CASE OF OBLITERATION OF THE PORTAL VEIN (PYLEPHLEBITIS ADHESIVA).

By WILLIAM OSLER, M.D., M.R.C.P. Lond., *Professor of the Institutes of Medicine, McGill University, Montreal.*

THROMBOSIS and suppurative pylephlebitis are the affections most commonly met with in the portal vein. A few instances of calcification and extreme fibroid thickening of the walls are reported. Organic occlusion, by conversion of the vein into a fibrous cord or mass of connective tissue, is a very rare lesion, as in cases of thrombosis life is usually terminated long before organisation of the clot can take place. The following instance of it presents many interesting features, anatomical and clinical:—

J. C., aged 28, admitted into the General Hospital, under my care, June 17th, 1881, in a condition of extreme exhaustion, consequent upon loss of blood by vomiting. My house-physician, Dr Andrew Henderson, obtained from him the following history:—Has always been strong and healthy; somewhat intemperate, but a steady worker. Has never had syphilis. No constitutional disease in his family. Last September, when engaged in some very hard work, was obliged to give up on account of weakness and dull heavy pain in the upper region of the belly. It was never very localised, and was not aggravated by eating. Patient had to be in bed most of the time, and at about the end of a month had an attack of hæmatemesis, vomiting more than two quarts. Did not leave his bed for some weeks; does not remember whether his legs or abdomen were swollen. Did not go to work until about April, when he got employment in a manufactory as fireman. Latterly, he was put to heavy work, piling bags of sugar, and yesterday (16th) he had to give up owing to feelings of great weakness. This morning he vomited a large quantity of blood, partly fluid, but mixed with clots. When admitted in the evening he was in a state of great exhaustion; surface blanched; pulse very small—135; temperature, 100°; respirations, 20. Shortly after getting to bed he

vomited about four ounces of dark altered blood. A peculiar cadaveric odour was noticed in the breath.

June 18th. At the morning visit patient was examined, and the following condition noted:—He is a large, well-made, muscular man; lies on his left side in a drowsy, semi-conscious state, but can be roused. Face and general surface blanched; no distension of abdomen; superficial veins not visible; no œdema of the feet. Examination of *chest*, negative; *abdomen* flattened; skin of marble whiteness; visible epigastric pulsation; on palpation, marked pulsation in umbilical region, forcible, vertical in direction; no tumour to be felt. On auscultation a remarkable double murmur was heard midway between the navel and tip of ensiform cartilage; to be heard also a little to the right of the middle line, but was very feeble to the left. There was not a cardiac murmur. *Liver*—edge could not be felt; area of dulness much diminished; could scarcely be detected in sternal line; was 3 cm. in mammary, and 4 cm. in axillary lines. *Spleen*—not to be felt below costal border; area of dulness increased, 11 cm. in vertical, 13 cm. in transverse, directions. Patient did not complain of pain during the examination. Bowels have not been moved; a large external pile, filled with coagula, was found on the right margin of anus. Urine clear, and normal.

19th. During the night patient vomited a large quantity of bright, liquid blood, soaking the bed and covering the floor in the neighbourhood. He was found in an apparently dying state, but rallied on the administration of stimulants. The examination at the visit did not elicit any new facts; the epigastric murmur was not so distinct.

20th. No further hæmorrhage, debility extreme, and a tendency to delirium. Slight œdema of feet. The cadaveric odour was very perceptible.

On the 21st and 22d patient remained in a state of profound exhaustion, and there were no additional abdominal symptoms. On the 23d there were repeated syncopal attacks, during one of which he died.

Autopsy.—Body well nourished; skin blanched; no distension of abdomen; cutaneous veins not visible; slight œdema of feet.

In abdomen the coils of small intestines were of a very dark

slate colour; peritoneum smooth; colon distended; no exudation; liver and spleen did not appear below the costal border. In thorax a few ounces of serous fluid in each pleura.

Heart was flabby and pale; chambers contained small clots; valves were healthy. Aorta normal.

Lungs pale, œdematous at bases.

