

CIRSOID ANEURISM OF THE SCALP*

WITH THE REPORT OF AN ADVANCED CASE†

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CONGENITAL telangiectases, vascular nevi, or angiomas, which occur so commonly on the face and scalp, may be the starting point of disfiguring aneurismal tumors. When the contributing arteries and the outgoing veins succeed in forming anastomoses through the cavernous bed of the nevus, the lesion becomes nothing more than a diffuse arteriovenous fistula. Such is the probable etiology of most so-called cirroid aneurisms in which the intermediary incidence of trauma in many cases seems definitely established.

In its mechanical effects, therefore, a cirroid aneurism resembles an arteriovenous fistula, with the difference that in the latter condition the communication is usually (though not invariably) single, whereas in the true cirroid the communications are multiple, and take place through the vascular meshes of the tumor.

The condition must have been recognized from the earliest times in view of its striking characteristics, and it would be an interesting study to trace in detail the early records of the lesion;‡ it is not at all improbable that some ancient example may have given rise to the legend of the serpents in the hair of Medusa which Perseus cured by radical though unprofessional measures.

It would appear that the term *anévrisme cirsoïde* (varix-like) was first used by Brescht in 1833, long after William Hunter's description of "aneurism by anastomosis." Though other designations were introduced, as *aneurysma serpentinum* by Cruveilhier, the term "cirroid," accepted by Verneuil and by A. Robert in 1851, as well as by Ch. Rubin in his excellent description of the erectile tumors three years later, has subsequently come to be the one generally employed in French and English literature.

In the German literature, however, the cases for the most part must be sought under other titles, due largely to the influence of Virchow, who proposed the term *aneurysma racemosum* or *ranken aneurysma*. Moreover, fully cognizant of the fact that a congenital angioma was often the starting point of these lesions, he designated them as *rankenangiom* or *angioma racemosum arteriale*.

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† To Dr. Harvey Cushing the writer is indebted for permission to publish this case. Obligations are also due to Mr. K. E. Appel, of the Harvard Medical School, for his aid in reviewing the literature.

‡ For anyone so disposed, the section in Rudolf Virchow's classical treatise on tumors (*Die Krankhaften Geschwülste*, vol. iii, p. 471 *et seq.*) would make a proper point of departure.

CIRSOID ANEURISM OF THE SCALP

Undoubtedly the cases in the early literature, many of which are cited by Virchow, comprise lesions of various kinds, not all of them in association with primary angiomas; for it is quite probable that arteriovenous fistulæ (more especially those with single or multiple communications involving the vessels of the face and neck) may arise through some faulty vascular development, which may leave one or more arteriovenous communications without the intermediation of a cavernous angioma.¶

From this preamble it may be gathered that there are several varieties of so-called cirroid aneurism arising in various parts of the body which deserve clinical and pathological differentiation. The present communication, however, will be limited solely to a brief consideration of the anastomosing aneurisms of one type and in one situation: namely, those which affect the scalp and which arise through the intermediation of a preëxisting vascular abnormality of a congenital, angiomatous nature.

In any outspoken example of this condition, the clinical picture is such a startling one that the case is likely to be reported. The extensive literature on the subject shows that these cirroid aneurisms more often occur on the face and head, whether for the reason that congenital vascular nevi are more common there or because the head is more exposed to the kind of trauma likely to provoke an aneurismal change in the preëxisting lesion is not apparent. Nevertheless, cirroid aneurisms of the advanced type observed in the patient whose case-history follows are not common. It is the first example of the condition in the *circa* 35,000 admissions to the Brigham Hospital.

