THE VASCULAR ABNORMALITIES AND TUMOURS OF THE SPINAL CORD AND ITS MEMBRANES

By

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To

MY PARENTS

AND

THE MEDICAL STAFF OF
THE NATIONAL HOSPITAL

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FOREWORD

By DR. GORDON HOLMES

Till recently vascular anomalies and tumours of the central nervous system had not attracted widespread attention, but during the past twenty years monographs and papers by Lindau, Cushing and Bailey, Dandy, Olivecrona and others have made neurologists familiar with these conditions in the forebrain. On the other hand, though many individual communications have appeared on abnormalities and new growths of blood vessels in the spinal cord and its meninges, in which single or small groups of cases have been recorded, there has been hitherto no attempt to make a comprehensive and systematic classification of these conditions and to correlate the clinical symptoms they present with their nature and pathology. Even the nomenclature of the different forms they assume has been confused and indefinite. This is mainly due to the fact that few adequate pathological descriptions are available, and that in many of those published by pathologists the clinical symptoms and the mode of their development have not been adequately recorded. It is to these facts that the difficulty in their clinical diagnosis and the common failure to recognise their presence during life are due.

Dr. Wyburn-Mason's researches, in the course of which he observed a large number of cases and collated and reviewed critically the literature on the subject, should in the first place serve to draw attention to their relative frequency and to their importance in the clinical pathology of the spinal cord. His attempt to correlate clinical symptoms with various types of vascular anomalies and tumours also provides a basis for accurate diagnoses and is therefore a valuable contribution to neurology. Further experience may modify some of the conclusions to which he has come, for there is no finality in clinical medicine, but I have no doubt that on the whole they will stand the test of time. The practical importance of differentiation of different types and of their accurate diagnosis is obvious from the fact that in many types present-day methods of surgical interference cannot be successful and frequently aggravates the patient's disabilities.

Dr. Wyburn-Mason's introduction of kymography with the use of contrasting substances for the purpose of visual demonstration of pulsations in abnormal vessels is a valuable contribution to the subject of this monograph.

Of the clinical importance and practical value of this monograph I have no doubt, and I can only express the hope that it will receive the recognition to which the expenditure of much time, energy and exact observation entitles it.

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PREFACE

THE subject of this book is one which has received little attention in the past. This has been due to two main causes. The first is the difficulty in clinical diagnosis, which is itself dependent on the second, the failure to distinguish between the various pathological types of vascular anomalies and tumours which may occur in the cord. Further difficulties have arisen on account of the confused nomenclature and frequently the course of the disease has been so prolonged that it has been impossible to follow cases to autopsy. In many cases of vascular abnormalities, the diagnosis has been one of "myelitis," and no attempt has been made to elucidate the pathological processes involved. In my opinion many cases of so-called "myelitis" are in reality examples of vascular abnormalities and such an underlying pathology should always receive due consideration, particularly if the symptoms are recurrent.

Vertebral angiomata are not considered here as a considerable literature has accumulated about the subject in the last few years.

I wish to thank all those who have helped me in compiling this book by allowing me access to case records and pathological material, and especially to the Honorary Staff of the National Hospital, Queen Square, Dr. J. G. Greenfield, who made many of the pathological reports, Mr. Norman Dott, Mr. Harvey Jackson, Mr. T. G. I. James, Professor Geoffrey Jefferson, and Mr. Wylie McKissock. To Dr. Gordon Holmes I would tender my sincere thanks for much helpful interest, criticism and encouragement. I should also like to thank Mr. J. Anderson, who kindly took the photographs.

Mr. George E. Deed of Henry Kimpton has been an unfailing help in all matters connected with publication.

R. WYBURN-MASON.

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VASCULAR TUMOURS OF SPINAL CORD

CHAPTER I

Historical

THE earliest references to vascular lesions of the cord are those of Hebold (1885), Gaupp (1888) and Kadyi (1889). Since that time many single examples have been described, especially by Elsberg, in several papers from 1916–32. Sargent reported 5 cases in 1925. Globus and Doshay (1929), and later P. Guillain (1933) reviewed the subject and Mathieu (1931) that of venous abnormalities only. The first two papers consisted chiefly of a catalogue of some of the cases previously described with a classification into two broad groups, the angiomatous malformations and the vascular tumours, after the manner of Cushing and Bailey (see below). The latter were divided topographically according to their relation to the cord and meninges into extradural, extramedullary and intradural, intramedullary, and vertebral. This subdivision, although clarifying the position, offered no clinical means of diagnosis and grouped pathologically unrelated lesions together. The pathological findings were only briefly mentioned.

