and no mural thrombus was present. Both internal carotid and vertebral arteries were removed in their entirety. A saccular aneurysm of the right internal carotid sinus 2.5 by 1.5 cm. was found (see Fig.). Sections through the aneurysm showed that the lumen was completely obliterated by laminated thrombus; microscopic examination of the wall of the aneurysm revealed atheromatous thickening and

Close-up of right carotid bifurcation, showing saccular aneurysm of right carotid sinus.

areas of thinning. The thrombus varied considerably in structure, some areas being organized while others appeared to be recently laid down. The innominate artery and right carotid artery proximal and distal to the aneurysm showed no luminal narrowing, and no stenosis was found at any point in the left carotid system. Neither subclavian artery showed narrowing, but both vertebral arteries were severely stenosed at their origins, less than half the diameter of the original lumen being preserved. No other sites of thrombosis and no ulcerating atheromatous lesions were seen at any point in the carotid and vertebral systems.

Brain.—The right cerebral hemisphere was softened and swollen, with grooving of the right uncus against the tentorium cerebelli. The large arteries at the base of the brain were affected by severe atheroma, which was also present in their smaller branches within the cerebral sulci. Despite this atheroma all the major arterial lumina were patent save that of the right middle cerebral artery, which was partially blocked. Sections showed organizing thrombus. Coronal slices showed that the enlarged right hemisphere was soft in the region supplied by the right middle cerebral artery. The cortex of this area was hyperaemic and also haemorrhagic anteriorly at the boundary with the territory of the anterior cerebral artery. Sections showed changes of recent infarction. Elsewhere in both hemispheres there were numerous older softened lesions ranging from 1 to 4 mm. in diameter, and these were seen in the putamen and caudate nucleus on the left as well as in the white matter. An old yellowish destructive lesion was found in the white matter of the left middle temporal gyrus and on section proved to be a sequel of an old haemorrhage. Recent haemorrhages of various sizes were seen in the pons. The medulla and cerebellum were normal.

DISCUSSION

This is the only atheromatous aneurysm of the carotid sinus which we have found in 500 necropsies where the neck arteries have been dissected for study, and we

have been unable to find any description of a similar case in the literature. Although the condition is uncommon, this case does raise several points of general interest.

We feel that the recurrent transient episodes can be reasonably attributed to emboli from the grossly aneurysmal right carotid sinus, and it is possible that a similar mechanism might be responsible for transient attacks in the absence of an aneurysm, by embolization of thrombus from the surface of the atheromatous lesions commonly found in the carotid sinus, or of atheromatous debris from ulcerated plaques. We have, however, failed to find any correlation between ulceration in the carotid and vertebral arteries and transient cerebral episodes (Schwartz and Mitchell, 1961).

The distribution of the brain lesions is of interest, for although the aneurysm was on the right side many of the attacks suggested involvement of the left hemisphere, and old infarcts were found in both hemispheres. If the lesions were caused by emboli we must account for this bilateral distribution. Dickinson (1961) has shown that in the presence of severe neck-artery disease the relationship between the arterial and cerebral lesions is unpredictable, and it seems likely that in our patient, who had very severe narrowing of both vertebral arteries as well as the right carotid, the flow pattern in the circle of Willis would be abnormal. Under these circumstances we feel that emboli from one carotid artery might easily find their way into any of the branches of the circle. The aneurysm was so circumscribed that had carotid angiography been undertaken during the phase of transient episodes, and the lesion outlined, reconstructive surgery might have prevented the final thrombotic episode.

We are grateful to Dr. E. M. Buzzard for permission to report this case, and to Dr. M. J. V. Bull, the patient's general practitioner, for information about the past history.

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Aortic Arch Syndrome and Polymyositis with L.E. Cells in Peripheral Blood

Absence of pulses in the upper limbs may be due to a number of causes (Ross and McKusick, 1953), and Lessof and Glynn (1959) pointed out that not all forms of arteritis of medium and large-sized arteries correspond exactly to those described by Takayasu (1908).

The following is the report of a case of polymyositis in which the arteries of the upper and lower limbs were involved by some form of occlusive disease, and in which lupus erythematosus (L.E.) cells were demonstrated.

CASE REPORT

A housewife of 54 was admitted to hospital on November 17, 1960. Her illness began in June, 1959, with aching in both thighs similar to that experienced after unaccustomed exercise. The pain was constant even at rest. Later she developed similar pains in the back of the neck and in both shoulders. The muscles in the affected areas felt tender, and she found it difficult to raise her arms above her head. In June, 1960, she developed claudication in the calves, the right forearm, and to a less extent in the left forearm. She had never experienced angina.

