SYMPTOMATIC PANCREATIC HETEROTOPIA OF THE PYLORUS ASSOCIATED WITH BILATERAL RENAL CORTICAL NECROSIS IN AN INFANT

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(RECEIVED FOR PUBLICATION MARCH 27, 1956)

It has been suggested that islets of heterotopic pancreatic tissue are sometimes of clinical importance in adults, and that, occurring in the stomach wall, they may produce symptoms of pyloric spasm (Faust and Mudgett, 1940; Krieg, 1941; Waugh and Harding, 1946). They have been noted occasionally in infancy, but symptoms have not been attributed to them (Branch and Gross, 1935; Craig, 1955). Their importance will obviously vary with their size and position, and probably, with the intrinsic irritability of the neuromuscular apparatus of the gut. Even at necropsy the condition could easily be overlooked, and the present case in a 14-weeks-old boy suggests that it should be considered in the search for an organic basis for persistent vomiting in early infancy.

A heterotopia occurs when tissue elements proper to one area of the body develop at some other site where they are not normally found. Heterotopias are relatively common, especially throughout the intestinal tract. Thus gastric glands may be found in the oesophagus, and either gastric glands or pancreatic tissue, or both, are often found in congenital diverticula and in enterogenous cysts. Heterotopias may show disproportion in the relative proportion of their constituent tissues, and this is especially so in pancreatic heterotopias, where duct elements often greatly predominate over acinar tissue. However, they can usually be separated easily from hamartomata. In these some tissue constituents, such as vascular channels, nerve tissue, bone or cartilage, though normal for the area, are excessive, are disproportionate to other elements, and form respectively angiomatos tumours, neurofibromatous nodules and osteochondromata and other defects of bone modelling.

Clinical History

This full-term boy weighed 7 lb. 8 oz. (3,400 g.) at birth. Delivery was normal and one previous child is alive and well aged 3 years.

He was admitted to hospital on the seventeenth day of life with projectile vomiting, which started on the tenth day, and with loss of weight. There was visible peristalsis and a palpable tumour in the epigastrum and four-hour retention of a barium meal. Three days later operation revealed a hard firm tumour at the pylorus described as typical of hypertrophic pyloric stenosis and Rammstedt's operation was done. He was discharged seven days later after an uneventful recovery.

Three days later he vomited again with green diarrhoea and was in hospital for two days. Ten days later at the age of six weeks, he was again admitted for vomiting and diarrhoea. No intestinal pathogens were isolated, vomiting ceased after 48 hours and he gained 9 oz. (250 g.) in 12 days in hospital.

At the age of 10 weeks he weighed 11 lb. (5,000 g.) but at 12 weeks he was readmitted for diarrhoea. Intestinal pathogens were not isolated, and three days later vomiting developed and he rapidly became dehydrated. Saline infusions were given. The vomitus was coffee ground and on the fifth and sixth days he passed tarry stools. The haemoglobin was only 55% and he was transfused with 200 ml. blood but without clinical improvement. Despite treatment of a stomatitis he became reluctant to feed, most of his feeds were vomited and the vomitus continued to contain altered blood. He died two weeks after admission aged 14 weeks.

Necropsy

The body was that of an emaciated male infant. Externally there was no congenital defect, no abnormal pigmentation and no oedema. The surgical incision was healed.

The thymus is atrophic, the lungs are slightly collapsed, reddish, and, though oedematous posteriorly, they are not consolidated. The great vessels enter and leave the heart normally and there is no abnormality of the valves or cardiac septa. There is no hiatus hernia and the position of the cardio-oesophageal junction in relation to the intact diaphragm is normal.

There are no abnormal peritoneal adhesions. The stomach is empty and is not dilated. The pyloric region is slightly hypertrophied, but the muscle is relaxed and soft and the canal admits a probe normally. Only the
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posterior quadrant shows distinctive thickening of the muscle wall extending proximally for 1 cm. from the pylorus. There is patchy congestion of the mucosa of the lesser curve near the cardia, but elsewhere it is pale and no ulcers or erosions can be found.

Apart from the kidneys the other organs show only some generalized atrophy. The kidneys are not recognizably enlarged, but externally and, even through their capsules, they appear unduly congested and dark in colour, with irregular areas of reddish-yellow mottling. On section the cortex shows a pale yellowish zone which occupies the outer one-third to one-half of the cortex. This is discontinuous and of irregular outline, but about two-thirds of the whole subcapsular zone is involved. A thin red line separates it from the capsule. It is bounded by a narrow irregular zone of deeply congested kidney tissue and, though firm, appears entirely structureless. The remainder of the kidney is congested, but otherwise the inner part of the cortex and the whole of the medulla show their normal pattern and the veins of the boundary zone and the radicles of the renal veins are free of thrombi. The pelvis and ureters are normal.

The meninges are unthickened and free of haemosiderin. The brain externally, and on section after fixation, is normal.

Histological Examination

Routine sections have been prepared from all organs and multiple sections from all four quadrants of the pylorus and from the kidneys. The routine sections show only terminal congestion and oedema of the lungs. The sections from the posterior quadrant of the pylorus show pancreatic tissue in the submucosa and in the circular and longitudinal muscle layers. Pancreatic acini and a grossly disproportionate number of pancreatic ducts are present. The acini appear inactive and the ducts are dilated and have a prominent mantle of somewhat immature connective tissue around them. Islet tissue is not present. Multiple sections show no ulceration of the overlying gastric mucosa. Numerous pancreatic ducts traverse the submucosa and must open separately into the stomach.