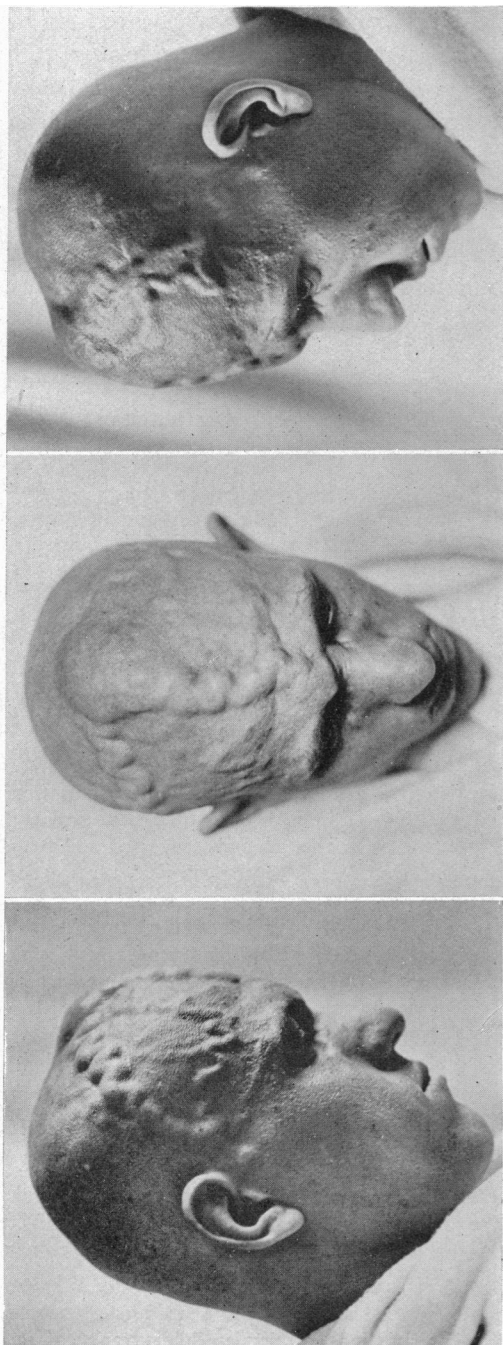
P. B. B. H. Surgical No. 17539. John A., an Italian wool-worker, aged twenty-two, entered the medical service of the Brigham Hospital, October 9, 1922 because of a chronic cough, "swellings" on his forehead, and headaches. Though suspected of having incipient pulmonary tuberculosis, he was soon transferred to the surgical service for treatment of his aneurism.

In 1918, while serving in the Italian Army, he had received some minor shrapnel wounds of the scalp, and was subsequently taken prisoner. His wounds which were confined to the occipital region, required no especial care, and though none of them appear to have affected the central region of the aneurism, he states that the top of his head was badly contused at the time.

To this trauma, he naturally ascribes the present condition, for soon afterward, he first noticed the pulsating swelling under the frontal hair margin, which has gradually increased in size. He soon began to be conscious of a roaring sound in his ears and at present this bruit and the startling appearance of the huge vein in his forehead constitute his chief annoyances.

Examination showed a vigorous, alert young Italian who, aside from a chronic bronchitis of doubtful origin, appeared to be in good general physical condition. The outstanding feature of his case lay in the lesion thus described by Doctor Cushing in a

¶ It is quite possible that the recent case reported by F. L. Meleney (*Surg., Gynec. and Obst.*, 1923, vol. xxxvi, p. 547) was of this type. The patient had a small arteriovenous aneurism under the chin. Doctor Meleney has supplied "a complete bibliography of the literature as given in the *Index Medicus* since 1879." This bibliography, however, is confined almost entirely to articles which have appeared under the title of "cirroid aneurisms" whereas almost an equal number of important articles, chiefly in the German literature and otherwise entitled, do not appear in his list.



