related to the rate of passage of R.P.P. back through the capillary walls. The second mechanism, on the other hand, would involve some delay after the injection before the R.P.P. content of the blood began to rise, corresponding to the time it takes for the R.P.P. to pass through the lymphatic system and hence into the blood stream. Longer delays would be associated with slower functioning of the lymphatic system, and it might be expected that the subsequent rate of increase of R.P.P. in the blood would be correspondingly slower.

With the exception of the result for the transected animals the variation of R.P.P. in the blood has followed the pattern expected from the second mechanism. In human subjects the initial delay is longer and the initial rate of increase of R.P.P. in the blood is much smaller in the patients with severe lymphoedema than in the control group of normal subjects. In the latter the observed values are also related to the degree of physical activity. It appears, therefore, that in the cases examined the lymphatic system is the most important mechanism for the transport of proteins from the tissue fluids. The iodide contamination of our R.P.P. made it, unfortunately, impossible to decide whether there is a return of a small proportion of protein directly to the blood through the capillary walls.

The transected animals are the only group in which blood absorption of R.P.P. appears to be taking place. Here the lymphatic route is destroyed and the normal physiological conditions are disturbed by trauma so that appreciable passage of R.P.P. in the reverse direction through the capillary walls becomes possible. Even in these conditions the removal of protein from the tissue, as measured by the initial rate of increase of R.P.P. in the blood, is considerably (about three times) slower than in the control animals.

The results of this investigation indicate that the behaviour of R.P.P. injected subcutaneously can be used as a test of lymphatic function. The method is useful as a research tool, but it is too expensive and time-consuming for routine clinical purposes.

Summary and Conclusions

The absorption of plasma protein labelled with radioactive iodine (R.P.P.) from the subcutaneous tissues has been studied in normal subjects and in patients suffering from lymphoedema. A similar investigation was performed in normal rabbits and in rabbits with a limb in which lymphatic transection had been performed.

The behaviour of radioactive plasma protein injected subcutaneously in the limbs of human subjects and animals is consistent with its removal by the lymphatic route.

Detachable direct absorption of protein into the blood stream occurred only after acute traumatic destruction of the lymphatic pathway.

The rate of removal of R.P.P. is slower in patients with severe lymphoedema than in normal subjects.

In normal subjects the rate of removal of R.P.P. is hastened by exercise.

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CASE OF REVERSE COARCTATION SYNDROME

AN AORTIC ARCH SYNDROME

BY

S. N. SEN GUPTA, M.B., M.R.C.P.Ed.

AND

J. C. GHOSH, M.B., D.T.M.

From the Department of Medicine, Calcutta National Medical College Hospital, Calcutta

The interesting clinical syndrome characterized by absence of pulsation in the arteries arising from the arch of the aorta, though very rare, has been reported from time to time. In this condition one or more of these arteries are involved in an obliterative process. A large variety of pathological lesions may be responsible for such arterial occlusion. Of these, a primary panarteritis of unknown aetiology is in a class by itself, and has been variously described as 'Takayasu's disease,' 'pulseless disease,' and 'young female arteritis.'

The largest series of pulseless disease seems to have been reported from Japan, and a comprehensive list of 58 cases was given by Caccamise and Whitman (1952) along with the report of their own case. Skipper and Flint (1952) summarized 16 cases of 'syphilitic' and 'non-syphilitic' origin and recorded two non-syphilitic cases of their own. Ross and McKusick (1953), in an extensive review of the world literature, analysed over 100 cases, 35 being previously unreported. They discussed the whole subject of pulse anomalies in the branches of the aortic arch under the title 'aortic arch syndrome,' a term first suggested by Frovig in 1946. Other possible aetiological factors besides the panarteritis, according to their analysis, are syphilis and trauma, with or without aneurysm formation, chronic dissection of the aorta, atheromatosis, congenital anomalies, mediastinal tumours, and thrombophilia. They concluded that syphilis is the commonest and primary atheroma a very rare cause of aortic arch syndrome.

