A CASE OF CONGENITAL OBLITERATION OF THE BILE DUCTS IN WHICH THERE WAS FIBROSIS OF THE PANCREAS AND OF THE SPLEEN.

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NAMES may cling though their significance may change with advance in knowledge. This is well exemplified by the title "congenital obliteration of the bile ducts." This term is used to describe a variety of conditions in which there is evidence that either at birth or shortly afterwards there is obliteration or complete absence of the bile ducts to a varying extent in different cases. Thus, in one case there may be obliteration of the duodenal end of the common bile duct, in another complete absence of the gall bladder, the cystic, hepatic, and common bile ducts, and

the other hand, Rolleston makes out a strong case for regarding the cirrhosis as the primary change and the obliteration of the ducts as one of the results of the cirrhosis.

The interest of the present case lies in the fact that not only were the bile ducts completely absent, the gall bladder altogether rudimentary in size and structure, and the liver in a state of typical biliary cirrhosis, but the spleen was fibrosed and the pancreas exhibited a cirrhosis in connexion with its ducts very similar to the cirrhosis in the liver, but with this difference—that whereas in the liver the process had gone on to obliteration of the ducts, in the pancreas the process had not advanced so far; the ducts were well formed and patent, and opened in the usual way into the duodenum.

I am not aware of any previous reference to a condition like this in the pancreas having been noted in connexion with these so-called cases of congenital obliteration of the bile ducts, and I regard the association as evidence in support of the view that the obliteration of the ducts is secondary to the cirrhosis, in contradiction to the theory

Fig. 1.—Under surface of liver (natural size). (a) Rudimentary gall bladder, (b) rudimentary cystic duct, (c) rudimentary hepatic duct.

between these extremes every degree of obliteration may occur. There is one feature, however, common to all these cases, namely, the liver is in a condition of cirrhosis, that variety of cirrhosis to which the term "biliary" or "monolobular" has been given, a form of cirrhosis similar to that which results from experimental ligation of the bile duct in dogs.

The view as to the association between the obliterated or absent ducts and the cirrhotic liver has undergone complete transformation. Thus, as Rolleston points out, John Thomson in his monograph, dated 1892, believed that the primary factor was a congenital malformation of the ducts, and that the biliary cirrhosis was secondary to this malformation. Later on, in his article in Clifford Allbutt's *System of Medicine* written in 1897, he regards the process as independent of any congenital malformation, and due to toxic bodies in the bile (like toluyldiamin) causing a descending cholangitis—that is, inflammation spreading from the fine to the main ducts. When, according to Thomson, the disease has gone on far enough to interfere with the free passage of bile from the liver, then biliary cirrhosis begins, as it does in the liver of animals whose common duct has been tied. On

that the cirrhosis results from the obliteration of the ducts.

The *post mortem* appearance of the organs suggests that in intrauterine life, as a result of the circulation of some toxic body, there had been set up a fibrosis of certain organs—namely, the liver, the spleen, and the pancreas (and perhaps of others which, however, had escaped notice, not having been submitted to microscopic examination); that this fibrosis in the liver and pancreas had its seat of election round the hepatic and the pancreatic ducts; and that while in the liver it had progressed to such an extent as to cause obliteration of the ducts, in the pancreas it had not gone on so far, and here the ducts had remained patent. The brunt of the poisoning would be expected to fall upon the liver, for the poison must reach the fetus from the mother via the portal vein. Some of it, however, would pass by the ductus venosus into the general circulation, and of this some would again find its way back to the liver by the hepatic artery, whilst some would reach the spleen by the splenic artery and some the pancreas by the pancreatic artery. The liver, therefore, would receive a double dosage, the bulk by means of the portal vein and a smaller quantity by the hepatic

artery. On the other hand, the spleen and the pancreas would only receive a relatively small quantity through the splenic and pancreatic arteries respectively.

