SUBARACHNOID HAEMORRHAGE

Coarctation of the Aorta and Successful Aortic Homograft

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Morgagni was thought first to have described the condition of coarctation of the aorta in 1760. Crafoord and Nylin (1945) discussed the surgical treatment of coarctation, describing two male patients, aged 12 and 27 years, in whom successful surgery had been performed. Surgical treatment was also described by Kirklin *et al.* (1952), in an infant ten weeks old and at the other end of the scale, Gross (1949), described a successful operation in a man aged 41.

Brock *et al.* (1953), in a discussion on the use of aortic homografts mentioned the work of Alexis Carrel on the preservation of homografts by refrigeration as early as 1912. Lichtenberg and Gallagher (1933) described what was then stated to be the only case of coarctation associated with intermittent leakage of a cerebral aneurysm diagnosed during life in a girl, aged 12, still living 18 months after the cerebral accident.

While it is now a well-known fact that coarctation of the aorta and intracranial vascular accidents are commonly associated, the clinical picture of coarctation, subarachnoid haemorrhage and successful aortic homograft in a patient, is considered sufficiently rare to warrant the following case report:—

Clinical Features

A male patient, R.J.S., aged 27 years was admitted to Walton Hospital, Liverpool on August 17, 1953. The history of his illness was mainly obtained from his wife and general practitioner on account of the confused and disorientated mental state of the patient.

He was perfectly well, and at work, until eight days prior to admission. He returned home and during the course of the evening he complained of a severe frontal headache, pain in the neck and back and blurring of vision, associated with three or four vomiting attacks. At the time he was mentally confused and his speech was slurred, he felt somewhat dizzy, and he retired to bed where he remained until his admission to hospital. These features gradually regressed for six days, but dur-

ing the next two days he had a further exacerbation of symptoms with complaints of frontal headaches, backache, an inability to move the right leg and a feeling of detachment from reality, together with a speech disability. There was no other relevant recent history available. He was known to have been discharged from the Army in 1943, suffering from a hypertension of a severe degree, i.e. 290/130.

Physical examination on admission revealed that he was drowsy, somewhat unco-operative, detached, disorientated and febrile. The pupillary reactions were normal, but the optic fundi showed early bilateral papilloedema and grade two hypertensive retinal arterial changes. There were no cranial nerve palsies. The arms possessed normal tone and power, the triceps and supinator reflexes were sluggish and equal and the biceps reflexes were absent. The right upper and left lower abdominal reflexes were present. The right leg was flaccid and weak with absent knee and ankle reflexes and there were bilateral extensor plantar responses. There was no neck stiffness and a doubtful Kernig's sign. The blood pressure in the arms was 200/120, with a pulse of 70 per minute. There was some cardiac enlargement and a rough systolic murmur was heard all over the praecordium, in the neck and the right side of the chest.

No abnormal arterial pulsations on the chest wall were seen at this stage. At a later stage it was observed that the femoral pulses were very difficult to feel, that they followed the radial pulses and the other lower limb arteries were not palpable. The femoral blood pressure was found to be 150/85. Suzman's sign was demonstrable, palpable anastomotic vessels were present over both scapulae.

Progress

Within 24 hours of admission his mental state improved, but at the same time further examination revealed a greater degree of papilloedema and retinal vein engorgement, more marked neck

stiffness and a definite positive Kernig's sign; a cardiac apical triple rhythm was also noted.

He was thus considered to be a case of subarachnoid haemorrhage due to hypertension with an associated right lower limb monoplegia and a dysarthria. The further signs described above indicated a probable cause of the hypertension, i.e. coarctation of the aorta. A lumbar puncture was performed and it revealed a uniformly blood-stained cerebro-spinal fluid under increased pressure with a xanthochromic supernatant fluid on standing. He remained very ill with urinary incontinence, and four days after admission he developed a paresis of the right arm and right lower face, the blood pressure remaining at 170/100. However, with treatment his general condition began slowly to improve and certain further investigations were carried out to determine the cause of his hypertension more accurately.

Investigations

Repeated urine examinations were completely normal and likewise the renal function tests. The blood and C.S.F. W.R. tests were negative and the C.S.F. was as described above. The intravenous pyelogram was normal and the Piperoxane Hydrochloride test was negative. The electro-cardiogram was normal. The chest radiogram and cardiac screening showed a hypoplastic aorta with marked rib notching and cardiac enlargement, compatible with the diagnosis of hypertension due to coarctation of the aorta.