In the kidney there is widespread and gross congestion affecting both glomeruli and small inter-tubular capillaries. This is especially marked around the necrotic areas and extends into them. The necrosis extends to the renal capsule though in some tubules immediately deep to the capsule isolated groups of cells survive. Towards the periphery of the necrotic tissue shrunk nuclei persist, the red blood cells retain their haemoglobin and there are some polymorphs. In relatively small areas towards the centres of the necrotic areas only the outline of shrunk tissue is recognizable. Tubules lying in the cortex deep to the necrotic zone and in the medulla show little abnormality, though collecting tubules often contain haemoglobiniferous casts. The blood vessels within the necrotic areas show necrosis of their wall and fibrin and breaking-down blood cells in their lumen, but outside the areas of necrosis the blood vessels are normal and there are no ante-mortem thrombi.

Discussion

It is not possible to determine whether this was a case of pyloric pancreatic heterotopia and congenital pyloric stenosis, or whether the apparent thickening of the pylorus seen at operation was due only to a strong reflex spasm of its musculature secondary to the heterotopic tissue. The division of some muscle fibres probably reduced, or abolished for a time the reflex spasm. When pancreatic heterotopias have caused symptoms they have rarely, if ever, done so directly by virtue of their size, but only by reflex disturbance of intestinal movement. Other factors, such as minor intestinal upsets, will influence the irritability of the bowel and symptoms may be expected to vary considerably from time to time.

Pancreatic heterotopias in the region of the pylorus have been mentioned occasionally in early infancy as an accidental finding (Branch and Gross, 1933; Craig, 1935). One of us (J.E.M.) has for some years included a section through the pylorus in the routine histology of his own and supervised necropsies on stillbirths, newborn babies and young infants. In this material, comprising over 500 cases, no other heterotopias have been encountered at this site. Heterotopias are congenital malformations present from birth. Routine inspection and one section might readily fail to reveal a small heterotopia. However, from this material there is no reason to suppose that the incidence of heterotopias in this area is higher than the 1 in 400 individuals suggested by Waugh and Harding (1946) from a review of the literature. Again, it may be emphasized that symptoms are probably produced in a few cases only. Heterotopias are commonest in the submucosa, where they probably produce little disturbance of neuromuscular activity. Again factors concerned with the irritability of the bowel are probably involved, since in the recorded cases symptoms apparently developed only in adult life though the condition must have been present from birth.

Blood loss into the intestine, suggested by the tarry stools, probably cannot be attributed directly to the heterotopia. Minute acute erosions in the stomach mucosa, commonest in the fundus and body, are extremely difficult to recognize as antemortem lesions at necropsy. They are non-specific and relatively common. Study of the mucosa over the heterotopia showed it was proper to the stomach and free of ulceration. Even in illness some acid secretion by the stomach is likely to persist as long as any secretion from the heterotopic pancreatic tissue, and it is difficult to appreciate how such a heterotopia could produce gastric ulceration by its enzyme secretion.

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The occurrence of bilateral cortical necrosis of the kidneys is of considerable interest. This condition has been described, and the theories of its origin discussed in detail, in the monograph of Sheehan and Moore (1952). Cortical necrosis may or may not be due to spasm of small blood vessels and the present case contributes nothing new to the discussion of the aetiology of the condition. Comparison with adult cases suggests that the small kidney of the infant is less rigid than that of the adult, and that congestion and even haemorrhage tends to be more marked. The recognition of different zones in the necrotic tissue is even more arbitrary than in the adult. The survey of Sheehan and Moore and the paper by Campbell and Henderson (1949) show that cortical necrosis is rarely described in infancy and childhood. This is true for the fully developed condition, but study of routine sections from cases of gastro-enteritis in infancy suggests that small focal areas of tubular necrosis occur not infrequently. In fresh and relatively well fixed material the focal change in the parenchymatous tissue appears distinctive, but it is difficult in these small and relatively terminal lesions with their scanty or absent interstitial reaction to produce satisfactory evidence that they are, indeed, ante-mortem lesions.

The lesions of cortical necrosis are entirely different from those of renal venous thrombosis (Morison, 1945). The tendency to involve the more immediately subcapsular portion of the cortex instead of the medulla and deeper cortex, and the absence of thrombi in vessels beyond the involved area is distinctive, even apart from the absence of thrombi in the veins of the boundary zone.

Summary
A pancreatic heterotopia involving the circular and longitudinal muscle of the posterior quadrant of the pyloric antrum in a male infant is described. An operation for hypertrophic pyloric stenosis was performed on the 20th day of life. Symptoms returned after 10 days and, until death at the age of 14 weeks, he had frequent episodes of vomiting and diarrhoea.

Influences increasing the intrinsic irritability of the bowel are probably always important as an additional factor, but in some cases of persistent vomiting the primary factor may be reflex spasm from such a lesion.

Circulatory failure developed and bilateral cortical necrosis of the kidney was found. The lesions resulting from this are entirely distinct from those of renal venous thrombosis.

References