Spleen greatly enlarged; weighed 675 grms.; was intimately adherent to the diaphragm and stomach. Capsule was very thick, in places wrinkled, and a firm, semi-cartilaginous plate existed at its convex border. On section organ cut with great resistance, creaking under the knife; the trabeculæ thickened, rough, and in spots gritty. Some of the veins were dilated, and contained thrombi. Near the hilus was a wedge-shaped calcified mass, the size of a walnut. The artery was very tortuous, and at the hilus presented a group of small saccular aneurisms, the size of large peas; the coats thickened, partly calcified, and one of them contained an old thrombus, which had softened in the centre.

Liver small, and closely united to the diaphragm and to the abdominal wall on the right side. It measured 25 cm. in breadth and 16 cm. from front to back. The shape was retained, but the left lobe was almost completely atrophied, only a small thin tonguelet remaining. The surface was smooth, but towards the right border and behind, many fibrous bands passed between the capsule and the diaphragm. The capsule on the under surface was opaque but smooth. On section, tissue uniform, pale reddish-brown colour; acini distinct, but no perceptible increase in the inter-lobular tissue. The anterior border and the remnant of the left lobe were firmer, and the connective tissue strands between the lobules could be seen. The hepatic veins were of full size. Portal canals not numerous, small; artery and duct distinct (condition of portal vein will be described under venous system).

Microscopic examination showed the liver cells to be somewhat fatty; the connective tissue on the greater portion of the right lobe was not specially increased, but at the anterior border and in the small portion of the left lobe the secreting substance was a good deal atrophied.

Gall-bladder contained a quantity of yellow bile. Gall duct normal. Hepatic artery almost double the usual size.

Stomach was capacious and contained a quantity of dark liquid mixed with food. Veins beneath the muscular coat could be seen dilated and tortuous. The mucosa was pale, here and there marked with spots of capillary injection. There were two small superficial losses of substance near the cardiac end; the tissue about them was not injected, and their bases were scarcely as deep as the submucosa. About the pylorus the membrane was mammillated, and on section very tough.

Intestines contained dark tarry fæces; mucosa pale throughout. Rectum presented a number of enlarged veins just within the sphincter, and the external tumour was found to be a collection of enlarged and thrombosed veins.

Kidneys of normal size, pale, a little firm. On section a large quantity of thin watery fluid oozed from the surface. Ureter and bladder normal.

Venous System.—On dissecting the gastro-hepatic omentum the portal vein was found to be obliterated from a point 2 cm. beyond its origin, and converted into an irregular, fibrous cord, with ill-defined margins, being matted with the surrounding tissues. In this state it entered the hilus of the organ, and penetrated the main divisions of the portal canals; no trace of the natural appearance of a vessel could be seen. On section the tissue was spongy, not indurated, and somewhat reddish in colour. There were no remnants of a thrombus, nor any cretaceous or calcified portions. The main branches within the organ were also occluded; the connective tissue of Glisson's sheath was abundant, and firmer than normal. Both artery and duct in these parts could be slit open readily. There was not any special contraction about the hilus, and the liver substance in immediate neighbourhood of the portal canals looked normal. The remaining portion of the portal vein and its branches were in the following condition:—Just beyond the junction of the splenic and superior mesenteric was a large saccular dilatation, the size of a walnut, with thickened walls, and here and there a calcified plate beneath the intima. It was in contact with the under surface of the right lobe close to the hilus. *Superior mesenteric* was much dilated; the terminal part, just behind the pancreas, presented several small sacculi, the intima of which contained atheromatous plates. The mesenteric

