FIG. 1.—Case 1. Lower end of left internal carotid artery dissection between adventitia below and media above. Media contains patchy islands of elastic tissue. (Elastic van Gieson. $\times 45$.)

FIG. 2.—Appearance of the internal carotid artery dissections in Case 2 (A) and Case 3 (B) on angiography. There is a tapering defect in Case 2 and an irregular defect in Case 3 (arrows).

FIG. 3.—Case 2. Upper end of dissection of right internal carotid artery between media above and adventitia below. (Haematoxylin and eosin. $\times 35$.)

FIG. 4.—Case 2. Showing cystic collections of ground substance in the media of the aorta. (Haematoxylin and eosin. $\times 35$.)

FIG. 5.—Case 3. Lower end of left internal carotid artery dissection between media above and adventitia below. There is an irregular distribution of elastic tissue in the media. (Elastic van Gieson. $\times 35$.)

FIG. 6.—Case 3. Thrombus in lumen of left middle cerebral artery is seen above, and below this blood is dissecting through and destroying the media of the artery. (Haematoxylin and eosin. $\times 100$.)

FIG. 7.—Case 3. Renal artery. The lumen is above and adventitia below. Patchy, irregular elastic tissue is present in the intervening media. (Elastic van Gieson. $\times 50$.)

cytes, leucocytes, and platelets from a normal or hyperplastic bone-marrow, but the relief produced in the cases described in this paper is far greater than could be accounted for merely by the removal of a heavy mass from the abdomen.

With regard to the question of hypersplenism, there is very little evidence to suggest this in any of these five cases. Most had slight anaemia, all had a moderate neutropenia before splenectomy, but this persisted for varying periods after splenectomy, in one case for as long as nine years. Platelets in the circulating blood were normal in four out of the five cases, and the usual rise of platelets after splenectomy occurred in all cases.

This condition, here called "giant follicle lymphoma of the spleen," is certainly rare. A sixth case with exactly similar features has been seen through the courtesy of Dr. R. J. McGill, of the Royal South Hants Hospital. Insufficient time has elapsed since splenectomy to judge the effect.

If bone-marrow biopsy was performed more often in cases of "aleukaemic lymphatic leukaemia" with large spleens, more cases might be discovered.

Summary

A description is given of five cases with massive splenomegaly, all of which presented features of a lymphoma very similar to that of lymphatic leukaemia except that the bone-marrow contained localized masses of lymphocytes instead of generalized infiltration. All were apparently cured by splenectomy.

Acknowledgments were made in the previous paper for help in the first four cases. The fifth case was under my own care at Charing Cross Hospital, and the splenectomy and liver biopsy were carried out by Mr. Norman Tanner. Dr. T. C. Studdert, of the Cumberland Infirmary, Carlisle, gave me information about a previous illness of one patient (Case 5) in which the spleen was found to be enlarged.

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Spontaneous Dissecting Aneurysms of the Cervical Internal Carotid Artery

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[WITH SPECIAL PLATE]

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Occlusion of the internal carotid artery just beyond its origin in the neck is a not uncommon finding at necropsy. Hultquist (1942) observed it in 4.4% of a series of 1,300 routine necropsies. In most cases the occlusion is caused by thrombosis superimposed on atheromatous degeneration of the artery concerned, and the condition is therefore much commoner in the older age-groups. As a rarer occurrence it is seen in individuals below the age of 40. This arbitrary division into these two age-groups can be made, as the pathogenesis of the condition seems to be quite different in the two groups.

The causes of obstruction of the internal carotid artery in this younger category have been adequately reviewed by Humphrey and Newton (1960). They found that in over half the published cases no adequate cause could be determined. Approximately half of the remainder were attributed to recent trauma, which has been discussed by Therkelsen and Hornnes (1963). Other possible causes mentioned being cerebral aneurysm and subarachnoid haemorrhage, hypertension, blood dyscrasias, chronic infection such as rheumatoid arthritis and rheumatic fever, neoplasms, cervical ribs, and the post-partum state. Recently an arteritis of the internal carotid has been suggested (Shillito, 1963). Atheromatous occlusion appears rarely in this age-group.

In the past five years 115 patients were found to have occlusion of the internal carotid artery in the neck. Ten of these patients were below the age of 40. Of these 10, three were related to recent cerebral trauma, two were associated with chronic infective processes elsewhere in the body, one followed a severe attack of epilepsy, and in one no associated

factor was observed. The purpose of this report is to put on record the remaining three cases in which a spontaneous dissecting aneurysm was responsible for sudden partial or complete occlusion of the lumen of the internal carotid artery near its origin.