Figs. 1, 2 and 3.—Condition before operation, October 18, 1922.

note dictated shortly before the operation, when the accompanying photographs were taken (Figs. 1, 2 and 3):

“Before this man’s head was shaved, there was discernible through the hair a faintly reddish, soft, warm, pulsating tumor measuring 6 by 6 cm., slightly elevated above the surface of the rest of the scalp and situated in the midline about at the coronal suture. Radiating from this tumor in all directions but chiefly laterally and anteriorly are huge, tortuous vessels, the appearance of which naturally suggests the snakes in the head of Medusa. Through the hair, the slightly prominent central area, owing to its discoloration and increased heat, gave the impression of an inflammatory process on the verge of ulceration. After shaving, however, it becomes evident that the central lesion is a reddish nevus, doubtless congenital, of the port-wine-mark variety which overlies a cavernous angioma.

“The enlarged and tortuous vessels which radiate from the lesion are chiefly confined to the anterior half of the scalp. The three main outstanding channels are evidently veins, though they show visible pulsation and have a palpable thrill. The largest of these is a broad vessel measuring nearly 4 cm. at its widest point which passes downward toward the root of the nose. There it bifurcates and extends on each side as far as the alæ. Another large tortuous vein passes off from the right side of the central swelling and extends down toward the anterior part of the ear, where, like the aforementioned vessel, it becomes gradually narrower and finally disappears. There are

similar vessels on the left side of the scalp, though less marked than those described.

“These vessels, which are obviously huge pulsating venous channels, are easily distinguished from the arteries, which, however, are likewise unduly prominent and

CIRSOID ANEURISM OF THE SCALP

unusually tortuous, and which can be palpated all the way from their points of appearance on the scalp directly into tumor. This is particularly true of the greatly dilated anterior branch of the right temporal, which can be traced directly into the site of the main central lesion; the same is true of the left temporal as well as the supraorbital arteries. The suboccipital arteries are likewise full and large, and pulsating vessels can be felt over the scalp in the occipital region as well as in the anterior areas where, however, they are particularly pronounced.

"While the patient is sitting up, it is possible to empty the large vessels by pressing upon them near their point of emergence from the main central lesion, proving that they are efferent veins. Moreover, they are visibly pulsating, and there can be no question, therefore, but that arterial circulation through some form of communication is thrown directly into them. The central tumor or swelling is pulsating and apparently contains a large lake of blood. Its tension varies considerably, depending upon the position of the patient, but even when he is sitting up and when all of the arteries possible are compressed, it still remains tense and pulsating.

"Pretty much over the entire scalp is heard an astounding bruit, which is audible to the patient and which is precisely like that accompanying an arterial venous aneurism. It is curious, however, that this bruit though heard all over the head is louder at the lower margins of the scalp, particularly in the temporal region and over the glabella than it is directly over

the central pulsating tumor itself, where, as a matter of fact, the bruit is scarcely to be heard.

"The X-ray plates show greatly dilated sphenoparietal grooves, and it is quite probable that there is a dilatation of the meningeal vessels corresponding to that of the extracranial vessels. The meningeal artery consequently may participate in the process, and it is possible that the dilated meningeals may communicate with the longitudinal sinus, and thus with the extracranial pulsating mass which has been described. This seems the more probable in view of the fact that when a tourniquet is applied around the head shutting off the extracranial arterial supply there is an enormous increase in the size of the veins of the 'caput' which distend to such a degree that rupture seems impending. This observation makes it appear that a bilateral ligation of the external carotids will be a necessary preliminary step to the operation."

Doctor Cushing's operative note November 4, 1922, is in large part as follows:

"Ligation of each external carotid artery. Reflection of scalp, disclosing cavernous

