HEALED DISSECTING ANEURYSM

BY

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Received February 2, 1946

Since Shennan (1934) published his well-known report on 300 cases of dissecting aneurysm the literature of this condition has become extensive, and Sailer (1942) stated that there were then some 500 published cases, of which 33 had been diagnosed before death. There is general agreement that in some 80 per cent of these cases death occurs within a few hours of an acute onset, and that most of those patients who survive the accident of dissection are cardiac cripples who usually die of internal hæmorrhage or of cardiac failure within a year or two. There are, however, exceptions, such as Hall's (1926) patient, a boy aged 17, in whom the dissection occurred after a race, with such good recovery that he led a strenuous athletic life for 15 years. The survival record seems to be held by Graham's (1886) patient, who lived for 30 years after the dissection.

There is general agreement, too, as to the diagnostic criteria of the acute phase of the dissection. These have been well summarized by East (1939), who at the same time reports a personal case, diagnosed before death, of a woman, aged 43, who survived 5 years after the onset, despite a pressure of 300/150 during this period. That the healed dissection may be diagnostically obscure is pointed out by Gouley and Anderson (1940), who describe six patients in whom the onset was insidious and the clinical picture was that of cardiovascular syphilis. Pain was absent in four, inconspicuous in one, and occurred only intermittently for three weeks before death in one. All exhibited cardiac decompensation for periods varying between two months and four years. They all had big hearts, aortic regurgitation, and a dilated aorta radiographically: the Wassermann reaction was positive in one case only.

Others who have described aortic regurgitation with normal cusps in dissecting aneurysms are Resnick and Keefer (1925), Borger (1906), and Letulle (1905).

The case we now publish was, like those of Gouley and Anderson, diagnosed as syphilitic aortic reflux and aneurysm, though the possibility of a dissecting aneurysm was considered. The case is also of interest as illustrating the remarkable capacity for effort on the part of this victim of a healed dissection.

A Dutch engineer, aged 42, was first seen on March 19, 1942. He had gonorrhœa in 1921; his Wassermann reaction was then negative. In 1936, when aged 36, he played a long set of singles at lawn tennis in Holland and became so abnormally distressed that he had to stop. A week later, at 9 p.m., he was seized with very severe epigastric pain, which radiated to the angles of the scapulæ, more severely on the right side. He said that for two or three nights he was walking about in agony, and that morphia had to be injected, and that the pulse was said to have been very irregular: renal colic was diagnosed. He stayed at home for a fortnight, and subsequently resumed work, after which he noticed a tendency to dyspnœa and palpitation on vigorous exercise. In February 1937 he was investigated in a Dutch hospital; the aorta was "enlarged" and though the "blood test" was negative, he was given a course of intravenous arsenic. After this he was well enough to swim, and to play tennis, but not with his former vigour.

In 1940 he escaped from Holland on foot with his wife and children, carrying much baggage. He was able to swim and play tennis after his arrival in England that summer.

During January and February 1942 he became increasingly dyspnœic on exertion, and there was paroxysmal dyspnœa at rest. When examined on March 19, 1942, he was a

well-built man, not dyspnoëic at rest, and presenting no evidence of congestive failure. Pulsation was seen and felt to the right of the sternum, where there was a diastolic shock. The presence of dilated venules along the line of diaphragmatic attachment suggested early vena azygos obstruction. The pulse was 92, with frequent premature beats; the blood pressure 140/60; and the arteries not unduly thick. On auscultation, a loud diastolic murmur was heard at the base and down the left sternal edge; the murmur was louder to the right than to the left of the sternum. No tracheal tug was felt. The knee jerks were brisk, and the pupils a little sluggish to light and not quite circular. On X-ray screening, the whole thoracic aorta was diffusely dilated. There was a considerable degree of left ventricular enlargement (Fig. 1), and the trachea was displaced to the right. His electrocardiogram showed numerous right ventricular premature beats, but was otherwise normal (Fig. 2, A).

FIG. 1.—Orthodiagram on 25/5/42, showing left ventricular enlargement and dilatation of the aorta.

