internally there was a well-defined granulation tissue zone.

Comment

This patient, aged 4 years and 4 months, presenting the typical appearance of a mongoloid defective, was admitted to hospital on account of diarrhoea and vomiting and died after an illness lasting 21 days. The primary cause of death was bronchopneumonia, recognized clinically and confirmed at necropsy. A diagnosis of diaphragmatic hernia was

made and, on radiological grounds, perforation of a hollow viscus was suspected; this proved to be due to a perforated acute duodenal erosion. It is interesting to consider how these various findings could be related.

It is obvious that in a patient suffering from bronchopneumonia a large diaphragmatic hernia must diminish considerably the chances of recovery. In fact, a diaphragmatic hernia may attract attention to itself on account of respiratory distress. The additional strain of the paroxysms of whooping cough, together with bronchopneumonia and the associated hernia, are sufficient to account for the unfavourable progress and outcome. The initial symptoms of diarrhoea and vomiting, the subsequent passage of blood by rectum, and the unexpected finding of a perforated duodenal ulcer are less easy to fit into the picture.

The problem can be approached from two angles: the incidence of peptic ulceration complicating diaphragmatic hernia; and the incidence of peptic ulceration in childhood.

Although there is evidence that gastric ulceration can be associated with a diaphragmatic hernia, there seems to be no known association between herniation and duodenal ulcers. Dunhill (1948), in 52 consecutive cases of diaphragmatic hernia at all ages, states that in seven cases (14%) bleeding, caused by venous congestion in the gastric mucosa, occurred. In only one case was a chronic gastric ulcer found. In a further 30 cases described by Harrington (1945) eight had haemorrhage from the stomach and, at operation, seven of these were, in the author's opinion, due to trauma at the edge of the diaphragmatic hernia.

Perforation of a gastric ulcer into a diaphragmatic hernial sac has been described by Lennox Gordon (1916). The patient, aged 56 years, gave a four-year history of indigestion and had been unfit for work for six months. He had had abdominal pain, vomiting, and constipation for four days before admission to hospital. Liver dullness was absent and the right chest was hyper-resonant. Death occurred before operation, and at necropsy a defect in the right dome of the diaphragm with a diaphragmatic hernia, containing the pyloric half of the stomach, the first and second parts of the duodenum, the right half of the transverse colon, and some coils of small intestine, was found. A perforation of the anterior wall of the stomach 1 in. from the pylorus had occurred, and this perforation had all the characteristics of an ulcer and seemed to have been of recent origin. Sangster (1936) reports the post-mortem findings in the case of a man aged 64 years, who had a defect of the left dome of the diaphragm. The hernia contained

the left lobe of the liver, a portion of the transverse colon, and loops of the jejunum in addition to the stomach, which was rotated so that the greater was higher than the lesser curvature. A perforation of the posterior wall of the stomach had occurred into the left pleural cavity.

There is no evidence of any association between the duodenal ulcer and the diaphragmatic hernia in the case now reported. The ulcer was of the acute variety and there is no reason to incriminate the diaphragmatic hernia which had presumably been present since birth. The hernia, moreover, was a ventral one through the left hiatus of Morgagni and contained only a portion of the transverse colon. The neck of the hernia was wide and there had been no strangulation; nor was there any evidence to suggest that there had ever been any other contents. Peptic ulceration, when it occurs in association with a diaphragmatic hernia, is usually gastric in site and of chronic type, and it arises when part of the stomach is itself within the hernial sac.

Summary

A clinical case of a typical mongoloid defective, aged 4 years and 4 months, is presented. He was

admitted to hospital on account of diarrhoea and vomiting. A diaphragmatic hernia was discovered. Death occurred from bronchopneumonia complicated by whooping cough. Necropsy demonstrated perforation of a duodenal ulcer. The relationship between peptic ulceration and diaphragmatic hernia is considered and the incidence of peptic ulceration in infancy briefly discussed.

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