CASE REPORTS

THROMBO-OBLITERATIVE VASCULAR DISEASE OF EARLY ONSET AND LONG DURATION

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The case reported below is a woman who had thrombotic obliteration of the abdominal aorta, the renal arteries, and the right subclavian artery. It seems worthy of recording in view of the early onset and long duration.

Case Report

A Chinese woman of 26 was admitted to hospital on December 28, 1960, for investigation because of persistent gross hypertension. She had first entered another hospital complaining of pain in the right lower quadrant of the abdomen for three days, a condition diagnosed as appendicitis for which appendicectomy was performed.

The patient stated that early in her childhood, at the age of 5 years, she started to have severe pain in the right leg on exertion, which could be relieved by taking a rest. As she grew up, she noticed that her right leg became progressively smaller and weaker than the left leg. In recent years, she could walk for only about 300 yards without pain, especially when she had to carry weight on her shoulders. She occasionally had headaches which were not associated with nausea, vomiting, or blurring of vision. She did not complain of palpitation and dyspnoea on exertion, and she never had any loin pain or hæmaturia. Menarche was at the age of 14 and subsequent periods were regular. She was married at the age of 21 but remained sterile.

On physical examination she was small in stature [height 5 ft. 2 in. (157 cm.) and weight 92 lb. (41.7 kg.)] The right lower limb was 1 in. (2.5 cm.) shorter than the left. The circumferences of the right thigh and leg measured at 4 in. (10 cm.) above and below the knee were both 1 in. (2.5 cm.) less than those of the left. All muscles of the right lower limb were weak in power but the knee and the ankle jerks were present. No sensory changes were detected. The skin temperature on the right was cold from below the knee but colour remained normal after rising and standing and there were no trophic changes in the toes. The femoral, popliteal, medial malleolar, and dorsalis pedis pulses in the right leg were all absent. The right radial pulse was weaker than the left. Blood pressure recorded in the right arm was 210/120 and that in the left 260/160 mm. Hg. The apex beat of the heart was not displaced. Heart rate was 90, rhythm was regular, and heart sounds were normal. A systolic bruit was distinctly audible to the left and just above and below the umbilicus. Another systolic bruit was heard in the right supraclavicular fossa. Examination of the fundi revealed normal discs, narrow retinal arteries, and exudates and hæmorrhages in both.

Blood picture was normal, and urine normal. Blood urea, 33 mg.; cholesterol, 165 mg.

Electrocardiogram showed evidence of a vertical heart with left ventricular hypertrophy.

The following findings were revealed by radiological examination.

Chest. The lungs were clear, heart size and shape were normal, calcifications were present in the right apex, which on fluoroscopy were seen to be extrapulmonary, and their situation and relation to the clavicle indicated that they were in line with the course of the right subclavian artery. No calcification was seen in the aortic arch or the thoracic aorta.

Abdomen (Fig. 1). Calcification was present along the left border of the 2nd and 3rd lumbar vertebrae and on either side of the lower margin of the 1st lumbar vertebra. Intravenous pyelogram showed normal dye excretion in both kidneys and confirmed that the areas of calcification on either side of the 1st lumbar vertebra were in the hilar region of the kidneys.

FIG. 1.—Radiograph of abdomen showing calcium in the hilum of each kidney, more in the right.

FIG. 2.—Percutaneous aortogram. Right common iliac artery not visualized. Right renal artery small with constriction near its origin. Left renal artery also showed local constriction. Both vessels showed calcification.

Percutaneous aortogram through the left femoral artery (Fig. 2) revealed that the aorta was well filled and showed intramural calcification at the level of the 2nd and 3rd lumbar vertebrae. The right common iliac artery was not visualized. The left common iliac artery and its branches were normal. The right renal artery was small and became constricted 2 cm. from its origin, followed by a post-stenotic dilatation heavily encrusted with calcified plaques. After dividing, the branches were small in calibre with arborizations normally distributed. The left renal artery was also constricted 1 cm. from its origin and beyond the post-stenotic dilatation, aneurysms outlined by calcification were present before the artery divided into two main branches of normal calibre. The nephrogram showed a smaller right kidney measuring 9 cm. by 4 cm. and a left kidney of 13 cm. by 5 cm.

In view of the bilateral renal involvement the case was considered unsuitable for surgical treatment. The patient was put on guanethidine sulphate, and her blood pressure was reduced from 260/160 to 170/100 mm. Hg in the left arm by a daily oral dose of 50 mg. The fundi showed remarkable improvement with complete resolution of the exudates and hæmorrhages three months later.

Discussion

The extensive involvement of the vascular system in this case raises the question as to whether it is an example of Leriche's syndrome (Leriche, 1923) or Takayashu's syndrome (Takayashu, 1908) especially as the cause is unknown. In Leriche's syndrome, atheroma of the abdominal aorta usually begins in one iliac artery, extends towards the bifurcation, and then invades both the aorta and the opposite common iliac artery (Leriche and Morel, 1948). In Takayashu's syndrome, the aortic arch is involved in a chronic progressive inflammatory process which affects all three layers of the proximal segments of the innominate, subclavian, and carotid arteries. It usually occurs in young women and has been known to affect other nationals besides the Japanese, including the Chinese (Chang, Chang, and Chiu, 1955). These two entities are therefore regarded as different diseases. However, the thoracic and the abdominal aorta may rarely be involved in Takayashu's syndrome (Ask-Upmark and Fajers, 1956; Correa and Araújo, 1958; Danaraj and Ong, 1959). When the origin of the

renal artery is involved in either of the two diseases, hypertension may develop as the result of the Goldblatt effect.

DeBakey, Creech, and Cooley (1954) stated that patients suffering from Leriche's syndrome were generally a decade younger than those affected by peripheral vascular disease. The youngest in their series of 22 patients was 33 years; among 32 patients reported by Starer and Sutton (1958) the youngest was 37; and in another series of 35 patients (Beaconsfield and Kunlin, 1953) the youngest was 27 years. However, Leriche's syndrome has been reported at all ages, even though rarely in infancy and childhood. Rothstein (1935), for example, reviewed 12 patients between the ages of 2 weeks and 12 years. Panarteritis of the aorta also rarely affects children as in the two cases of Danaraj and Ong (1959), whose ages were 7 and 13 years respectively. In the case reported the thrombo-obliterative process in the abdominal aorta apparently started when the patient was only 5 years old, at which age she first experienced intermittent claudication. The stunting of the right lower limb in length as well as in girth is consistent with long-standing inadequate blood supply dating back to the growing period.

At the time when the patient was investigated she was 26 years old, 21 years after the onset of symptoms. While the insidious nature of Leriche's syndrome is well recognized as a characteristic of the disease, the long duration of symptoms in this case is comparable only with that of one case in the series of Beaconsfield and Kunlin (1953) who also had symptoms at 21 years. In other reports durations of symptoms of only 12 years (DeBakey *et al.*, 1954) and 13 years (Starer and Sutton, 1958) were recorded. The duration of symptoms in Takayashu's syndrome ranges from 6 months to 14 years (Gibbons and King, 1957), and the average time from onset of symptoms to the time of reporting or to death was 5.1 years in 26 cases reviewed by Ask-Upmark (1954).

Summary

A young Chinese woman of 26 was found to have malignant hypertension, and aortography showed slight narrowing of the abdominal aorta, complete occlusion of the right iliac artery, partial occlusion of the right and left renal arteries, and aneurysms in the left renal artery. Calcification of the right subclavian artery was also demonstrated radiologically. Symptoms of arterial insufficiency in the right leg started at the age of 5 years. The hypertension responded to treatment by guanethidine sulphate.

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