The diagnosis was aortic aneurysm, with aortic regurgitation. In view of the acute onset after violent physical effort, the possibility of either a ruptured valve or of a dissecting aneurysm was discussed. The latter diagnosis was considered to be improbable, because, apart from the onset, the clinical picture was typically that of a non-dissecting thoracic aneurysm. Moreover the radial and femoral pulses were equal, and it was thought that the combination of a dissecting aneurysm with free aortic regurgitation would hardly allow such a degree of physical activity as the patient had proved himself to be capable of. In spite of the fact that his Wassermann reaction was negative he was treated with mercury and iodide, and subsequently with stovarsol orally. When seen on May 25, 1942, he was working at his office full time, dyspnoëic only on hills, and sleeping well with only one pillow. Dr. Anwyl Davies thought that in spite of the negative W.R. the condition was almost certainly syphilitic and advised further treatment with mercury and iodide.

In January 1944 he was still hard at work and almost symptomless, except for what

FIG. 2.—Electrocardiograms. (A) Normal rhythm on 24/1/44. (B) Auricular fibrillation with ectopic beats on 21/5/44.

FIG. 3.—Orthodiagram taken on 9/1/45, showing further enlargement.

sounded like a paroxysm of auricular fibrillation during influenza a month previously. He had spent some months in Sweden, flying there and back under war-time conditions. Radiographically both heart and aorta were a little bigger. The electrocardiogram was unchanged.

In April 1944 there were further paroxysms of auricular fibrillation, lasting about two hours, and his condition had deteriorated when seen on May 21, 1944. Fibrillation had now become permanent, with multiple ventricular ectopic beats (Fig. 2, B). He was strongly advised not to follow the army of occupation into Holland, but felt that his knowledge of

the Dutch canals and their pumping stations was so unique that he insisted on going. He went by air to Brussels in September 1944. On his return he was able to work in his office only two or three days a week. He was increasingly dyspnoeic on slight exertion, but his ventricular rate was easily controlled by a small dose of digitalis, and there was no congestive failure. In January 1945 an orthodiagram showed some further enlargement to the right (Fig. 3). Subsequently his condition deteriorated rapidly, and on March 4, 1945, he was admitted to St. Thomas's Hospital on account of increasing paroxysmal dyspnoea without evidence of congestive failure. On March 17, at 9.30 p.m., there was a sudden onset of sternal pain with severe dyspnoea, rapid pulse, sweating, and pulmonary oedema. He improved under morphia, but died within a few minutes at 10.30 a.m. on March 18.

AUTOPSY REPORT

The body was that of a well-nourished adult male.

Heart. A moderate excess of straw-coloured fluid was present in the pericardial sac. The heart was greatly enlarged, both sides showing the increase, but particularly the left. On section, the left ventricle showed hypertrophy, the thickness of its wall varying between 1.5 and 2.0 cm. Its cavity was dilated, the maximal internal diameters being approximately 7 cm. horizontally and 9 cm. in the superior-inferior direction. The right ventricle showed these changes to a corresponding degree, the thickness of its wall varying between 0.6 and 1.0 cm., and its internal diameters being both approximately 8 cm. The ventricular muscle appeared homogeneous and of normal consistency. The left auricle contained a thrombus in its appendix of approximately 1.5 cm. in diameter.

The tricuspid and pulmonary valves were normal. There was slight thickening of the edge of the posterior cusp of the aortic valve, and atheroma of the aortic cusp of the mitral valve. The aortic ring appeared dilated, its diameter being about 3 cm. The site of the intra-pericardial part of the aorta was occupied by a soft spherical mass having maximum diameter of 7.5 cm. (Fig. 4). The swelling lay superior to the right ventricle and anterior to

FIG. 4.—Heart and intra-pericardial aneurysm (anterior aspect). (A) Aneurysm.

the right auricle, compressing the latter posteriorly. Its surface was smooth, shiny, and for the most part of pale, blue-white colour. The mass was not adherent to any of the surrounding cardiac tissue except at the line of junction.

The coronary arteries were normal except for very slight patchy atheroma.

Aorta. On section, a horizontal rent in the intima was seen, extending from above the middle of the left posterior cusp round the left side across the anterior surface to the right lateral aspect, terminating just posterior to the union of the anterior cusp with the right posterior one. The maximal width of the rent was 2.5 cm.; its length was approximately 8 cm. The edge of the rent above the left posterior cusp was formed by free intima, which extended for about 3 cm. along its length. Here the floor was recently formed blood clot, part of the surface of that filling the intra-pericardial aneurysmal dilatation (Fig. 5). For the rest of the rent, the intima was bound by adhesions in an irregular manner to the media,

FIG. 5.—The same after opening. (A) Left ventricle. (B) Aorta. (C) Aperture into aneurysmal cavity showing surface of blood clot within.