Rolleston and Hayne describe the cirrhosis of the liver in these cases of congenital obliteration of the bile ducts as of a mixed type, partly multilobular, that is—ordinary

The child was born at full term and appeared healthy. It was breast-fed for the first fortnight, but since then had been fed on milk and barley water. There was no jaundice at birth, and normal meconium was passed. On the fourth day jaundice appeared, and at the same time the motions became "white like milk and very frequent," and from this time onwards the child became gradually thinner and more and more

Fig. 2.—Section of pancreas ($\times 360$ diameters). The ducts are well formed, lined by regular columnar-celled epithelium, and lie embedded in dense strands of fibrous tissue.

Fig. 3.—Section of liver ($\times 300$ diameters). The bile ducts everywhere small and in places completely obliterated, lie obliterated in dense strands of fibrous tissue.

portal cirrhosis, the effect of the poison that enters by the portal vein—and partly monolobular or biliary cirrhosis, the effect of the poison that reaches the hepatic lobules by the branches of the hepatic artery.

In the present case we must assume that the poison came into action at a very early period of embryonic life, since in the case of the liver it not only led to cirrhosis and an obliterative cholangitis, but it also determined a faulty formation or rather a complete arrest of development of the large bile ducts outside the liver as well as a very faulty development of the gall bladder itself. In contrast to this condition of the hepatic ducts were the pancreatic ducts, for embedded in fibrous tissue as they were inside the pancreas, externally they opened on to well-formed papillae in the duodenum and apparently functionated during life satisfactorily. This contrast between the bile and the pancreatic ducts is well explained by the liver receiving a very much larger amount of the poison than the pancreas, as pointed out above.

The particulars of the case are briefly as follows:

A female infant, 10 weeks old, was admitted into the Hospital for Sick Children, Birmingham, under the care of Dr. G. W. Powell, on February 28th, 1905, for jaundice and diarrhoea. Both of these symptoms appeared on the fourth day of life, and had persisted ever since. The general development of the infant was good, but it was wasted, weighing only 9 lb., and it was deeply jaundiced. The anterior fontanelle was depressed. There were no external signs of congenital syphilis. The heart and the lungs were normal. The liver was palpable 1 in. below the costal margin in the right vertical nipple line, and the free edge was sharp and hard. The spleen was firm and palpable two fingerbreadths below the costal margin.

yellow. The father was healthy, but the mother did not look very strong, and said that she had always been somewhat delicate. She lost the sight of the left eye in infancy, and had had the eye excised when she was 8 years old. She had rheumatic fever at 14 years of age, and a second attack three years later. She was an only child, and was left an orphan at so early an age as to have no knowledge at all of her own parents. She had had eight pregnancies. Her two eldest children were alive and healthy, aged 7 and 6 years respectively. The third

child died at 6 months of age of "wasting disease," the fourth at 8 months of age of "convulsions," and the fifth at 8 months of age of "broncho-pneumonia." These three children were all healthy at birth. The sixth pregnancy ended in a miscarriage at six months, and the infant the subject of this article was born next at full term, and since its birth there had been another miscarriage at two months. The mother showed no signs of syphilis, congenital or acquired, but her parentage and appearance suggested descent from a poor stock, and her child-bearing history is certainly suggestive, at all events, of some chronic toxæmic condition during the later years of her married life. This toxæmia was possibly (some would say probably) due to syphilis, but no further evidence in this direction was obtainable.

The mother was not aware of any similar case of infantile jaundice having occurred in any other member of her family. There is little more to record. The child, after its admission, gradually be-

came weaker, and died from asthenia six weeks later—that is, four months after birth. The jaundice persisted to the end. On March 17th some urine drawn off by the catheter was found to contain bile pigment. There were two or three motions every day, and they were "white, like thick milk." The temperature was subnormal during the entire six weeks the child was under observation. At no time were there any hæmorrhages. The weight at death was 8½ lb. The child was treated with mercurial inunctions, but without the least improvement at any time.

Fig. 4.—Section of spleen ($\times 60$ diameters). The trabeculae are represented by thick strands of fibrous tissue.