We obtained the opinion of a Neurosurgeon, Mr. A. Sutcliffe Kerr, on the patient's cerebral condition and he reported that the likely diagnosis was subarachnoid haemorrhage due to a ruptured berry aneurysm on the anterior communicating artery, causing a right hemiparesis and dysphasia and a partial dysarthria.

It was thought that an angiogram at the time was unlikely to give information that would be a guide to treatment, and that it would be wise to use conservative measures for six weeks.

Treatment

He was treated with rest in bed and physiotherapy. Later, he was given a test dose of a Hexamethonium compound by injection which reduced the blood pressure from 180/100 to 150/80, and subsequent therapeutic use of hypotensive agents kept the blood pressure at a temporary lower level. By means of physiotherapy it was not long before the patient was able to move his limbs and the right hemiparesis eventually recovered completely.

We now considered the question of surgical treatment of the coarctation of the aorta, and

having been seen by Mr. J. K. B. Waddington, he was subsequently admitted to the Regional Thoracic Surgical Unit for exploratory thoracotomy, the operation being carried out on January 13, 1954.

At operation an extensive collateral circulation was present, which made it rather tedious getting into the chest. Inside the chest a constriction of the aorta, 3 mm. in diameter, about 6 cm. below the origin of the left subclavian artery and about 1.80 cm. in length, was found. It was decided that in view of his age, and the extent of the gap likely to be produced after excision of the coarctation, an aortic graft would be more satisfactory than an end-to-end anastomosis, although the aorta itself appeared healthy.

Therefore, the coarcted area was excised, and a piece of freeze-dried human graft, 4.5 cm. in length, was inserted into the 'gap' and sutured with interrupted silk sutures; the graft in this case was taken from the Artery Bank at Walton Hospital, which was under the direction of Mr. A. F. Murphy, F.R.C.S.

The pre-operative blood pressure was 240/120 and immediately post-operatively it was 180/110. He was kept in hospital for a month and at the end of that time the circulation in the lower part of the body had considerably improved with good femoral pulsation.

He was last seen by me on July 20, 1956. He told me that he was extremely fit and active at work, and that he had had no further symptoms of any kind since the operation two and a half years ago.

The blood pressure was 160/100, the heart slightly enlarged and a grade III apical systolic murmur was present. A short soft systolic murmur was also audible in the left infra-scapular region and right posterior axilla, but very much softer than before operation. The arterial pulses in the lower limbs were all present and the femoral blood pressure was 130/95.

Discussion

It is proposed to discuss this case briefly, pointing out some of the more interesting features which are considered to be of clinical value. Many authors have pointed out the necessity of early recognition and diagnosis of coarctation of the aorta, stressing the high mortality rate in untreated cases before middle life and the feasibility of curative surgical treatment in the young patient with a subsequent good prognosis and freedom from some of the grave complications of the condition.

The other aspect of this case, namely the presence of subarachnoid haemorrhage, also complicates the problem and affects the ultimate

prognosis in later life. Blackford (1928) discussed a large series of cases of coarctation, pointing out the death rate of 254 out of 323 cases, mostly in males, before the age of 40 and stressing the importance of early diagnosis.

Riefenstein and Levine (1947) surveyed 104 autopsies. They noted the high mortality rate and stated that although some people live a long and useful life, the abnormality brings many hazards to its possessor. A particular note was made of intracranial lesions in which patients died with subarachnoid haemorrhage due to ruptured intracranial aneurysm. These cases were responsible for 10.6 per cent. of all deaths in the series at an average age of 28. The aneurysms were either single or multiple and probably of the congenital type.

Early diagnosis was considered essential in view of the fact that at a later age the aortic wall might become thinned and dilated, surgical treatment then being impossible. The frequently associated subarachnoid haemorrhage from intracranial aneurysms militated against the hopeful expectation of surgery, but surgery was the only real cure. The various causes of death and their relative frequency were discussed by Gross (1949), when he reported a series of 60 patients, between the ages of 5 and 30 upon whom he had operated. There were seven deaths, and 41 patients were completely relieved of hypertension. The author stressed that in older patients arteriosclerotic aortic changes develop which increased the risk of operation, but some excellent results had been obtained in patients in the late twenties. Very young children, on the other hand, have small aortae which cannot be worked on and it may be found that in adult life the anastomosis may not be adequate. Aortic graft was suggested as an alternative measure where end-to-end anastomosis was found impossible. This patient is nearing the upper age limit for operation, although still within the acceptable range, Gross (1949), Crafoord (1945).