FIG. 6.—More detailed picture of the split. (A) Left ventricle. (B) Rent in aorta, showing adhesion of split intima to underlying media.

which in this area also formed the floor (Fig. 6). The maximal width of the intimal tear was at the site of the visible portion of the blood clot and the adjacent area of the medial floor.

At the line of contact of the above-mentioned blood clot and medial floor, i.e. approximately superior to the junction of the anterior and left posterior cusps, there was free space which communicated superiorly and slightly to the right with a dissection in the posterior part of the ascending aorta. That part of the vessel immediately inferior to this free space showed slight saccular dilatation and gross irregularity of surface, much of which was due to localized atheroma.

It was apparent that the free space above mentioned formed the path of flow for the blood into the dissected false passage, and the small saccular dilatation and roughening of the aortic surface below was brought about by the impinging of the blood here on its passage through. The aneurysmal dilatation into the pericardium and the rent with a free intimal edge indicated a much more recent change.

The dissection, which initially lay posterior, tracked slightly to the left as it progressed upwards. The aneurysm thus formed posterior, inferior and right lateral relationships. The left lateral relationship was formed by the left pulmonary artery. The diameter of the dissection was here approximately 3 cm.

In the region of the arch of the aorta the false passage lay posterior and slightly inferior. There was only very small penetration upwards round the beginning of the innominate, left common carotid and subclavian arteries, reaching a maximum of 0.5 cm. These vessels thus formed the most superior border of the false passage (Fig. 7). As the aorta curved down-

FIG. 7.—A view of the aorta further on. (A) Descending thoracic aorta. (B) Innominate artery. (C) Left common carotid artery. (D) False passage.

wards the false passage changed its posterior and inferior relationships to a posterior and slightly right lateral one, but when the diaphragm was reached it lay exactly posterior. At the bifurcation of the abdominal aorta the false passage lay right lateral (Fig. 8). Here the true aorta appeared continuous with the left common iliac artery, whereas the false passage descended down the right one for approximately 4.5 cm., 0.7 cm. above the division into the internal and external iliac branches. The site where the false joined the true arterial channel was clear cut. The former lay at first right lateral and then anterior to the first part of the true right common iliac channel, with the result that the free edge of union lay posterior to the former and anterior to the latter. The opening of the true right common iliac artery into the aorta had become displaced as a result of the dissection, appearing as a slit-like branch of the aorta where it became continuous with the left common iliac artery.

The false passage showed atheroma with ulceration and calcification on the wall not shared with the true passage, the change being most severe in the right common iliac artery 1.5 cm. up from the site of junction with the true iliac artery. It extended, however, also upwards to a point approximately 4 cm. from the bifurcation. There was no

FIG. 8.—Bifurcation of the abdominal aorta. (A) The distal end of the aorta. (B) Left common iliac artery. (C) Atheromatous plaque in false passage. (D) Line of union of false passage with the true right common iliac artery.

FIG. 9.—Cross-section of aorta at the level of the diaphragm. (A) True passage. (B) False passage.

change to a similar degree on the common wall in the corresponding places. The true aorta was also moderately atheromatous in all its extent. The posterior wall of the false passage showed in addition great unevenness of surface, raised white areas standing out from depressed more grey-white areas. There was also considerable wrinkling of the surface, particularly in a longitudinal direction.

Cross-section of the two channels at the level of the diaphragm revealed the internal diameter of the aorta when compressed to be 2 cm. and that of the false passage 3 cm. Thus there was a tendency for the latter to flank the former on its lateral aspects (Fig. 9).

The thickness of the true aorta varied from 0.15 cm. to 0.2 cm. The wall common with the false passage was only slightly less thick than the unsplit portion. The posterior wall of the false passage measured 0.10 cm. The cross-section of the common wall was clearly made up of three layers, a central yellow portion and slightly thicker light grey portions on either side. On the anterior aspect of the true passage the yellow layer became compressed and the inner light grey layer was proportionately more thick. The rest of the wall of the false passage was composed of light grey tissue only. The outer wall of the whole was covered by a very thin layer of softer pinkish-grey tissue.

The weight of the heart together with the intra-pericardial aneurysm and its contents was 1190 g.