Recently two more cases of 'pulseless disease' have been reported in the Anglo-American press (Barker and Edwards, 1955; Trias de Bes *et al.*, 1955). From India the first case of aortic arch syndrome was published by Shikhare (1921), and so far as we know the case reported below is the second. Our case is of interest not only because the syndrome itself is rare but also because it belongs to a very rare aetiological variety.

Case Report

A sweeper of a veterinary hospital, aged 51, was admitted to hospital complaining of low pyrexia, breathlessness, productive cough, and generalized body ache of ten days' duration. He had been a chronic sufferer from productive cough for a long time, with occasional exacerbations, especially in the winter months, but his activities had not previously been limited by breathlessness. He had occasional attacks of vertigo, though they were never very severe. A hospital discharge ticket in his possession showed that he was sent home a fortnight previously after successful treatment of a hypopyon ulcer of his right eye.

The patient was thinly built and looked very ill, with sunken eyes and cheeks. Bilateral lenticular opacity and a scar on the lower part of the right cornea were present. The fundus oculi could not be properly visualized because of the lens opacities. The palpebral conjunctiva looked rather pale. He was slightly cyanosed; the respiratory rate was 42 a minute. Clinically the chest was grossly emphysematous. The breath sound was vesicular in character with

prolonged expiration, and plenty of rales and rhonchi were heard all over the chest. The cardiac apex was not palpable and the heart sounds were faintly audible. There was no cardiac murmur. The tender liver was palpable 1 inch (2.5 cm.) below the costal margin, and the caecum was thickened and slightly tender. There was no clinical abnormality in the nervous system.

Pulsation in the radial arteries on either side was completely absent, although the arteries were well palpable at their usual site. Further examination revealed absence of pulsation in the brachial, axillary, subclavian, carotid, and temporal arteries of both sides. The vigorously pulsatile aortic arch was, however, easily palpable at the suprasternal

notch, but its wall seemed to be unusually hard to the touch. There was good pulsation in the abdominal aorta and in all the arteries of the lower limbs. The dorsalis pedis was visibly throbbing. Several small tortuous and pulsatile arteries were also visible over the abdominal and chest walls.

It was obviously impossible to record the blood pressure in the upper limbs. Over the popliteal arteries it was 220/75 mm. Hg. The patient had a temperature of 99.8° F. (37.7° C.) and his pulse rate was 120 a minute. The extremities were warm. The patient was co-operative and was talking intelligently.

On further interrogation he denied any history of severe trauma on the chest. He had never suffered from chest pain or paralytic attacks and firmly denied a past history of syphilis.

Fluoroscopy in the out-patient department showed patchy opacities scattered in both lung fields. The heart size appeared to be within normal limits. The aorta was unfolded and extensively calcified.

Treatment with penicillin and oxygen was immediately started, but the patient died suddenly, rather unexpectedly, within a few hours before complete investigation could be made. The leucocytes numbered 10,600 per c.mm., with 82% neutrophils, and the blood Wassermann reaction was negative. A skiagram taken immediately after death confirmed the fluoroscopic findings. The patient obviously died as a result of bronchopneumonia.

Necropsy Findings

Only a limited post-mortem examination was allowed, and the arch of the aorta with about 1 inch (2.5 cm.) of each of its branches could be removed. A piece of the adjacent part of the right lung was also taken out.

The aortic arch was of cartilaginous consistency and cracked under pressure. When split open its walls were found to be plastered with innumerable calcific plaques, and a big greyish-white friable clot about 5 by 1.5 cm. was found

attached to its upper wall, completely occluding the mouths of all three branches. Less than about two-thirds of the diameter of the arch was patent (see Fig.). There was no aneurysmal dilatation of the arch.

The left common carotid artery was flattened in the antero-posterior direction and its cross-section was oval in shape. The cut ends of both the left common carotid and subclavian arteries were found occluded with a firm greyish-white mass which could not be removed from their lumina. The whole length of both these arteries felt like solid cord. The distal ends of the right common carotid and subclavian arteries were, however, patent, although the innominate artery would not admit a probe. When cut across, the latter was also found stuffed with a firm clot.

