OBSTRUCTION OF INFERIOR VENA CAVA BY A PERSISTENT EUSTACHIAN VALVE IN A YOUNG ADULT

BY

R. E. ROSSALL AND R. A. CALDWELL

From the General Infirmary at Leeds and the Department of Pathology, University of Leeds

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The Eustachian valve—or valve of the inferior vena cava—is an embryological structure which may persist into adult life as a rudimentary, crescentic, fenestrated membrane lying in the anterior wall of the inferior vena cava at its junction with the right auricle.

Franklin (1948) has comprehensively surveyed the literature concerning the mode of development and possible functions of the valve, and has added many descriptions of its structure and its incidence in mammals. No convincing cases of a persistent Eustachian valve leading to obstruction of the inferior vena cava have been described. This justifies the following description of a case where an exceptionally large persistent Eustachian valve produced almost complete obstruction to blood flow from the inferior vena cava to the right auricle. Had the cause of this obstruction been diagnosed during life surgical treatment might have been feasible.

Case Report

The patient, a youth aged 19, presented himself for National Service medical examination in April, 1951. Although in good health, he had noticed the development of varicose veins of the legs two years previously, and on examination numerous distended superficial veins were seen on the anterior abdominal wall. He was referred to a hospital in Bradford, where this finding was confirmed, the direction of blood flow being upwards towards the axillae. Hepatomegaly and gynaecomastia were noted. Results of investigations were as follows: Hb, 90% ; R.B.C., 4.3 m. per c.mm.; W.B.C., 7,000 per c.m.m. with normal differential; serum bilirubin 1.4 mg.%; plasma proteins 8.3 g.%; (albumin 5.0, globulin 3.3); blood urea 29 mg.%; urine, albumin negative on seven occasions, urobilinogen and urobilin absent. An intravenous pyelogram was normal. The kidneys concentrating the contrast medium satisfactorily.

Obstruction of the inferior vena cava was diagnosed and a laparotomy performed. The abdomen contained a small amount of straw-coloured fluid and the liver was noted to be enlarged and congested with a small, hard, red nodule on the under surface. The renal veins and inferior vena cava were normal, and no evidence of neoplasm was found.

It was concluded that obstruction of the inferior vena cava was present at or above the site of entry of the hepatic veins.

He remained under observation until October, 1951, when he was readmitted to hospital for venography. Attempts were made to introduce a cardiac catheter into the inferior vena cava via a saphenous vein, but this proved impossible. The other saphenous vein was cannulated and attempts made to outline the vena cava with contrast medium, both through the cannula and the cardiac catheter, but in each case the medium entered the superficial abdominal veins and the inferior vena cava was not demonstrated.

He remained relatively well, doing farm work, until July, 1953, when he became ill with malaise and diarhoea followed by epistaxes.

He was admitted to a hospital in Oxford in August, 1953, when he was found to be febrile (105° F. on occasions), slightly jaundiced, and to have numerous large tortuous veins in the abdominal wall (Fig. 1) and on the back. Ascites was present, the liver was enlarged 10 cm. below the costal margin, and healed varicose ulcers were noted on the legs.

The results of investigations were as follows: Hb, 80%; W.B.C.s, 8,300 c.mm. with normal differential; E.S.R., 74 mm. in one hour; prothrombin time, 35% of normal; marrow hyperplastic, no abnormal cells seen; blood culture sterile; sputum and gastric lavage for tubercle bacilli negative; faeces negative for pathogenic organisms; urine negative for albumin on 19 occasions, and Strep. faecalis found on one occasion only; serum agglutinins for Salmonella and Brucella abortus negative; blood urea 37 mg.%; plasma bilirubin 1.8 and 2.2 mg.%; plasma alkaline phosphatase 23 and 25 units; thymol turbidity negative; colloidal gold negative; plasma proteins 6.3 g.% (albumin 2.9). A chest radiograph showed a high diaphragm and a barium swallow was suggestive of varices in the lower third of the oesophagus.

Penicillin, aureomycin, and streptomycin were administered in turn, and while on streptomycin his temperature settled and there was subjective improvement.
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The cause of his febrile illness remained obscure, and he returned to Bradford for domestic reasons.

He remained moderately well until February, 1955, when he developed a further febrile illness with rigors and epistaxes.