Case 1

A housewife aged 40 complained of a generalized headache for 12 hours and then suddenly developed a complete aphasia and right hemiplegia. Clinical examination revealed a drowsy individual, completely aphasic, with a right homonymous hemianopsia and a right hemiplegia. She was normotensive and apart from a systolic murmur over the cardiac apex there were no other signs of systemic disease. Her level of consciousness deteriorated. Angiography demonstrated a complete obstruction of the left internal carotid artery 1.5 cm. above its origin, no contrast medium entering the cranial cavity. It was noted that the pineal gland, which was calcified, lay to the right of the midline. Her condition continued to deteriorate and she died some 40 hours after the onset of symptoms.

Necropsy revealed no significant abnormality in the bodily organs. There was no myocardial hypertrophy and the important arteries showed only slight atheroma, with a few linear lipid plaques in both common carotid arteries. The left internal carotid artery was occluded by a dissecting aneurysm of the posterior wall which began 1 cm. above its origin and extended up to the base of the skull. A small intimal tear was present 4 mm. above the lower limit of the dissection. There were only a few small lipid plaques at the origin of the left internal carotid artery, below the dissection. The angiography puncture holes were 4 cm. lower down in the left common carotid artery, well clear of the dissection.

Examination of the decalcified skull base showed that the dissection ceased abruptly at the beginning of the bony carotid canal. Above this the lumen of the left internal carotid artery and the origins of all its branches contained fluid blood, with the exception

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of the three large branches of the left middle cerebral artery, which were occluded by ante-mortem thrombus. There was massive, recent pale infarction of the cortical and ganglionic distribution of the left middle cerebral artery.

Histological examination of the left internal carotid artery showed the dissection of recent blood in a plane between the media and adventitia (Special Plate, Fig. 1). There was cystic medial degeneration of the media of the vessel both at and below the level of the dissection. A similar appearance was seen in the thoracic aorta. The medial cysts contained material staining bright green with Alcian green and probably representing an excess of mucopolysaccharides. There was no abnormal vascularization of the media. In addition to these changes in the ground substance of the media of the left internal carotid artery and aorta, the left internal carotid showed a patchy distribution of elastic fibres in the media. This arrangement of the elastic tissue at the origin of the internal carotid arteries was also seen in the two subsequent cases. Control material from necropsies on subjects of similar age with apparently normal carotid arteries and without hypertension was examined, and it was found that the arrangement of the elastic fibres in the media of the origin of the internal carotid arteries was identical to that present in the three cases of dissecting aneurysm. It was concluded that this appearance represented the usual transition between the large elastic arteries and the muscular arteries, although Anderson and Schechter (1959) thought it to be pathological.

Case 2

A man aged 35 complained of an attack of blurred vision in the right eye and pain in the right cheek some 12 hours prior to sudden loss of consciousness. He was found to be stuporous, with the eyes deviated towards the right and a left hemiplegia, hemihypoaesthesia, and hemianopsia to menace. He was normotensive with no other signs of disease. Lumbar puncture revealed a clear fluid under increased pressure containing 70 mg. of protein per 100 ml., no cells being seen. A right carotid arteriogram revealed a severe stenosis of the internal carotid artery in the neck some 2 cm. long (Special Plate, Fig. 2 A). Contrast material entered the intracranial cavity but the middle cerebral vessels did not fill. There was evidence of displacement of the midline structures to the left, and air studies confirmed the presence of generalized swelling of the right hemisphere. His condition deteriorated and he died before further treatment could be carried out.

Necropsy showed some early hypertrophy of the wall of the left cardiac ventricle which averaged 19 mm. in thickness, the heart weighing 390 g. There was minimal atheroma of the large arteries of the body. A few small lipid plaques of atheroma were present at the bifurcation of both common carotid arteries and in the first centimetre of both internal carotid arteries. The angiography puncture holes in the right common carotid artery lay over 4 cm. below the origin of the internal carotid artery. There was a 1 by 1.5 cm. recent dissection of blood into the posterior wall of the origin of the right internal carotid artery. This had a smooth intimal surface with no evidence of mural thrombus on or above it. As in Case 1, the blood in the internal carotid artery and its branches was fluid, except for the large branches of the right middle cerebral artery, which were occluded by ante-mortem thrombus.