The other important matter for consideration is the mode of presentation of these cases.

Analysis of the history shows that this patient was first known to have hypertension in 1943, aged 17. It is thought important to stress here the necessity of following up all lines of clinical investigation at the first opportunity in such cases, to ensure accurate diagnosis, particularly in a curable complaint.

The recent history of the illness which brought him into hospital showed two clear 'episodes,' separated by a few days of partial recovery. Abbott (1928) has pointed out that the intermittent leakage phenomenon is a diagnostic sign of a small repeated rupture of an aneurysmal sac

and in the present case it seems the most likely explanation in view of the confirmatory evidence of lumbar puncture and subsequent clinical progress. Abbott stated that in view of the extreme difficulty in locating these very small aneurysms it was safe to assume the presence of a ruptured cerebral aneurysm in practically all cases of so-called spontaneous cerebral haemorrhage occurring in coarctation of the aorta before the end of the second decade of life.

The particular value of the examination of the chest for collateral circulation and the lower limb circulation for alteration in pulses and blood pressure is illustrated in this case, where no other cause for his hypertension could be found, either clinically or pathologically.

Doraman and Beck (1954) illustrated some of these points in their description of a case of subarachnoid haemorrhage, coarctation and hypertension in a boy of 18 years. He had two attacks of headache, each lasting a few days, interrupted by a symptomless period of two weeks; and in addition he had a left third nerve palsy. A successful operation was performed, and the peripheral lower limb pulses then became palpable.

A report on the neurological complication associated with congenital stenosis of the aortic isthmus was made by Woltman and Sheldon in 1927. General manifestations were headache 44 per cent., hemiplegia 34 per cent., convulsions in 19 per cent. cases. At necropsy the cause of death was ascertained as follows: cerebral haemorrhage 37.5 per cent., cerebral softening 19 per cent. and aneurysm with rupture 9.4 per cent. Three of the main features in the case presented were headache, right hemiplegia and dysarthria. It should be emphasised that more than one cerebro-vascular accident may occur before the fatal one and this might even give time for surgery in the intervening period. Sealy in 1953, reported a series of 28 cases in which 22 were operated on between the ages of 16 months and 31 years, and one not so treated, which subsequently died, had an exploratory thoracotomy, but excision of the coarctation was deferred as a graft was not available. It has been suggested by Brock (1953) that operation on patients in their late teens or older should not be performed unless a homograft was available at the time. Crafoord (1948) in a paper read to the International Conference of Physicians, stated that in his view exploratory thoracotomy was no more dangerous than laparotomy and is certainly justified in order to determine whether or not resection could be performed. However, cardiac catheterization, angiocardiology and other methods of investigation have much reduced the necessity of this procedure.

Sealy (1953) points out in some detail the indications for surgical treatment of coarctation. He states that it has its uses, particularly as a prophylactic measure to prevent rupture, cardiac failure, endocarditis or cerebral catastrophes. He discusses the use of grafts and anastomoses, emphasising that over the age of 30 it is rather a hazardous procedure.

Finally, there is a report by Gross (1951) of 19 cases of coarctation treated by homologous grafts. In this series 80 per cent. of the patients had the blood pressure restored to normal, a fact which so far has been observed in the present case.

Summary

A case report of a patient suffering from subarachnoid haemorrhage and coarctation of the aorta, treated successfully by aortic homograft, is presented.

The apparent rarity of such a remarkable sequence of events is indicated.

Some attempt has been made to show the importance of discovering the exact cause of hypertension in the younger patient at the earliest age whenever possible (the gap of 10 years, between 1943 and 1953 in this case being noted), and the modes of presentation of these cases are described,

particularly in relation to cerebro-vascular catastrophes.

Early diagnosis and recognition of the condition is stressed, and this is shown vitally to affect the prognosis.

The value of an artery bank being available for the use of thoracic surgeons dealing with these cases is emphasised.

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* These articles have not been quoted in the text. Each one of these articles is a classical contribution to this branch of medicine.