Post-mortem Examination.—Body well developed, but markedly wasted. No signs of congenital syphilis. No enlargement of glands, no haemorrhages, no oedema. Marked jaundice of entire skin, conjunctivae, and all the internal organs. The pericardium was healthy. The heart weighed $\frac{1}{2}$ oz., and was healthy. The pleurae were healthy. Each lung weighed $2\frac{1}{2}$ oz., and exhibited small areas of collapse along the posterior surface, but otherwise was crepitant throughout. No ascites, no peritonitis, no enlargement of mesenteric glands, but a few enlarged dark-coloured glands at the hilum of the liver. The stomach and intestines presented nothing abnormal. The spleen weighed 1 oz., was firm and enlarged. The kidneys each weighed $\frac{1}{2}$ oz., and with the suprarenals appeared normal. The ureters, bladder, uterus, and appendages were healthy. The pancreas appeared small, somewhat shrivelled, and tough. The liver weighed 7 oz. It was dark olive-green in colour, and its surface was roughly granular, like morocco leather. To section it was very tough, and obviously markedly cirrhotic. The gastro-hepatic omentum was dissected with the greatest care, and the hepatic artery and the portal vein were traced to both lobes of the liver, but no signs of a common bile duct could be found, not even a fibrous band passing from the gall bladder to the duodenum. The ductus venosus was traceable, connecting the portal vein and the round ligament. In the fissure of the gall bladder there lay a whitish wrinkled tube $\frac{1}{8}$ in. in diameter and $\frac{1}{2}$ in. in length. Its free end (the fundus) was somewhat bicornuate, and did not reach the anterior margin of the liver by $\frac{1}{2}$ in. The opposite end was continuous with a dark-coloured ovoid swelling which lay in the transverse fissure. On splitting longitudinally the narrow whitish tube it was found to be divisible into two nearly equal lengths. The anterior half was smooth lined, the posterior half showed spiral foldings of the mucous membrane, and the suggestion forced upon one was that the former represented a rudimentary gall bladder, and the latter the cystic duct. The wall of this tube throughout its length was thick, and it contained clear, yellow-tinged, thin fluid. The dark-coloured ovoid swelling above mentioned lay in the transverse fissure, and appeared to be continuous with the whitish tube. It was $\frac{1}{2}$ in. long, and $\frac{1}{4}$ in. in width, ovoid in shape, and dark green in colour. Its wall was thin, and it was distended with a viscid green-coloured fluid quite different to the contents of the whitish tube. Its interior was smooth lined and was coated with a green slime. No communication existed between this ovoid swelling and the whitish tube. Its true inferior surface was free, its superior surface inseparably associated with the hepatic tissue in the transverse fissure. Possibly this may have represented a rudimentary hepatic duct which had failed to communicate with the gall bladder.

On opening the duodenum two well-marked papillae (a small and a large one) were present opposite to the head of the pancreas. Bristles could be passed into these for about an eighth of an inch towards the pancreas. They were the openings of the pancreatic duct into the duodenum.

The liver, spleen, and pancreas were sectioned, and examined microscopically. In all these organs the characteristic feature was the large amount of fibrous tissue present. There was, however, a great contrast between the condition of the bile ducts and that of the pancreatic ducts, and this contrast is well brought out by the excellent photomicrographs so kindly taken for me by Dr. John T. Hewetson.

The liver shows fibrous bands of enormous thickness permeating the entire organ. They are entirely interlobular, for nowhere do they extend into the lobules. The striking feature of these fibrous tissue strands is the enormous number of bile ducts they contain cut in every direction, some longitudinally, others transversely. For the most part the lumina of these ducts are conspicuously absent, but here and there they may be seen as spaces filled with inspissated bile readily distinguished by its bright green colour. The cells lining the lumina are low cubical in type, and show well-stained nuclei. There is, however, a great contrast between these cells and the cells corresponding to them in the pancreas. In the pancreas they are columnar in shape, regular in their distribution, and enclose a widely-open lumen, while in the liver they are more or less cubical in shape, more irregularly disposed around a lumen which is everywhere narrow and in many places obliterated.