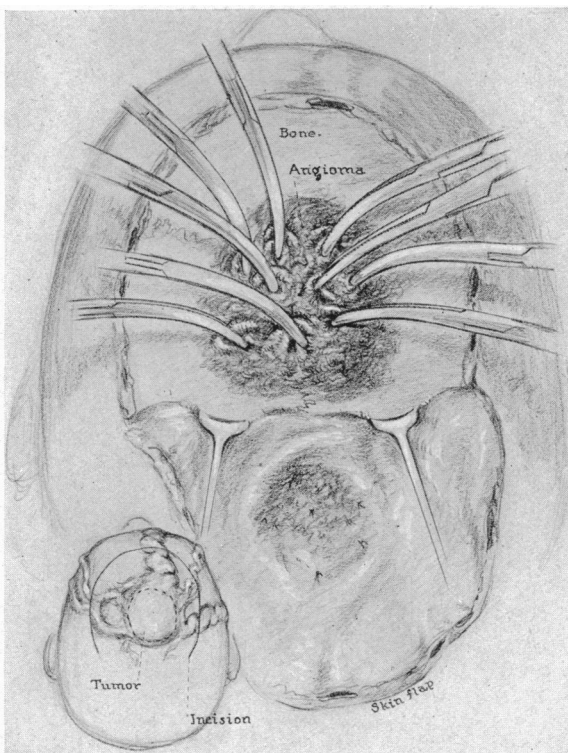
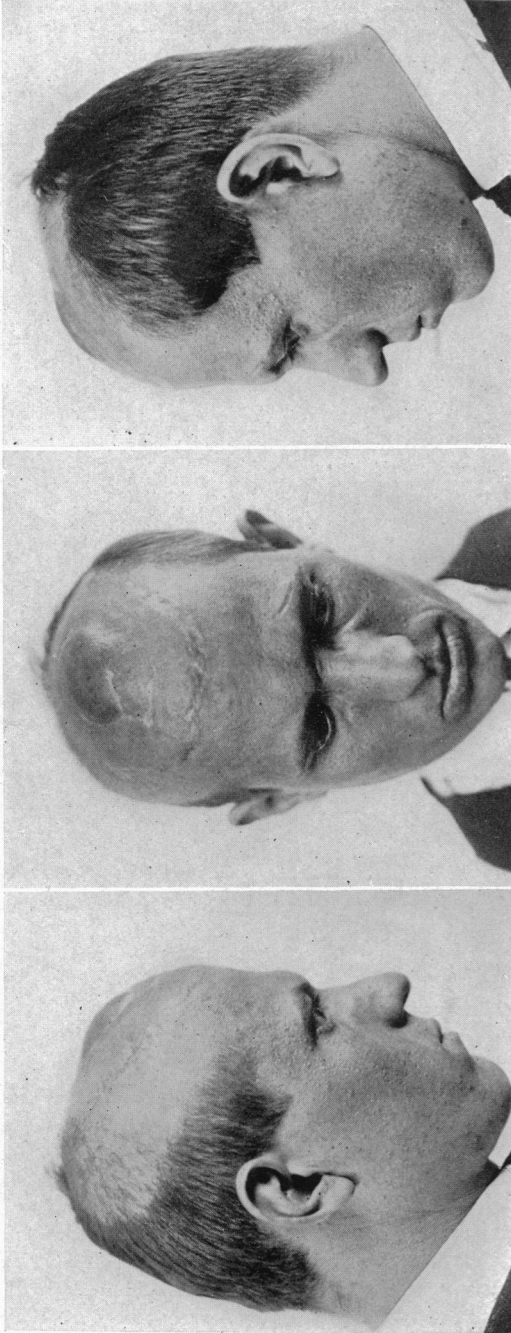


FIG. 4.—Operative sketch indicating procedure which was followed after extensive carotid ligation.



FIGS. 5, 6 and 7.—Condition February 2, 1923, three months after operation following radiotherapy.

angioma. Puckering of angioma by multiple sutures. Replacement and suture of scalp in layers.

“The external carotids were exposed, first on the right side where the vessel was immediately ligated then on the left, where a provisional ligature was thrown around the vessel and a bulldog clip applied so that the circulation might be reestablished should a slough of the scalp be threatened.