Branches of aorta. The intercostal artery openings were patent in the true aorta, and some were linked with corresponding openings in the false passage by thin strands of fibrous tissue. The celiac, superior mesenteric, inferior mesenteric, left renal, and left testicular arteries opened direct into the true channel. The right renal and right testicular joined the false channel, corresponding apertures being present also in the common wall. All the lumbar branches of both sides opened into the false passage. The corresponding apertures in the true passage were patent, and some of these, like the corresponding intercostal vessels, were linked by fibrous strands to the false passage openings. The middle sacral artery was not involved in the dissection and therefore opened into the true passage immediately above the bifurcation.

Respiratory system. There was a large excess of straw-coloured fluid in the left pleural cavity. Firm adhesions were present between the parietal and visceral pleuræ on the right side, especially in the basal region. A small excess of pleural fluid was present in this cavity also. The trachea and main bronchi were full of frothy straw-coloured fluid. The lungs (right—1021 g.; left—794 g.) were extremely œdematous, but apart from patchy basal collapse, slight congestion, and emphysema they showed no further macroscopic abnormality.

The *liver* (2168 g.) showed moderate nutmeg change.

The *spleen* (298 g.) was firm, cut crisply, the cut surface being of deep purple colour.

The *left kidney* (383 g.) was congested.

The *right kidney* (326 g.), in addition to this change, contained an infarct in its upper half. The surface area involved was approximately 2 cm. square, having an irregular contour. The central part was light yellow and immediately surrounding it there was a zone of congestion. On section the infarct was shown to be wedge-shaped, the apex reaching the rim of the pelvis.

The remaining *abdominal organs* showed congestion.

There was a cortical adenoma in the *left adrenal*, 1.2 cm. in diameter.

The head was not opened. There was no macroscopic abnormality of mouth, larynx, pharynx, and thyroid.

HISTOLOGY

Aorta. Sections were cut of the vessel (1) at the site of the rent and (2) just superior to the diaphragm. They were stained with hæmatoxylin and eosin, Van Gieson's stain, a modification of Weigert's elastic stain, and with Scharlach R for fat.

(1) The section was taken longitudinally through the aortic wall to include an area of the rent where the media formed the floor, and where the pulmonary artery was contiguous with the vessel. The media of the aorta here showed advanced degeneration. There was extensive fibrous replacement, diffuse mucoid degeneration and a few patches of necrosis. In one or two places the mucoid change had led to the formation of small lakes in the medial tissue. No fatty degeneration was demonstrated. The intima was a little atheromatous and the adventitial coat had undergone hyaline fibrosis.

(2) The split had taken place in the media approximately at the junction of the inner two-thirds and outer third (Fig. 10). The internal elastic lamina and the stripe of the true channel were not easily seen throughout the whole circumference of the vessel. The intima

FIG. 10.—Photomicrograph of aorta at the level of the diaphragm (modified Wiegert's stain). (A) Lumen of false passage. (B) Intima of true passage. (C) The site of split in the media.

showed marked hyperplasia for about a third of its circumference, i.e. for the extent of nearly the whole of the wall not shared with the false channel. This hyperplasia was most marked between the internal elastic lamina and the stripe; the hyperplastic tissue showed also a hyaline degeneration. In the area of intima contiguous to the split in the media there were some fair-sized capillary blood vessels.

The false channel partly overlapped the true channel. The wall showed a variable thickness. The wall common with the true passage consisting of approximately two-thirds of the original media was thicker than that elsewhere. The thinnest portion was that diametrically opposite this common wall where there were only a very few elastic fibres. The media showed a progressive diminution in thickness between these two points. Under low power, in the section stained with hæmatoxylin and eosin, the false passage showed an inner layer of pale staining tissue, an intermediate zone staining light pink, the original media giving a deep pink, and lastly the outer adventitial zone. It appeared thus that a new "intima" had been formed. The elastic stain section showed that in both the inner and intermediate zones there was still considerable elastic tissue, the density of this becoming less as the lumen was approached, this diminishing content of elastic tissue accounting for the progressive lightening of the stain in the hæmatoxylin and eosin section. In both these zones the elastic tissue was very degenerate, contrasting sharply with that in the normal media. As a result of the high content of elastic in the new "intima" the inner surface of this false tube showed fine corrugation. Its surface was covered by a single layer of flattened endothelium. At that angle formed by the medial split which was not associated with intimal hyperplasia in the true passage there was a small valve-like projection from the "intimal" surface of the false passage. This consisted of a central core of dense elastic surrounded by a rim of less dense and degenerate elastic.