The whole subject is clouded by a loose and confusing nomenclature, one name being used to indicate two or more pathological conditions, and, conversely, a single pathological condition being designated by different names. Thus one and the same condition has been described as aneurysmal varix, cirsoid aneurysm, vascular dilatation, telangiectasis, racemose angioma, Rankenangiom (of German authors), angioma, hæmangioma and pial varix.

Of attempts at classification of vascular tumours in general, Virchow's is one of the best. He divided them into two large groups, namely, *angioma cavernosum* and *angioma racemosum*, the former being characterised by the absence of parenchyma between the blood-filled spaces, whereas in the latter each vessel forms a unit itself and the tumour consists of a jumble of vessels which are separated from one another by parenchyma. This second group, which has been called the "hamartomata," was subdivided according to the developmental stage of its vessels, into those with thin-walled vessels like capillaries, which Virchow termed *telangiectases*, and others, whose vascular walls suggest either arteries or veins—*angioma racemosum arteriale sive venosum*. Sometimes the blood flows directly from artery to vein through widened channels without an intermediate capillary bed. Virchow called such a short-circuit an *arterio-venous aneurysm*.

The work of Lindau (1926), Cushing and Bailey (1928), Roussy and Oberling (1931), and Bergstrand, Olivecrona and Tönnis (1936), has

added greatly to our knowledge of vascular abnormalities of the brain. Lindau drew attention to the frequency in which angiomatous cysts of the cerebellum were associated with an angiomatous tumour of the retina, previously known as v. Hippel's disease, and with multiple cysts and tumours in various viscera, such as the pancreas, liver, kidney, spleen and suprarenals. He also mentioned that tumours similar to those in the cerebellum might occur in the cord. Cushing and Bailey deal chiefly with cerebral lesions and divide vascular abnormalities of the central nervous system into two subdivisions—the vascular malformations and hæmangioblastomata. The former are distinguished by the presence of nervous tissue between the parts of the tumour and are essentially anomalies ; the latter are neoplasms and formed not only of vessels of different calibres, but also of a fine network of reticulum. They correspond to the tumours described by Lindau. Roussy and Oberling likened their structure to that of the reticulo-endothelial system and suggested the name angio-reticulomata for them. In addition, they claimed that in some cases glia formed an integral part of the growth and to such tumours (found in the cerebellum) they applied the name angiogliomata. Cushing rejected the distinction and considered these tumours as vascular gliomata. No such tumour has been reported in the cord.

Bergstrand *et al.* divide the conditions into :

(1) *Angioma cavernosum* (where there is no neural tissue between the vessels).
(2) *Angioma racemosum venosum.*
(3) *Arterio-venous aneurysm.*
(4) *Hæmangioblastoma* (LINDAU).
(5) *Telangiectasis.*

They maintain that, in the brain, angioma arteriale is indistinguishable from arterio-venous aneurysm pathologically and histologically, and that angioma racemosum venosum and arterio-venous aneurysm are distinguishable only physiologically, in the effect of the latter on the general systemic circulation and the presence of pulsation and a murmur.

However, there are objections to all the above classifications. Thus—(1) Some lesions that have no nerve tissue between the vessels, and hence should be grouped as true tumours, are more allied to the malformations—notably the cavernous angiomata. (2) In some cases malformation and tumour are associated in the cord (see below). One cannot, therefore, contrast tumour and anomaly too rigidly with one another. (3) There is good evidence that " cavernous " angiomata develop from telangiectases (Unna) and that both may occur in the same case, or that such a vascular lesion may in one part contain parenchyma between individual blood spaces, but elsewhere none may be visible. Hence it seems that there is no justification for dividing " cavernous " angiomata from telangiectases.

CHAPTER II

Classification

THE following groupings seem best to fit these lesions of the cord. They can be divided into vascular abnormalities and vascular tumours. Of the first group the purely venous anomalies consist of single or numerous abnormal veins which lie in the pia and invade the cord—the angioma racemosum venosum. This must be distinguished from the venous dilatation occurring below a spinal tumour.