branches were moderately enlarged. *Splenic vein* admitted the index finger freely, and all its branches were dilated. Several of those on the anterior margin of the spleen were full of firm thrombi. The vasa brevia from the fundus of stomach were dilated, and some of the larger branches contained thrombi. The left gastro-epiploic was almost as large as the splenic, and at the curvature presented several large dilatations, one of which admitted the top of the thumb. The gastric vein emptied into the dilatation just beyond the junction of the superior mesenteric and splenic, and was also enlarged. The walls of all of these vessels were thickened, the intima a little roughened, and in spots calcified. The *inferior mesenteric* was moderately enlarged; the hæmorrhoidal branches were distended. *Inferior cava* normal; openings of hepatic veins presented nothing unusual. Among its branches the lumbars appeared large, particularly one passing by the side of the third lumbar vertebra. To the left of the aorta was a large vein nearly equal in size to the inferior cava; it terminated below by two branches, one of which passed over and joined the junction of the external and internal iliacs on the right side; the other joined the common iliac of the left side. The appearance of the parts after dissection suggested a double inferior cava. Unfortunately the liver, together with stomach, pancreas, and spleen had been removed before this condition was detected, so that the upper termination of this vessel could not be made out. Two large branches joined it above, but their connection could not be traced. The *iliacs* were large; many of the branches of the internal divisions were thrombosed. The *diaphragmatic* veins formed a close plexus, particularly in the œsophageal region, which united with the veins of the coronary and lateral ligaments of the liver. The *œsophageal* veins were numerous and large, and formed a rich network about the cardia. The veins in the suspensory or round ligaments were not dilated. In the thorax the lower intercostal veins were very large, particularly one running along the lower margin of 10th rib. The *vena azygos major* almost equalled the inferior cava in width, and admitted the index finger easily; the azygos minor was also of large size.

Remarks—Such a case as the above presents many points of

interest. Sudden and violent hæmatemesis in a young man; no ascites, no enlarged abdominal veins, small liver, large spleen, and a localised murmur in the epigastric region—the diagnosis was not easy, but it lay, I thought, between cirrhosis of the liver, splenic anæmia, and an aneurism. For cirrhosis as a cause of the hæmatemesis were—history of spirit-drinking, diminished volume of liver, increase in size of spleen, and the existence of piles; against were—the age of patient, and the absence of many important signs, as gastric or intestinal catarrh, ascites, and enlarged veins. The well-nourished state of the man, the moderate enlargement of the spleen, and normal aspect of the blood, were opposed to the idea that the primary trouble was splenic. The suddenness of the attack, the brightness of the vomited blood, together with the existence of a localised murmur in the epigastric region—the origin of which remains obscure—suggested the occurrence of a small aneurism, either of the aorta or one of the branches of the cœliac axis; but the hæmorrhage in September, the absence of any pulsating tumour, and the state of the liver and spleen, seemed fatal to this view. Altogether, in spite of the absence of many of the important symptoms usually present, the most satisfactory diagnosis appeared to be portal obstruction from cirrhosis. The evident reduction in the volume of the liver was strongly in favour of this view, and as I had also met with several instances in which severe hæmorrhage had been the initial symptom, I was the more inclined to regard it as an anomalous case of this nature. The history of a hæmorrhage in September, followed by an obscure illness of some months' duration, pointed to a chronic malady.

The history of the case offers no clue to the cause of the obliteration of the vein, but we may suppose it to have taken place in the way in which veins usually become occluded, viz., by the formation of a thrombus which organised, and was ultimately converted into a fibroid cord. Apart from marasmatic conditions, in which portal thrombosis occasionally occurs, coagulation of blood in the portal vein is met with—1st, as an effect of compression, as in cirrhosis, in which the pressure is exercised within the liver, or in tumours in the neighbourhood of the hilus, which compress the main trunk in the gastro-hepatic omentum; 2d, by extension of inflammation from the

bile passages, as in cholangitis from obstruction by gall-stones; and 3d, by the extension of inflammation or transference of emboli from suppurating or ulcerative foci in the territory of the portal vessels, but in these instances the thrombi which form rapidly soften, and suppurative pylephlebitis is the result. I have met with cases of pylethrombosis from the above causes, but, so far as can be ascertained, none of them have prevailed in this case. The only possible source which is suggested by the *post-mortem* is the cretaceous area in the spleen, representing the final stage of a small abscess or infarct, which, when in an active state, might have induced, by direct extension or embolism, the pylethrombosis.