The adventitia surrounding the whole vessel was comparatively normal. A few elastic fibres were present. Collagen was a little more dense outside the free wall of the false passage where the elastic media was thinnest. The small vascular channels were dilated and full of blood and there had been hæmorrhage into the surrounding tissue. Round one of these vessels there was an accumulation of small round cells.

In summary, the histological picture confirmed the clinical conclusion that dissection down the aorta had occurred a considerable time before death. The period of active repair had passed and endothelium had formed on the surface of the false passage. Further, the appearance of atheromatous degeneration with surface ulceration and calcification in those parts of the false passage not shared with the true passage, without any corresponding change in the wall common with the true passage, is added evidence that the dissection was of old standing.

Kidneys. Both congested. Right kidney, four recently infarcted areas: an artery in relation to the largest of these contained a small recent thrombus. The capsule and the perirenal fatty connective tissue overlying the infarcted areas showed organizing chronic inflammatory granulation tissue. Adrenal congested with cortical adenoma.

Liver and spleen. Venous congestion, and slight fatty degeneration of liver.

Lungs. Slight congestion. Œdema. Emphysema.

SUMMARY AND CONCLUSIONS

A case of healed aneurysmal dissection of the aorta, of nine years' duration, is described. For some years the patient was able to live a very strenuous life, till symptoms of left ventricular failure appeared, the diagnosis then being syphilitic aneurysm with aortic regurgitation.

The autopsy findings are reported in detail.

There can be little doubt that the original dissection occurred in 1936, nine years before death. The diagnosis at this time was renal colic, and it is interesting to find that the right renal artery arose from the false channel. Renal colic and hæmaturia have been described in reports of acute dissections, which sometimes involve one or both renal arteries.

The histological appearances of the aorta at the site of the intimal tear conform with those of idiopathic cystic medio-necrosis of the aorta (Erdheim, 1929; Moritz, 1932; Roberts, 1939; and Davies, 1941). It is not unreasonable to assume that these changes were present to a degree at the time of the original accident.

Our thanks are due to Mr. A. E. Clark, for his assistance with the photographs, and to Dr. R. W. L. Todd, who referred the patient to us.

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EDITORIAL NOTE ON ANOTHER HEALED CASE

With the approval of the authors the notes of another healed case are added. Dr. A. G. Gibson has given me details of a woman where recovery was complete but unluckily she had no relatives from whom a history of the original illness could be obtained. She died from a cerebellar hæmorrhage and all that was known was that she had worked as a domestic servant for the last six years of her life without any serious illnesses or disability.

The whole aorta consisted of a double tube from just below the origin of the left subclavian artery to the left iliac artery. The figure (Fig. 11) shows the opening of the true aorta.

Autopsy by Dr. A. G. Gibson

A well-nourished woman, aged 46.

Heart. Hypertrophied. Sclerotic thickening of one aortic valve cusp. Atheroma at base of aorta and anterior leaflet of mitral. Both ventricles hypertrophied. No pulmonary embolism. Coronaries thickened but patent.

Thyroid. Large degenerating cyst in upper pole of l. lobe.

Aorta. Beginning from the descending part of the arch just beyond the left subclavian artery was another tube attached to the inner and posterior wall, heading down through the abdominal aorta to end in the left iliac artery one inch from its origin from the aorta. There was a wide opening at its origin in the aorta.

Lungs. Both collapsed.

Abdominal viscera. Normal except for some congestion.

Kidneys. Left: small, congested, slightly granular; arteriosclerotic scars. Right: diminished cortex; as left. Suprarenals. Both hypertrophied cortex.

Brain. Bleeding into both lateral ventricles (liquid). Clotted blood in the 3rd ventricle, in the iter and the 4th ventricle. Massive hæmorrhage in the middle part of the cerebellum which had reached the surface of the r. lobe. Some bleeding into anterior r. middle and posterior fossæ. Right sphenoidal sinus double. Lower one thickened mucosa and slight pus. Maxillary antra nil.

FIG. 11.—Photograph of the aorta, showing the opening of the true aorta and a healed dissecting aneurysm.

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