Angioma racemosum venosum shall, according to Virchow, resemble a varicocele, *i.e.*, a knob of widened veins peripherally to a capillary bed. The vessel walls in different parts of the same vessel may be of different structure ; in one part a transverse section of a vessel has a well-developed elastica interna and muscularis and in others both of these are absent. The interna is occasionally so thick that the lumen is almost obliterated and sometimes absent, and in its place there is loose connective tissue. In contrast to the telangiectases (including cavernous angiomata) the venous racemose angiomata regularly affect the pia with ectasia of the pial vessels, especially in the cord.

The arterial anomalies have received scant attention previously, but in my experience they are at least as common as venous. According to Virchow's definition, angioma racemosum arteriale must consist only of arteries, both efferent and afferent vessels being such. A number of cases are described in the literature as occurring in the cerebrum and a few in the cord (*e.g.*, Pappenheim's case), but it is doubtful to what extent this description is correct. In most cerebral cases they have been confused with arterio-venous angiomata, to which they correspond both clinically and histologically. Such lesions of the cord are often referred to in the French literature as cirsoid aneurysms.

The arterio-venous aneurysm is a short cut between the arterial and venous system, either by way of a large artery communicating with a vein, or through a vascular knot replacing the capillary bed. The result is an increase in the size of the afferent artery and the efferent vein. In other parts of the body, if the vascular knot is large, it produces such an increase in the blood volume that cardiac hypertrophy results. Pulsation of the vascular knots and a blowing murmur over them are two of the classical signs of the condition, but only vascular pulsation has been recorded in the cord. The use of the term aneurysm in this connection seems dubious and apt to cause confusion. The term angioma is preferable, being justified by usage, but is misleading in suggesting a new growth rather than a malformation.

In the cord there are certain other arterial anomalies which deserve attention ; these may be associated with congenital heart disease. Occasionally the term racemose arterial angioma has been applied

to these. Sometimes an aneurysmal dilatation may occur in one of the spinal arteries in spinal meningo-vascular syphilis.

Another group is formed by the telangiectases with which the so-called cavernous angiomata may be included, the reasons for which have been already mentioned. Such lesions may occur within the cord or lie extradurally.

The vascular tumours include the hæmangioblastomata. They are characterised by autonomous growth. The hæmangioblastomata (Lindau's tumours) may occur in the cord or on the nerve roots, and in some cases may be associated with very enlarged and distended veins with, or without, the presence of syringomyelia, which alter the clinical picture. Similar tumours occur extradurally.

Finally, several cases of lymphangioma within the spinal canal have been reported, but their existence in most cases has been very doubtful.

Up to the present about 140 cases have been found in the literature but only about 120 are adequately described.[1] A further 67 cases are added.

The following classification will be adopted in this description :

(A) **Abnormalities**

(I) Venous Abnormalities

(*a*) *Secondary venous abnormalities*, that is, those occurring below a tumour of the cord, or associated with arachnoiditis or calcification of the cord.

(*b*) *Angioma racemosum venosum*, that is, extensive venous varicosities affecting the pia and central regions of the cord.

(II) Arterio-venous Angioma.

(III) Arterial Anomalies

(*a*) Associated with congenital heart disease.

(*b*) Alone.

(IV) Syphilitic Aneurysm of the spinal arteries.

(V) Telangiectases, including so-called cavernomata or cavernous angiomata.

(B) **True Tumours**

(VI) Hæmangioblastoma or Hæmangio-endothelioma.

(*a*) *Angio-reticuloma*, or *Lindau's tumour*, occurs in the cord or on a nerve root and may be associated with

[1] Since this was written O. A. Turner and J. W. Kernohan (*Arch. Neurol. Psychiat.*, 1941, 46, 444) have made a pathological study of 46 cases of vascular malformations and tumours of the cord. Their classification is essentially the same as that adopted here, except that they divide the true tumours into a large number of different types which, in my opinion, is unnecessary. They included 3 cases of epidural varices, a type which I have not seen. In 1 case of arterio-venous angioma of the cord, the muscles and skin of the corresponding metameres were likewise permeated by abnormal vessels,

syringomyelia, or with similar tumours elsewhere in the nervous system and cysts in other organs.

(*b*) *Extradural hæmangioblastoma.*

(VII) Lymphangioma.

Reference will be made in the appropriate sections to the frequency of these different abnormalities. Rassmussen, Kernohan and Adson (1940) found that of 557 intraspinal lesions, 8·5 per cent. belonged to the group of extramedullary vascular tumours, and that of 64 intramedullary tumours, 7·5 per cent. were vascular in nature.