There was massive, mainly pale, recent infarction in the cortical distribution of the right middle cerebral artery, overlapping into the parasagittal portion of the anterior cerebral cortical distribution on that side. Secondary haemorrhage and necrosis were present in the right thalamus, occipital lobe, and mid-brain.

Histological examination showed the dissection in the right internal carotid to lie between the media and adventitia, as in Case 1 (Special Plate, Fig. 3). Similarly there was a cystic change in the media, with the cysts containing material that stained strongly with Alcian green. This was present in the right internal carotid artery both above and below the dissection and was especially prominent in the thoracic aorta (Special Plate, Fig. 4).

Case 3

A housewife aged 38 complained of left-sided headaches and ear pain. Two days later she was found unconscious and on arrival at hospital was drowsy with a right hemiparesis. Her condition deteriorated, she developed a fixed dilated pupil on the right side and

a flaccid right hemiplegia. She was normotensive with no other signs of systemic disease. Lumbar puncture revealed a fluid containing a normal level of protein and no cells. Left carotid angiography showed an irregular filling defect in the left internal carotid artery (Special Plate, Fig. 2 B). Air studies demonstrated gross generalized swelling of the left hemisphere. The patient's condition continued to deteriorate despite intensive dehydration therapy and she died less than 24 hours after the ictus.

Necropsy showed no significant abnormalities in the body organs except for dense, fibrous pleural adhesions over the right lung, which itself appeared normal. The left cardiac ventricle was not hypertrophied. There was non-stenotic, fibro-lipid atheroma in the carotid sinus on both sides, but atheroma was not present in either common or internal carotid artery. The lumen was obliterated 2.5 cm. above the origin of the left internal carotid artery by a recent dissection of blood into the wall of the artery, which ascended to the base of the skull. When this was decalcified and cut, the dissection was seen to end at this point, but the lumen of the internal carotid artery was occluded by recent ante-mortem thrombus extending up into the left middle cerebral artery and its main branches. There was almost total pale infarction of the cortical and ganglionic distribution of the left middle cerebral artery.

Histological examination of the origin of the left internal carotid artery showed the dissection to be in a plane through the deeper or more external layers of the media (Special Plate, Fig. 5). As in the two previous cases there was an apparent excess of mucopolysaccharides in cystic collections in the media of the artery. This was also the case with the comparable portion of the right internal carotid artery, the thoracic aorta, and both renal arteries. Histological examination of the thrombosed left middle cerebral artery showed that there was in fact recent dissected blood through the deeper layers of its media in addition to the recent thrombus in the lumen (Special Plate, Fig. 6).

Discussion

From a clinical standpoint there is little outstanding in these three case histories. Careful inquiry of the referring hospitals and the patients' general medical practitioners failed to reveal any history of recent injury, migraine, epilepsy, or chronic infective disorders. One patient (Case 3) showed old healed pulmonary tuberculosis which had been treated over 20 years earlier. All three patients presented with a sudden ictus following 12 to 48 hours of headache. One patient (Case 2) gave a story of a characteristic symptom of internal carotid occlusion, that of blurring of vision in one eye. All three patients on clinical examination presented evidence of a gross lesion affecting the appropriate cerebral hemisphere. Examination of the neck revealed no striking signs to suggest the cause of the cerebral lesion.

Radiologically a tapering type of filling defect on angiography appeared to be the characteristic finding, although this is commonly seen in patients with atheromatous lesions. A striking picture was that of an irregular wavy defect suggesting folding of the separated intima within the lumen of the vessel (Fig. 2), and this appearance has been described in dissecting aneurysms occurring within the cranial cavity (Spudis *et al.*, 1962). In these instances the appearance was likened to "saw teeth." The displacement of the midline structures, radiologically, merely reflects the severity of the cerebral infarcts responsible for the patient's death.

The aorta is the usual site of dissecting aneurysms and these lesions may well extend into the major branches of this vessel. However, although rare, these lesions do occur in peripheral vessels, some 48 instances having been reported in the English literature in the past 40 years (Foord and Lewis, 1959). Of these peripheral vessels the carotid circulation appears the most commonly involved, although many of the reports include cases almost certainly due to angiographic needle puncture (Crawford, 1956) and trauma to the neck. The renal and coronary vessels are also infrequently the site of this condition. In each case the symptomatology appears to be that of sudden vascular insufficiency in the part of the body supplied by the affected artery.