“As shown in the accompanying sketch (Fig. 4), a horseshoe-shaped incision with its base posteriorly was then made around the central pulsating angiomatous mass. By the usual method of finger compression of the scalp, this incision was carried down through the galea and the huge vessels at each side were caught by clamps. Naturally there was no arterial bleeding but the large veins remained as full apparently as before the carotid ligations. On reflecting the flap of the scalp in the subaponeurotic layer, the chief angiomatous tumor was exposed. It seemed to lie more or less within its own pericranial capsule from which the scalp could be dissected away without great difficulty. Fortunately, this central lesion had a great deal of fibrous tissue so that it could be caught by a series of heavy curved clamps (cf. sketch) as the scalp was peeled away from it. Subsequently a multitude of through-and-through silk sutures were taken where the tumor had been gathered up in these clamps: thus the growth was thoroughly ‘puckered’ in the hope that thrombosis might be encouraged.

“There was no special difficulty in this procedure and the hæmostasis was fairly complete. The scalp was then replaced and closed carefully in layers as usual without any special attention being paid to the individual dilated vascular channels. The bulldog

CIRSOID ANEURISM OF THE SCALP

clip was then removed from the left external carotid, but the arteries in the scalp promptly filled and it was evident that there would be some bleeding. The provisional ligature therefore was tied before closing the wound in the neck."

The circulation in the scalp seemed perfectly good when the usual dressing was applied. However, a very slight superficial slough of the flap margins occurred as the broad scars in the accompanying photographs (Figs. 5, 6 and 7) make evident. The central lesion was subsequently radiated to encourage endothelial destruction and thrombosis.

As is evident from this description, the operation consisted in an attempt to interrupt the vascular channels by an incision encircling the central lesion in the hope of producing thromboses which would cure the fistulous character of the lesion without removing the benign angioma itself. The operation was less formidable than anticipated and it is possible that the cavernous portion of the angioma underlying the galea might have been subsequently removed. This would have meant carrying the dissection down to the skull and removing the tumor together with the pericranium, but, as stated in Doctor Cushing's note, it was assumed that the lesion had vascular connections through the skull with the sinus longitudinalis and was possibly fed by the meningeal vessels. The outcome of the operation showed that this was apparently a mistaken idea: and though the central angioma therefore might have been removed, it is an innocent vascular anomaly in which the possible acquirement of further aneurismal characteristics seems remote. The lesion remains quiescent at the present writing, a year after the operation.

Comment.—As stated in the introductory paragraphs, it is the purpose of this paper merely to put on record another outspoken example of cirroid aneurism of the scalp with no pretense of covering the full literature of the subject. A large number of these cases have been recorded and many of them have been successfully treated either with amelioration or with cure by a great variety of surgical procedures—by single or bilateral carotid ligations; by multiple circumferential ligations; by attempts to obliterate the lesion with the galvanocautery; by acupressure; by the injection of thrombosing or scar-forming substances; or indeed by extirpation.

Of the many remarkable examples in the literature, possibly the case best known to surgeons was that reported by H. Müller¹ in 1891 from the clinic of Paul Bruns. For his paper was accompanied by a drawing (Fig. 8) which has passed down through several generations of texts and has consequently familiarized many with the appearances of the lesion on post-mortem dissection. As is so common, the patient had a red birth mark at the hair margin of his forehead which greatly increased in size and by his twentieth year had acquired the appearances shown in the dissection. Bruns first ligated the right external carotid and on attempting the same thing on the other side encountered severe bleeding and was compelled to ligate the left common carotid. Hemiplegia followed with a fatality and Ziegler made the studies which have given us this striking illustration of the lesion.

¹ Ein Fall von arteriellem Rankenangiom des Kopfes. Beitr. z. klin. Chir., 1891, vol. viii, pp. 79-91.