CHAPTER III

Venous Abnormalities

THE criteria of normality of the veins of the cord are difficult to define as considerable variation may occur. The subject was investigated by Kadyi (1889), who published the only study of the subject. In a general survey of the vascular supply of the cord in 26 cases, he found that dilatations and tortuosities of the larger or smaller veins of the cord are not uncommon. In eight patients various degrees and several types of varices of the spinal cord veins were found. Even in the spinal cords in which the veins presented a normal appearance, he frequently noticed that the course of the veins on the dorsal aspect of the cord was somewhat tortuous. He, therefore, concluded that a careful study of a larger number of cases would disclose transitional forms, with gradual change from the normal condition to the marked varicosities, in which the venous trunks and their branches form a bizarre network and are often so extensive as to cover the surface of the cord completely.

(*a*) **Secondary Venous Abnormalities.** Whenever pressure occurs on the cord from an intra-spinal or vertebral tumour there is, as is well known, an engorgement and enlargement of the veins below the tumour, due presumably to the obstruction of the spinal circulation. This increase in the size of the veins is never very great however, but results in the so-called Froin's syndrome—an increase in the protein content, with xanthochromia of the cerebro-spinal fluid below the tumour. In some cases, notably with very vascular tumours, actual exudation from the tumour may contribute to the increase in the protein content and xanthochromia of the fluid (Greenfield and Carmichael). Ayer has shown that immediately above the tumour there is also an increase in the protein of the fluid, which decreases progressively the higher up the spinal canal the fluid is removed. This increase only extends over a few segments usually. Cushing and Ayer have shown also that, in tumours of the cauda equina, increase in the protein content and xanthochromia occur *above* the tumour.

A similar enlargement and engorgement of the veins occurs in association with arachnoiditis and arachnoid cysts, but is probably a secondary result to the venous obstruction caused by kinking of the vessel walls. An example of this condition was described by Lafora, when a recurrent arachnoidal cyst of the cauda equina was associated with an enlargement of the pial veins to some moderate degree. This type of enlargement and engorgement of the veins is never very marked however. In addition, a case has been reported by Hare and Everts in which, following trauma to the spine, calcification occurred in the cord, and this was associated with enlargement and increase in

the number of veins on the cord above the lesion. The calcified area was visible on radiography. Alterations in the spinal veins may occur also, secondary to thrombosis of the superior longitudinal sinus (McLean).

All the above types of venous enlargement, however, are comparatively slight in degree and they are mentioned only for completeness.

(*b*) **Angioma Racemosum Venosum.** This is the commonest type of vascular abnormality of the cord, according to the number of published reports of cases.

Occurrence. It is difficult to obtain accurate figures of the frequency of occurrence of this type of lesion. Cornil and Mosinger found it five times in 104 intraspinal tumours, whereas Schoenbauer found it twice in 80 laminectomies for spinal lesions. Elsberg states that six times in 130 successive laminectomies for spinal disease, he found one or several enlarged posterior spinal veins on the surface of the cord. Adson and Ott met it three times in 112 laminectomies for spinal tumours, in 27 of which no tumour was seen (but in 4 cases it was found at autopsy), *i.e.*, in 89 spinal tumours. Dandy found it four times in 75 spinal tumours, Linnep four times in 153 laminectomies (64 being intradural and extramedullary), and Sick once in 50 laminectomies. It would seem, therefore, that it occurs in 3–4 per cent. of cases of spinal tumour, not an inconsiderable number of cases.

The first description of such a condition was that of Gaupp in 1888, and Kadyi mentions it, but gives no clinical data. Since that time cases have been fully described by the following : Sargent, 4 cases ; Globus and Doshay, 4 cases ; Ritter, 2 cases (case 2 is referred to by Jungling) ; Rand, case 2 (case 1 was one of extradural varices, and not the condition under consideration, although included by Globus and Doshay as such) ; Elsberg (1916, 1917, who mentions 6 cases in all, but one was associated with an intramedullary tuberculoma) ; Spiller and Frazier ; Blahd ; Mühsam ; Frazier and Russel ; Lindemann ; Benda ; Löwenstein ; Perthes, 2 cases ; Hackel, case 2 ; Sachs ; Rosenhagen ; Gropalli ; Krause ; Kreiger-Lassen ; Kortzeborn ; Nonne, 2 cases ; Voss, case 2 ; Anderson and Dellaert ; Puusepp, 3 cases ; Roger, Arnaud and Alliez ; Bucy ; Marinesco and Draganesco (1935) ; Mathieu, 2 cases ; Belloni ; Sterling and Jackimowicz ; Fracassi ; Schoenbauer, cases 35 and 36 ; Alexander, case 2 (case 1 was a " dural varix ") ; that is 48 cases in all. Descriptions given by the last two authors are, however, unsatisfactory. In addition to the above, passing references to cases have been made by Adson and Ott (3 cases), Dandy (2 cases), Linnep (4 cases) and Sick. This brings the total up to 61 cases, only 44 of which are described in any detail. Hackel's case 1 was insufficiently described to know its nature. In my collection of 67 cases of vascular abnormalities and tumours of the cord, 14 cases of this type occur (cases 1–14), bringing the total up to 75