On admission to the General Infirmary at Leeds in April, 1955, he was critically ill, jaundiced, and had a high swinging temperature. Gross ascites and bilateral pleural effusions were present. The liver edge was 12 cm. below the costal margin, and the tortuous veins in the abdominal and chest wall were a striking feature. Penicillin, aureomycin, and streptomycin were given in turn, but there was no response, and he died after a short downhill course.

Necropsy

External Appearances.—The body was that of a poorly nourished young adult male of medium build and large stature. The skin and conjunctivae showed a moderate degree of jaundice. Pitting oedema of the feet, legs, and thighs was marked. The abdomen was grossly distended and showed numerous striae and many dilated tortuous veins (Fig. 1).

Internal Examination.—The chief findings are as follows:

Cardiovascular System.—The heart was not enlarged (left ventricular thickness 12 mm., right ventricular thickness 4 mm.). The mitral valve showed a small, dark, wart-like vegetation close to the border of the anterior cusp, but the remaining valves, the chambers, and the myocardium appeared healthy. The coronary arteries and aorta were normal.

The inferior vena cava was almost completely obstructed at its junction with the right auricle by an abnormally large persistent Eustachian valve (Fig. 2), which arose from a cord-like roll of firm tissue encircling the inferior vena cava at its termination and lying within its wall (Figs. 2 and 3). When the vessel was opened, the valve appeared as a thin fibrous diaphragm almost completely obstructing the lumen but leaving a small deficiency lying posteriorly. On closing the vessel, the valve was thrown into redundant folds which hung down into the inferior vena cava. From its inferior surface several tenuous fibrous tags were dependent, to one of which was attached a firm, smooth, ovoid, yellow thrombus measuring 2 x 1.5 x 1.5 cm. approximately. When extended the valve was semicircular, measuring 4.4 cm. along its free upper border and 2.6 cm. in depth. It was of uniform thickness (about 1 mm.) and showed a fine irregularly wrinkled surface which appeared to be covered by unbroken endothelium. The inferior vena cava was of normal calibre.
several sharply defined opaque yellowish nodules of varying size (Fig. 4).

The gall-bladder and pancreas were normal.

Genito-urinary System.—The kidneys were normal in shape, but moderately enlarged, the left measuring 15 × 9 × 6 cm., the right 14.5 × 7 × 7 cm. Surrounding the upper half of the right kidney was a broad band of yellowish induration in which patchy abscess formation was present.

The cut surface of the right kidney showed a large thick-walled abscess cavity in the mid-zone close under the cortex, with several small abscess cavities in the surrounding tissue. Elsewhere there was marked congestion. The cut surface of the left kidney showed chronic congestion.

The branches of both renal veins contained post-mortem thrombus. The ureters, prostate, and testes were normal.

The pus yielded a pure growth of Staphylococcus pyogenes aureus on culture.

Spleen.—This was moderately enlarged (15 × 10 × 7 cm.), firm and congested, the cut surface being uniformly dark red.

Peritoneum.—The peritoneal cavity contained a large quantity of clear greenish fluid with much

throughout, but a small mural thrombus extended along the anterior wall from the orifice of the right renal vein to the hepatic veins.

Respiratory System.—The left pleural cavity contained 900 ml. and the right 500 ml. of clear yellow fluid. Numerous dense fibrous adhesions were present over the left upper lobe and a soft fibrinous exudate over the right lower lobe. The diaphragmatic pleura on the right side showed marked fibrous thickening.

The trachea and bronchi were slightly congested and contained greenish mucopus.

Both lower lobes were partially collapsed, more pronounced on the left side, and their cut surfaces showed marked congestion and slight oedema.

Alimentary System.—A number of submucosal varices were present in the lower third of the oesophagus, but the stomach and intestines were not distended and appeared normal.

The liver was moderately enlarged (28 × 19 × 14 cm.) and bile-stained and showed a fine external mammillation. The right lobe was indented posteriorly by a large thick-walled abscess which contained creamy yellow pus. The cut surface of the liver showed diffuse chronic venous congestion and in the upper half of the right lobe there were


fig. 4.—The cut surface of the right lobe of the liver showing the nodular hyperplasia.
suspended fibrin and greenish gelatinous material throughout, but no definite pus was seen. Dense fibrous adhesions were present between the liver and the diaphragm. The appearances were consistent with a low-grade peritonitis.