To be sure, many other examples occur in the German literature, the surgical aspects of which are chiefly emphasized, some of them representing very formidable conditions. An excellent résumé of the subject up to 1883 was given by Hermann Kümmell.² One of the better known cases subsequently reported is that by Paul Clairmont³ in 1908, the lesion having been attacked in two stages without preliminary carotid ligation. Another outspoken case reported by Max Kepler⁴ was operated upon by Heineke in

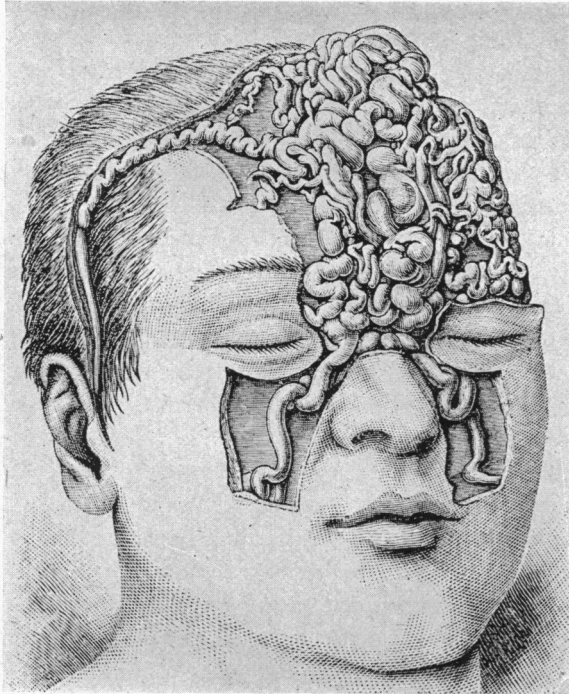


FIG. 8.—Dissection of a plexiform angioma of the forehead. (After H. Müller.)

1910: a huge cavernous cirroid of the parieto-occipital region, a very desperate and bloody procedure carried out in two stages with a subsequent skin-graft.

The French literature likewise contains a good many comparable examples and recently Noordenbos of Amsterdam has reported a case⁵ very similar to the one the subject of this report. At the age of twelve the patient had applied for removal of a small vascular tumor on his forehead, but operation was thought inadvisable. Twenty years later, because of its enormous increase in size and its tendency to bleed, inter-

vention was imperative. At operation the external carotid and frontal arteries were ligated on both sides. Catgut sutures were then passed around each afferent vessel and tied over a small roll of gauze. The whole tumor mass was then excised down to the periosteum. Subsequent skin-graft resulted in complete recovery.

The question of an intracranial extension of the lesion which was feared in the case herein reported has arisen in the minds of a good many others and

² Zur Behandlung des Angioma arteriale racemosum. Arch. f. klin. Chir., 1883, vol. xxviii, pp. 194-213.

³ Zur Behandlung des Angioma arteriale racemosum. Arch. f. klin. Chir., 1908, vol. lxxxv, p. 549.

⁴ Zur Behandlung des Aneurysma arteriale racemosum. Beitr. z. klin. Chir., 1912, vol. lxxviii, pp. 521-536.

⁵ W. Noordenbos and R. de Jong. Aneurysma arteriale racemosum van het Hoofd. Nederlandsch Tijdschrift voor Geneeskunde, 12 Oct., 1918, vol. ii, pp. 1224-1235.

CIRSOID ANEURISM OF THE SCALP

has been thought probable in view of the fact that in a number of patients (*e.g.*, Clairmont's) epilepsy has supervened or else there have been definite evidences of mental deterioration. Indeed, the question was raised as long ago as by Guérin (1870) as to whether an extensive cirroid of the scalp could come into communication with the intracranial vessels and sinuses. It would appear, however, as was true of our patient, that there was no real ground for this apprehension and seemingly in all other cases in spite of the apparent grooving of the skull by the huge vessels, operation has disclosed no alteration in the skull or evidence of intracranial communication. There nevertheless have been reported a good many examples of aneurismal varices which have originated within the cranial chamber. This, however, is quite another subject.