Of the dissections occurring in the carotid and vertebral arterial tree the majority occur peripherally within the cranial cavity, the middle cerebral being the most often involved (Mair, 1952; Norman and Ulrich, 1957); another common site being the basilar artery (Wolman, 1959). On the other hand, dissecting aneurysms involving the proximal arteries in the neck are rare. Only one previous case of a spontaneous aneurysm similar to the present three cases has been found (Anderson and Schechter, 1959). Two other cases were due to trauma (Northcroft and Morgan, 1945) and hypoplasia of the vessel concerned (Jentzer, 1954). In the case described by Anderson and Schechter a 41-year-old man rapidly developed a progressive unilateral cerebral infarction and was found at necropsy to have a long stenosis of the internal carotid artery from a dissecting aneurysm. They concluded that medial degeneration, such as that described by others in the aorta and in common carotid dissection in Marfan's syndrome (Austin and Schaefer, 1957), was the cause of the aneurysm in their case and commented on the fact that their patient had suffered from long-standing cavitating tuberculosis. Although one patient in the present small group had had tuberculosis this had completely healed some 20 years previously.

Various conditions have been cited as a causative factor in the production of intracranial dissecting aneurysms. Trauma seems to have played a part in many. Wolman (1959) thinks that they are due to congenital medial defects such as are thought to give rise to congenital berry aneurysms, but he seems alone in this view. Syphilitic arteritis has been found in two cases. Migraine also has been described as occurring with this condition on two occasions. Medial degeneration has been described pathologically in three cases. It has been suggested that haemorrhage from vasa vasorum might account for sub-intimal dissection in the cerebral arteries (Nedwich *et al.*, 1963). However, vasa vasorum have not been demonstrated in the media or intima of cerebral arteries in the absence of atheroma or other arterial disease, so that this explanation is unlikely. Whatever the cause may be in the more distal vessels there seems little doubt that the few spontaneous dissecting aneurysms which have occurred in the large arteries of the body have been associated with changes in the media of both the vessels concerned and those elsewhere. It may be that the process affecting the larger arteries of the body is different from that occurring in the smaller arteries containing more smooth muscle and less elastic tissue. This view is suggested by Foord and Lewis (1959).

What may be referred to as the "transitional" distribution of medial elastic tissue is found both at the origin of the internal carotid artery and in the renal artery (Special Plate, Fig. 7). This was demonstrated in the three present cases and in controls. The coronary arteries are also muscular arteries arising from an elastic artery, and the reported cases of dissecting aneurysms at these three sites suggest that this transition may predispose to or predetermine the likely site of dissection in the presence of some other causative factor such as trauma or cystic medial degeneration as in the present three cases. However, Wright (1963) has shown that an excess of acid mucopolysaccharide is not an uncommon finding in the arteries of relatively young people. He found it in the renal and common carotid arteries of apparently normal individuals between the ages of 30 and 40. Thus perhaps too much emphasis should not be placed on this factor until more is known about its frequency and its relation to dissecting aneurysms.

Other points of interest in these three cases are the fact that the dissection in Case 1 ceased at the entrance to the bony carotid canal, suggesting that the rigid restriction of the canal

prevented further dissection. However, this was not so in Case 3, where dissection was present in the middle cerebral artery, for it seems unlikely that this was a separate dissection from that in the internal carotid. There is no evidence in any of the three cases that the thrombus present in the middle cerebral artery was embolic, and the smooth intimal surface of a recent dissection with intact endothelium is unlikely to be the source of emboli. In Case 3 the thrombus was continuous from carotid into middle cerebral and was probably propagated. In Cases 1 and 2 it may represent stasis, occurring some time after the carotid lesion and occurring in those arterial branches furthest away from any collateral supply.

It seems unlikely that any form of treatment would have been of any avail in the three cases described. It is possible that if this condition were recognized at an early stage by the presence of a localized tapering or irregular defect of the internal carotid artery on angiography, immediate thromboendarterectomy might affect the course of other patients. It is also suggested that this condition is commoner than reports would suggest, and that it may account for an appreciable proportion of sudden obstructions of the internal carotid artery in the neck in patients of the younger age-group.

Summary

Three cases of spontaneous dissecting aneurysms of the internal carotid artery are described. The important points common to these three patients were: (1) All were in the fourth decade of life and came to necropsy within six months of each other. (2) There was no history or necropsy evidence of trauma to the head or neck in any of them. (3) Dissection of blood through the deeper or external layers of the media of the non-atheromatous origin of the internal carotid occurred in all of them. (4) In each case the ipsilateral middle cerebral artery contained thrombus. (5) Ipsilateral cerebral infarction occurred in all cases. (6) In all there was cystic degeneration of the media of the internal carotid and other arteries.

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