cases.[1] Many cases of unexplained spinal lesions probably belong to this group.

Pathological Anatomy. The variations from the normal have been mentioned above (Kadyi's findings). An analysis of nearly all the cases recorded shows that the pathological changes are remarkably uniform from case to case, but have never been accurately described. This condition has been confused by many writers with arterio-venous angioma. Failure to recognise the essential pathology has caused surgeons to attempt procedures which have led to disastrous results and the majority of observers to state that it is impossible to diagnose this condition pre-operatively. It will be shown that, in most cases, this should be possible with a fair degree of certainty.

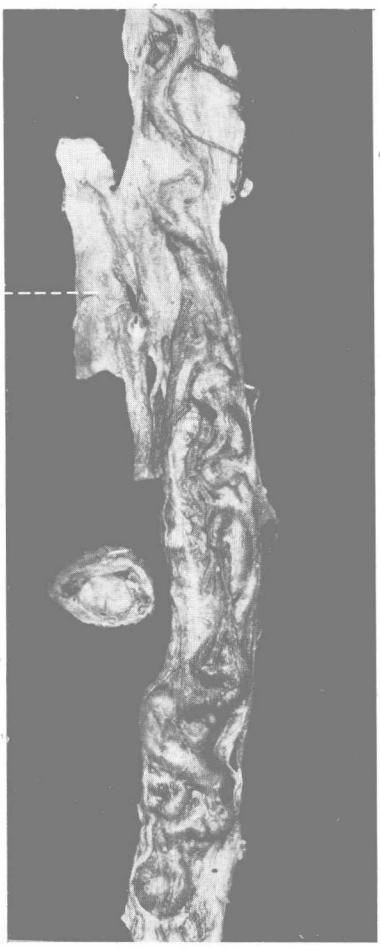

FIG. 1.—Angioma racemosum venosum of lower part of cord. Dorsal surface (case 5).

In every case reported the *condition affects the lower part of the cord and in no case has it extended higher than the D6–5 segment.* On reflecting the dura the lower part of the cord is usually seen to be covered on its *dorsal surface* by a mass of sinuous, anastomotic, turgid, blue pial veins. These are often in two layers and completely hide the cord. Many veins are seen to enter the cord from the mass. Passing upwards the condition gradually diminishes in extent, but it often extends down to the cauda equina. Sometimes the vessels are confined to a few segments only, as in case 10, or there is a localised knot of widened abnormal vessels. Occasionally, as in Elsberg's, Sachs', Rosenhagen's and Globus and Doshay's (case 2) cases, the vein is single and then it tends to occur in the lower dorsal region on the left side (Elsberg). The lateral and anterior surfaces of the cord are very much less affected than the dorsal surface, but the anterior spinal veins are usually larger than normal and more

[1] Since this was written 3 further cases have been reported by M. Schope (*J. Neurol. Psychiat.*, 1941, 171, 810), K. Hüber (*Ibid.*, 1941, 171, 799), and P. Delmas-Marsalet (*Presse Méd.*, 1941, 49, 964), but owing to the war the references are unavailable. In the last case repeated symptoms occurred in pregnancy and the typical lipiodol radiographs were obtained.

tortuous. The venous mass is drained by means of enlarged radicular veins. At its upper limit a large tortuous posterior spinal vein courses upwards for a few segments. (Fig. 1 is a photograph of such a condition.) In some of these cases there are meningeal adhesions and thickening present in the region of the angioma. However, the