Sections of the pancreas show healthy acini, well-marked islands of Langerhans, and well-developed ducts. These ducts are lined by columnar epithelium with well-stained nuclei. They are, however, seen to be embedded in conspicuous strands of fibrous tissue which is moderately cellular in structure. The lumina of the ducts are large and patent, and appear quite normal.

In the spleen the trabeculae are represented by dense fibrous strands. The pulp and the Malpighian bodies appear quite normal.

In concluding my account of this case, I wish to acknowledge my thanks to Dr. G. W. Powell for his permission to publish it, to Dr. A. E. R. Weaver for his excellent drawing of the liver, and to Dr. John T. Hewetson for the photomicrographs of the microscopic sections.

The bibliography on this subject will be found at the end of John Thomson's article in Allbutt's *System of*

Medicine, vol. iv, p. 257; and further references will be seen in a paper by Rolleston and Hayne in the *BRITISH MEDICAL JOURNAL*, vol. i, 1901, p. 758; and in Rolleston's *Diseases of the Liver*, p. 639.

THE RINGWORM QUESTION IN ELEMENTARY SCHOOLS.*

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SINCE January this year some 195 cases of ringworm in children have come under my care at the West London Hospital, Hammersmith, and 69 at the Hospital for Diseases of the Skin, Blackfriars, making a total of 264; 89 of these had not recently been attending school, some being under school age, while others had been kept away from school for a considerable time before coming under my notice. In a few cases, no doubt by inadvertence, no note was taken in the hospital casebook in reference to the school attendance.

The schools which the children had attended up to the time of coming to the hospital, or up to a short time before, were noted in 175 cases—125 at the West London Hospital, and the rest at the Hospital for Diseases of the Skin. Each of the hospitals serves a wide area; the cases at Hammersmith come from 49 county council and other elementary schools, and those at Blackfriars from 41. Several of the individual schools have furnished within the period more than one case—for example, Brackenbury Road School no less than 8, St. John's, Hammersmith 7, St. Dunstan's 6, Fulham Palace Road 6, the British School, Brentford, 6, and William Street, Queensmill Road, and Sacred Heart, 5 each; but the majority of the schools in my list are each responsible for only one or two of my cases.

It is, of course, obvious that the cases of ringworm from these ninety schools, which have come under my personal notice within the period of six months, represent but a small proportion of the total number of the school children with ringworm; many of these, no doubt, go to other hospitals and dispensaries, and many of them to the general practitioners of the neighbourhood.

The figures which I am able to bring forward from my individual experience are, however, in themselves enough to show (1) how widely spread the disease still is among the metropolitan school children, and (2) what a large number of elementary schools now are, or have recently been, contaminated by ringworm. I may here state that many of the patients at the time of coming under my care were really bad cases, the disease having existed and remained undetected by the school teachers and nurses for a considerable time.

It may perhaps be asked by those who are satisfied with the existing state of affairs what proof there is that these children acquired the ringworm at the schools. I freely admit that some of them may have contracted it elsewhere, but there is no getting away from the fact that all of them had been attending at the schools, often for weeks or even months, without detection; and every one conversant with the disease and with the usual conditions of school life will admit that they must all of them have been sources of infection for the other children. I have no doubt myself that in the large majority of the cases the mothers were quite right in saying that the ringworm had been "caught at school."

Many years ago, and again in 1905, in a paper read before the Royal Institute of Public Health, I pointed out the inadequacy of the measures adopted by the authorities against the spread of ringworm and other contagious diseases of the skin in the elementary schools, but as far as I can judge from the cases that are still coming under my notice at the two hospitals, there does not seem to have been much improvement since then, in spite of the fact that the Education Department of the London County Council has to a certain extent increased its medical staff. I further venture to say that it is not likely that we shall see much improvement until the medical inspection of school children becomes a reality,

* Read at the International Congress of School Hygiene, London August, 1907.