Very little has been said in the foregoing paragraphs of the cases which have been published from American clinics—all of them, to be sure, very briefly recorded. The older medical literature antedating the great medical indices, if thoroughly perused, would of course be found to contain the records of some of these surprising lesions. Attention may be drawn to two of these early reports at least. One of them was put on record in 1853 by Dr. R. D. Mussey,⁶ Professor of Operative Surgery in the Miami Medical College at Cincinnati, Ohio, who described his case under the caption of "Aneurismal Tumors upon the Ear Treated by Ligation of Both Carotids." He speaks of the lesion as being "like an aneurismal varix which had arisen in a congenital nevus."

Another example will be found in a volume published in 1867 by J. Mason Warren,⁷ then surgeon to the Massachusetts General Hospital. The case, similar to our own, was described as a so-called cirroid aneurism of the scalp arising from a congenital nevus starting about at the midline in the roots of the hair "which gave him a very formidable aspect"; and Doctor Warren goes on at length to tell of the many and varied procedures which were carried out by ligatures thrown over needles, followed by the applications of caustic potash repeated between twenty and thirty times to destroy the central lesion.

There are doubtless other similar cases buried in the early American literature. The following list, which may be incomplete, will nevertheless serve to assemble most of the cirroid aneurisms of the scalp which since 1887 have been put on record in this country.

(1) T. M. MARKOE of New York (*Phila. Med. News*, 1887, vol. I, p. 270). A brief report of a case following trauma, with ligation of both external carotids and probable improvement.

(2) HERMAN MYNTER of Buffalo (*ANNALS OF SURGERY*, 1890, vol. xi, p. 93). A case with ligation of one external carotid followed by circumferential ligation of vessels. Improvement but no late report.

(3) WILLY MEYER of New York (*N. Y. Med. J.*, 1892, vol. lvi, p. 214). A very brief report possibly of an arteriovenous fistula rather than a true cirroid aneurism involv-

⁶ *Am. J. Med. Sc.*, 1853, vol. xxvi, p. 333.

⁷ *Surgical Observations with Cases and Operations*. Ticknor and Fields, Boston, 1867. Case cclxii, p. 451.

D. C. ELKIN

ing the temporal region. Primary ligation of external carotid with excision of aneurism. In the discussion, cases were mentioned by J. D. Bryant and Kammerer.

(4) WILLIAM D. HAMILTON of Columbus, Ohio (N. Y. Med. J., 1894, vol. lx, p. 550). Ligation of one common carotid. Improvement but no late report.

(5) W. S. FORBES of Philadelphia (Phila. Med. News, 1895, vol. xlvi, p. 663). A case almost exactly like our own, with central pulsating tumor following trauma. Treatment, acupressure of entering vessels and subsequent excision of central tumor. Recovery.

(6) WILLIAM B. COLEY of New York (ANNALS OF SURGERY, 1901, vol. xxxiv, p. 414). Unilateral case with involvement of right temporal region; not advanced. Ligation of right external carotid followed by direct attack on tumor. "Nearly one hundred ligatures were applied." Recovery, but result uncertain.

(7) CARL BECK of New York (ANNALS OF SURGERY, 1903, vol. xxxviii, p. 496). "Aggravated case" following trauma. Ligation of temporal and other arteries with extirpation of tumor. Profuse hæmorrhage. Recovery.

(8) RUDOLPH MATAS of New Orleans (New Orleans Med. and Surg. J., 1908-9, vol. lxi, p. 469). Brief report at a society meeting probably a true post-traumatic arteriovenous fistula, not a cirroid case, though so reported. A bilateral, pulsating exophthalmos was present following fracture of the base of the skull. Ligation of both external carotids. Temporary relief with slowly progressing recurrence.

(9) E. S. JUDD of the Mayo Clinic (St. Paul Med. J., 1916, vol. xviii, p. 48). Perhaps the most extreme example of the condition on record. Ligation of both external carotids followed a week later by bilateral reflection of scalp and dissection of vessels. Probably complete cure.