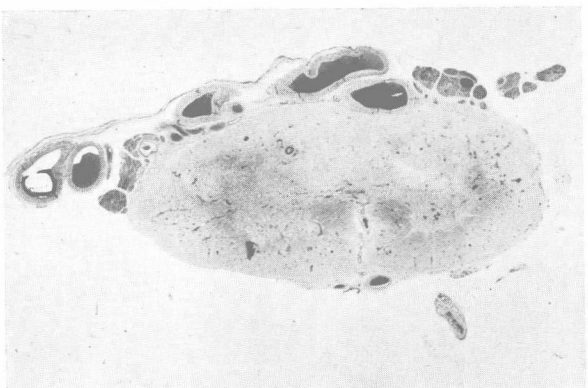

FIG. 2.—Cross-section of lumbar cord showing large veins on dorsal surface, numerous small intramedullary vessels and loss of cord structure (case 10).

external appearances of the cord are only part—and a minor part—of the condition, contrary to the impression gained by surgeons at operation. Macroscopically it is often difficult to distinguish an arterio-venous from a purely venous angioma. This is best done by their topographical position, by the region of the cross-section of the cord which is affected and by the presence of arterial vessels entering the mass. It is possible that some cases described as venous, *e.g.*, Perthes' case, were really arterio-venous in type.

If the cord is sectioned and examined microscopically, in the involved regions abnormal vessels are seen extending from the anterior median fissure into all parts of the grey matter and white matter adjacent to it. The nervous tissue is replaced by very numerous small blood vessels, chiefly capillaries, pre-capillaries and venules (Figs. 4 and 5), which destroy the cells of the grey matter and cause degeneration of the white matter with considerable gliosis in many cases (Fig. 6). The changes gradually decrease in extent on ascending the cord. The white matter shows much less change, but Weigert-Pal staining reveals degeneration of the ascending tracts, especially those in the ventro-lateral columns, and this may reach the cervical region. The pyramidal tracts are also markedly affected, but normally only in the later stages are the posterior columns involved to any extent and not most severely, as one would expect from the external appearances of the cord (Fig. 6). If the condition is of long standing there is diffuse glial overgrowth of the posterior columns, secondary to the ascending degeneration. The small vessels in the grey matter are thick-walled and

show hyaline changes and decrease in size of their lumina. The vessels in the dorsal pia are very wide. They are thick-walled and abnormal in structure (Fig. 3). The thickening involves all coats. The intima is very thick and often shows the presence of hyaline changes and an internal elastic lamina, like that in arteries. Their lumina are frequently partially or completely obliterated by organising or fresh thrombus. The media is also thickened and shows hyaline changes and often the presence of smooth muscle fibres.

In the late stages the affected part of the cord is completely structureless and soft, and shows overgrowth of glia in all areas. There is no evidence to suggest that a local solid tumour, with or without syringomyelia, ever develops from this condition, but occasionally

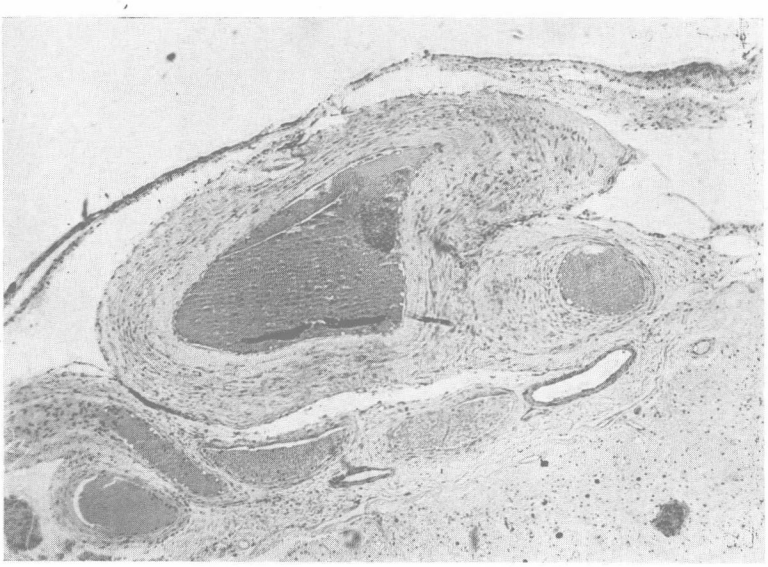

FIG. 3.—Cross-section of vessels on the dorsal surface of the cord (case 10).

small spaces are found in the grey matter. The appearances described were present in 4 of my series (cases 3, 5, 10 and 13) which were examined post-mortem, and Nonne, Sterling and Jackimowicz, Globus, Mühsam, Lindemann, Kreiger-Lassen, Fracassi, Kortzeborn, Hackel, Krause, Benda, Gaupp, Anderson *et al.*, and Marinesco *et al.* also found them. No evidence of active proliferation of the endothelium of the blood vessels is present and no mitotic figures.