The brachial and radial pulses were absent on the right and weak on the left. Pulsation in the carotid arteries was normal on the right, but possibly diminished on the left. All pulses in both legs below the femorals were absent. There was a systolic murmur over the origin of both subclavian arteries. The right hand exhibited Raynaud's phenomenon. Blood-pressure in the left arm was 120/70 and unrecordable in the right arm. Oscillometry of her right arm showed no pulsations, and the pulses in the left arm were probably reduced, as they were below the knees. Blood-pressure taken by an indwelling needle (right femoral artery) was 200/75.

The spleen tip was palpable. The shoulder- and hip-girdle muscles were tender. There was no obvious wasting, but the affected muscles appeared weak. Limb reflexes were very brisk but equal on both sides. Visual acuity in the right eye was only 6/60 owing to a refractive error, but was 6/9 in the left eye. Funduscopic examination was negative apart from a right retinal cyst. There were no cataracts.

Investigations.—E.S.R. 80 mm. in one hour, haemoglobin 70%, white blood count 7,400, differential count normal. Serum creatine 1.2 mg./100 ml. and urine creatine 41 mg./24 hours. W.R. and R.P.C.F.T. negative. Blood urea 48 mg./100 ml. Serum protein electrophoresis, normal pattern. Serum cholesterol 210 mg./100 ml. An electromyogram showed only minor abnormalities (low-amplitude polyphasic pattern) in the left biceps and left rectus femoris. A muscle biopsy (right deltoid) showed a diffuse variation in muscle-fibre calibre, some appearing moderately atrophic. Occasional fibres were undergoing necrosis and phagocytosis. There was a slight increase in endomysial fibrous and fatty tissue. Collections of small round cells were seen, which appeared related to the walls of blood vessels. There was a generalized increase in interstitial cells. Biopsy of the left temporal artery showed some increase in intimal fibrous tissue but no giant cells. An E.C.G. showed changes compatible with postero-lateral ischaemia. Chest x-ray examination showed nothing abnormal. Several L.E. cells were seen in one sample of peripheral blood, and some cells described as "suggestive" were previously seen elsewhere. The antinuclear factor was negative. (This last investigation was carried out three weeks after starting steroid therapy.)

Aortography showed that the right subclavian artery terminated just distal to the origin of the right vertebral artery, the right arm receiving its arterial supply through periscapular vessels. The left subclavian artery narrowed markedly just beyond the origin of the vertebral artery and the distal portion of the subclavian artery, and that portion of the brachial artery included on the films showed pronounced narrowing and irregularity of the lumen. There was slight narrowing of the origin of the right renal artery and a scar at the lower pole of the right kidney. Gross irregularity with narrowing of the lumen was present in both femoral arteries.

As there was evidence of a myositis, it was decided to try corticosteroids. Prednisone 10 mg. t.d.s. was started; this dosage was subsequently reduced to 5 mg. b.d. The patient soon lost the aching and stiffness of muscles, and there was great improvement in power. However, the claudication of her right forearm remained. The right subclavian artery was therefore explored by Professor J. B.

Kinmonth. There was no pulsation beyond the second part. The artery was opened and was found to be obliterated by old organized thrombus. This thrombus was removed, and some recent thrombus distal to the block was removed by suction.

The tissue removed consisted of parts of an organizing thrombus and parts of an artery wall, the intima of which was fibrotic and showed attached thrombus. There was focal chronic inflammatory cell infiltration of the intima. No haematoxyphil bodies were seen. The small amount of media included in the sections was normal. Post-operatively the radial pulse did not return, but the patient noticed a marked improvement in the ischaemic symptoms in the right arm.

COMMENT

The cause of the vascular lesions in this case is uncertain; atheroma (Shucksmith, 1959) is possible. The histological appearance of the limited amount of tissue removed from the site of the subclavian block was compatible with, but not diagnostic of, atheroma. On the other hand, the evidence of myositis, and its response to steroids, the raised E.S.R., the splenomegaly, and the L.E. cells suggested the presence of a generalized connective-tissue disease, and it is possible that the arterial lesions were also part of this process.

Polymyositis can occur in disseminated lupus erythematosus (Keil, 1940), and vascular lesions and Raynaud's phenomenon are not uncommon. The involvement of major arteries, however, is rare, though Keat and Shore (1958) describe a case of disseminated lupus erythematosus in which there was gangrene of both legs due to involvement of the anterior and posterior tibial arteries. Lessof and Glynn (1959), discussing their third case of "pulseless disease," state that they know of no case of disseminated lupus erythematosus previously described with lesions of the major vessels arising from the aortic arch. In their case pulses were absent in both arms and there was evidence of diminished blood flow in the left foot. L.E. cells were present and the antinuclear factor was positive. In the present case, despite the presence of L.E. cells, the picture was atypical of disseminated lupus erythematosus in view of the negative antinuclear factor (Weir *et al.*, 1961).

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