Histology.—Examination of sections of the aorta revealed gross atheromatous changes in all stages of development. There were large areas of ulceration and calcification. No cellular infiltration was found in the media or adventitia. The lumen of the left common carotid artery was filled with a mass of connective tissue which was continuous at places with the proliferating cells of the subintimal layer. In the left subclavian artery there was a loose connective-tissue mass with numerous intercommunicating capillaries and scattered areas of fibrous-tissue formation. A similar picture of recanalizing thrombus was found in the lumen of the innominate artery. No pathological change was evident in the walls of any of these arteries. The section of the lung showed areas of consolidation with surrounding emphysema suggestive of a bronchopneumonic lesion.

Discussion

The interesting presenting features in this patient were the absence of pulsation in all the arteries arising from the aortic arch and a high systolic blood pressure in the vigorously pulsatile arteries of the lower limbs, together with evidence of collateral circulation over the abdominal and chest walls. This resembles the clinical syndrome of coarctation of the aorta upside down, and we have preferred to call it a reverse coarctation syndrome. The term "reverse coarctation" has been criticized because the lesion here is not congenital (Barker and Edwards, 1955). This critical consideration does not apply when we mean a clinical complex only. We are, however, conscious that the particular syndrome under consideration has been already described under a large number of inadequate terms, and we may be responsible for suggesting another which is essentially descriptive and not nosological.

Obliteration of pulsation of all the arteries arising from the arch of the aorta is not an uncommon feature of aortic arch syndrome. It was present in 29 of the 40 best-documented cases discussed by Ross and McKusick (1953). Thrombosis occurring independently in any of these arteries, due to primary pathological conditions in their walls, can produce loss of pulsation in individual arteries. One or other of them may also, on rare occasions, be the site of embolic occlusion. In such cases the term "aortic arch syndrome" seems inappropriate. When all the branches of the aortic arch are obliterated the pathological lesion almost invariably involves the aortic arch itself, and there should not be any hesitation in calling it an aortic arch syndrome.

The main symptoms due to loss of cerebral blood supply such as vertigo occurred in 25, syncope in 15, hemiparesis and hemiplegia in 13, and convulsion in 8 of the 40 cases referred to. In our patient there was only a history of occasional vertigo, which was never very severe. His intelligence and memory seemed fairly good. Facial hemiatrophy and atrophy of facial bones with perforation of the nasal septum have been described as trophic changes due to diminished circulation in the face (Harbitz, 1926; Marinesco and Kreindler, 1936). In our patient, sunken eyes and cheeks may indicate atrophy of retro-orbital tissue and facial muscles. But such an appearance is not very uncommon in some otherwise normal persons. Poorly healing infections of face and scalp and premature cataract have also been reported (Ross and McKusick, 1953). The patient had a healed hypopyon ulcer in his right eye. He also had

bilateral cataract, which is by no means uncommon at his age. It is, however, possible that reduced blood supply and age were complementary factors in the opacification of the lenses. Ophthalmoscopic assessment of the condition of retinal blood vessels was not possible in the presence of the cataracts.

A rather intriguing point is how the cerebral blood supply was maintained. Obviously the supply through the left common carotid artery was completely cut off. Although the right common carotid was patent it was not in communication with the main current of aortic blood, since the innominate artery was totally occluded. There was no expansile pulsation in it. We may be justified in assuming that the process of occlusion was slow enough to allow collateral channels to open up adequately through whatever communication was available. The only other arteries that can carry sufficient blood to the brain are the vertebrals. The left subclavian artery was completely occluded as far as it could be removed. Whether the thrombus extended and involved the left vertebral artery is unfortunately not known. The lumen of the right subclavian artery was patent. It seems reasonable to conclude, in spite of the small material evidence, that anastomotic channels were established between the intercostals, internal mammary, and lateral thoracic arteries which fed the right subclavian, common carotid, and vertebral arteries.