_endocrine Glands and Nervous System._—No gross abnormality was seen. Several of the viscera were preserved as a block specimen and it was thus not possible to weigh the individual organs.

_histology_

_Eustachian Valve._—The valve was covered by flattened endothelium and was largely composed of moderately cellular fibrous tissue (Fig. 5), intermingled with numerous small elastic fibres, which have become somewhat condensed in the central part of the valve (Fig. 6). Small arteries and many dilated capillary venules were also present.

_FIG. 5._—Section through the Eustachian valve. Haematoxylin and eosin, × 90.

_FIG. 6._—Section through the Eustachian valve to demonstrate elastic tissue. Weigert’s elastic (Lawson’s modification), × 90.

_FIG. 7._—Section of the valve commissure. Haematoxylin and eosin, × 30.
The thickened commissure was largely composed of hyaline fibrous tissue and presented a laminated appearance (Fig. 7). A narrow band of elastic tissue fibres was present (Fig. 8), continuous with the elastic fibres of the valve itself. This band became more superficial as it approached the right auricle, merging with the subendocardial elastic layer (Fig. 8).

No cardiac muscle fibres were present in the valve itself, but a small group of fibres was seen in the angle between the right auricular wall and the commissure (Fig. 7).

The small vegetation on the mitral valve was composed of cellular fibrous tissue containing a few haemosiderin-laden macrophages and was covered by endothelium. It appeared to be organizing thrombus.

The myocardium showed no significant abnormality.
Liver.—There was diffuse cirrhosis affecting the portal tracts associated with centrilobular fibrosis and periportal sinusoidal congestion (Fig. 9). There was an increase in reticulin linking the central zones with resulting isolation of groups of liver cells (Fig. 10). The portal tracts showed slight, patchy round-cell infiltration with proliferation of small bile ducts.

The macroscopic yellowish nodules consisted of hyperplastic liver cells.

Spleen.—There was fibrous thickening of both the capsule and the trabeculae, together with reduction in size and number of the Malpighian corpuscles. The pulp showed diffuse fibrosis and a few small para-arterial haemorrhages were present. The appearances are consistent with portal hypertension.

Kidneys.—Apart from the acute abscesses in the right kidney these organs showed only marked congestion.

The sections taken from the other viscera confirmed the macroscopic findings and no other significant abnormality was found.

Discussion

Obstruction of the vena cava by a persistent Eustachian valve does not appear to have been recorded previously, although Bennett (1950) reports a case in which obstruction may have been due to such a structure. From the measurements quoted in his case report, however, it is difficult to determine the level of the membranous obstruction, which appeared to lie above the orifices of the hepatic veins yet to be 5 cm. below the termination of the inferior vena cava in the right auricle. The drawing accompanying the text of his report suggests that the membranous obstruction could well have been a persistent Eustachian valve, but it is difficult to define the attachment of the base of the membrane.

The clinical picture of inferior vena caval obstruction presented by our patient did not materially differ from previous descriptions (Thompson, 1947; Jonas and Lawrence, 1954).

The absence of albuminuria has previously been noted (Jonas and Lawrence, 1954), and this is presumably a reflection of the efficiency of the collateral venous circulation, whereby there is no persistent elevation of the renal venous pressure.

The nodular hyperplasia of the liver, also noted by Jonas and Lawrence (1954), is doubtless a late result of hepatic cirrhosis following raised hepatic venous pressure, and the histological features presented here accord with Sherlock's description of cardiac cirrhosis (Sherlock, 1951). There was no evidence of long-standing local obstruction of the hepatic veins, and the gross anatomical disturbances found in the liver form a striking contrast to the relatively normal anatomy and functional state of the kidneys, both organs presumably being subjected to the same order of back pressure.

Previous descriptions of the histology of the Eustachian valve have been made by Gätzi (1929) and by Benninghoff (1930), and no material difference from their accounts has been found in the present case.

The largest persistent Eustachian valve recorded is that described by Gätzi, which measured 19 mm. in height, but as no measurement of length or clinical details accompany his statement it is difficult to draw comparisons and to infer at what dimension symptoms of obstruction are likely to occur.

Summary

A case is presented of inferior vena caval obstruction with fatal termination due indirectly to persistence of an abnormally large Eustachian valve. This valve appears to be the largest so far recorded, and the clinical and necropsy findings are reported in detail.

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References