The state of the liver is worthy of note. Though shrunken, particularly in left lobe, the greater part of the organ was smooth and not in the least cirrhotic. In the few instances of chronic occlusion which have been reported, the condition has been variable. In Cruveilhier's case [1] in which the obstruction must have lasted for years, the organ is described as smooth and healthy. In others it has been cirrhotic. Solowieff [2] has produced a fibroid condition of the liver by inducing occlusion of portal branches in the dog, but that this is not an invariable sequence, in man, is shown by this, as well as other cases. Nor is there any good anatomical reason why it should occur. After complete exclusion of portal blood from the organ, the lobular capillary plexus continues filled, as the venules which collect the blood from the capillaries of the hepatic artery empty directly into the portal interlobular vessels, and the blood-supply is in this way maintained. Hence the function of the gland is not materially interfered with, and bile continues to be formed from the blood furnished by the hepatic artery, which may, as in this case, undergo a compensatory enlargement. That the arterial blood can in this way act as substitute for the portal supply is well shown by such a case as the one under consideration, which forms an interesting counterpart to the one of aneurism of the hepatic artery [3] which apparently demonstrated that the converse is not true, but that, as Cohnheim and Litten

[1] *Atlas d'anat. path.* livr. xvi. Pl. 6.
[2] *Virchow's Archiv*, lxii.
[3] *Canada Med. and Surg. Journal*, 1877, Drs. Ross and Osler.

state,[1] the portal blood cannot replace the hepatic if the latter be completely excluded from the organ.

There is no more interesting subject of study than the way in which channels of collateral circulation are established in occlusion of large vessels. In the case of the portal vein, numerous opportunities for this purpose are afforded in cirrhosis of the liver, in which the obliteration of many interlobular branches necessitates the development of circuitous routes, by means of which the blood-current in the portal system is equalised, and the reduced carrying capacity of its vessels counterbalanced. In some instances, so adequate is this compensation that the cirrhosis may reach an extreme grade without producing symptoms. These collateral channels have been fully described by Sappey, and are chiefly:—(1) the anastomoses between the coronary veins of the stomach and the œsophageal plexus, which discharges into the lower intercostal and azygos veins, and also communicates with the diaphragmatic vessels; (2) veins passing in the coronary and suspensory ligaments of the liver and in the adhesions which often form between the liver and diaphragm; (3) in some cases a small vein in the round ligament dilates enormously, and affords free communication between the portal vein and the epigastric vessels. Some regard this as a redistended umbilical vein, but Sappey states that it is one of the small vessels which he describes as the *venæ portæ accessoriæ*. I have recorded an instance[2] of advanced cirrhosis, with great narrowing of the portal branches, in which no symptoms of increased blood-pressure existed in the portal system owing to the presence of this vein, which was as large as the little finger; (4) certain veins, forming what is known as the system of Retzius, which, originating in parts of the intestinal canal, and anastomosing with the radicles of the portal vein, discharge into the inferior cava or its branches; (5) the communications which exist between the superior and inferior hæmorrhoidal plexuses. In this case the collateral circulation appeared to have been carried on by the first, fourth, and fifth of these channels. There were extensive communications between the gastric and œsophageal veins, and through the latter with the azygos and liver intercostals. The *vasa brevia* and others about the fundus

[1] *Virchow's Archiv*, lxvii. [2] *Montreal General Hospital Reports*, 1880.

of the stomach were highly developed, and joined the dense network about the cardia and the diaphragm in the immediate vicinity. Many of these branches were plugged with thrombi. Doubtless a large share in the supplementary circulation was taken by the veins of the system of Retzius and the peritoneal branches emptying into the cava. The large vessel to the left of the aorta may have been a greatly distended azygos minor which Henle figures as joining with the left iliacs, but unfortunately its connection could not be made out owing to the removal of the viscera before the nature of the lesion was suspected. The blood in it probably reached the azygos, which was of large size. In the case of obliterated vena cava, recorded in this *Journal*,[1] I met with a similar vein. The hæmorrhoidal plexuses were not greatly distended, but the branches of the internal iliacs, particularly on the left side, were very large, and many of them contained thrombi. The epigastric veins were not dilated.

The collateral circulation must have existed for some time perhaps for years, and was fully compensatory. The somewhat sudden onset of the final symptoms may reasonably be attributed to interference with this free circulation by the thrombi in the gastric veins, and in branches of the internal iliacs.

[1] Vol. xiii.