The presence of tortuous pulsatile arteries on the chest wall evidently lends further support to such a conclusion. From the existence of similar arteries on the abdominal wall it appears that these collaterals were also reinforced with a supply from the external iliac and femoral arteries. While this can also explain the circulation in the right upper limb, it is not easy to suggest any possible channel of supply to the left, which was also apparently well nourished and warm. It would certainly be interesting to know if the left vertebral artery, and the subclavian artery beyond it, were patent.

The popliteal blood pressure was recorded with a standard arm cuff, and due allowance for this should be made. But the high pulse pressure is striking. It is probably a common expression of the extensive collateral anastomosis and atherosclerosis.

Unfortunately a complete post-mortem examination was not possible. In compliance with the sentiments of the patient's relations we were obliged to content ourselves with removing only the aortic arch with the proximal parts of its branches and a small piece of the right lung. From the clinical and fluoroscopic findings it was thought that the mouths of the three branches were blocked with atheromatous lesions, which are known to occur predominantly round the ostia of aortic branches. The necropsy findings of the big ante-mortem thrombus were very much unexpected. Thrombus formation in an aneurysmal sac is not very uncommon. But in its absence it is surprising how such a big clot can form in the arch where the turbulence of blood is very great indeed. Ross and McKusick (1953) found only three examples in 44 patients, and in all of them the thrombus was in syphilitic aneurysmal sacs.

In our case the histological picture of an atheroma of the aorta was unmistakable. Primary atheroma of the aortic arch is rare. Ross and McKusick think that trauma may play some part in the pathogenesis of atheroma in the arch. Our patient gave no history of physical violence on the chest, at least of any severe degree which he could remember. Atheromatous changes can occur on pre-existing syphilitic aortitis. There were no pathological changes suggestive of such a lesion in the media and adventitia. Furthermore, there was no history of syphilis and the blood Wassermann reaction was negative. The only abnormality found in the left subclavian, left common carotid, and innominate arteries was the presence in them of organizing and recanalizing thrombi. There was absolutely no pathological lesion in their walls. It may naturally be concluded that the primary atheromatous ulcer on the wall of the aortic arch helped in the formation of the thrombus which gradually increased in size and extended into all the three branches of the arch of the aorta.

Summary

A case of aortic arch syndrome characterized by absence of pulsation in all the arteries of the upper limbs and neck, with high systolic blood pressure in the vigorously pulsatile arteries of the lower limbs and evidence of collateral circulation on the abdominal and chest walls, is described. From its resemblance to the clinical syndrome of coarctation of the aorta it has been called reverse coarctation syndrome. The patient died as a result of associated bronchopneumonia.

The patient had very few symptoms and physical signs referable to impaired blood supply to the head and neck and to the upper limbs. The possible collateral channel of supply is discussed.

Only a limited necropsy study was possible. There was extensive atheroma with calcification in the wall of the aortic arch. The upper third of its lumen was occupied by an ante-mortem thrombus which extended into and completely obliterated the lumina of the innominate artery, and the left common carotid and subclavian arteries as far as they could be removed. The right common carotid and subclavian arteries were patent. Histologically, organizing and recanalizing thrombi were found in the occluded arteries. Except atheroma in the wall of the aortic arch no other pathological lesion was found in the walls of any of the arteries.

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INTERMITTENT HYDRARTHROSIS

BY

S. MATTINGLY, M.B., M.R.C.P., D.Phys.Med.

Late Senior Registrar, Department of Physical Medicine and Rheumatism, the Middlesex Hospital

"Of periodicity in disease, no more striking example can be quoted than the recurrent effusion into joints, with little or no febrile disturbance or local signs of inflammation, which goes by the name of intermittent hydrarthrosis."

Garrod (1910).

Intermittent hydrarthrosis is a condition of unknown aetiology characterized by transient joint effusions which recur at regular intervals; one or both knee-joints are usually affected, but occasionally other joints such as the hip, ankle, or elbow are involved (Ragan, 1949). The periodicity of the effusions was first noted in 1852 by C. H. Moore (1864), a surgeon at the Middlesex Hospital.

The onset is often sudden, the effusion lasting two to six days and recurring in cycles of 7 to 11 days, though extremes of 3 and 30 days have been reported. The duration of joint swelling and the length of each cycle