Apart from the typical findings just described, cases also occur in which the pial veins are either unenlarged or only slightly so, but the lesions within the cord are typical. Such a case recently came under observation (case 67). This is of considerable interest, as it recalls the condition which Foix and Alajouanine, in 1926, called "subacute necrotic myelitis." They described two cases, both in males, in which

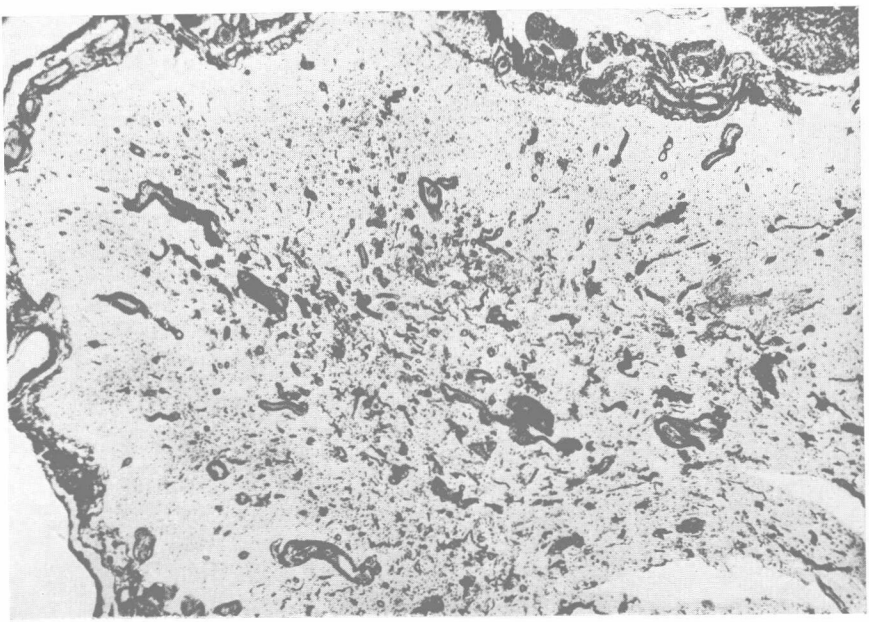

FIG. 4.—Cross-section of cord of a case of angioma racemosum venosum (case 13), showing the enormous increase in the number of small vessels in the grey matter. (Hæmatoxylin and v. Gieson × 30.)

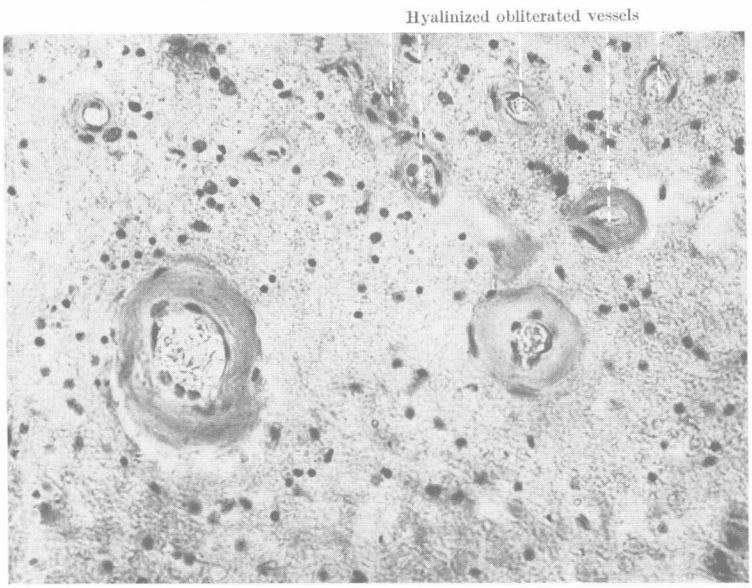

FIG. 5.—High-power view of part of anterior horn in lumbar region (case 5), showing numerous small thick-walled vessels with marked hyaline degeneration and partial obliteration of their lumina. The nerve cells have almost completely disappeared.