Cerebral Angiomas

Advances in Diagnosis and Therapy

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Preface

The basic principles of the management of cerebral arteriovenous malformations were established during the first phase of the neurological attack on these problems between 1930 and 1960. The leaders were CUSHING, BAILEY and DANDY, but principally OLIVECRONA, and in Germany TÖNNIS. The experience gained showed that complete excision of the arteriovenous angioma was the only certain cure, and therefore was the procedure of choice.

In the present second phase important advances should be made and indeed are occurring. New diagnostic techniques such as total angiography, selective and superselective angiography, intraoperative and fluorescein angiography, and the EMI-scanner have been developed. The pathophysiological aspects have been further investigated by indirect and direct measurement of local and general cerebral blood flow.

Parallel with these developments operative technique itself has been improved and modified by new methods. A more aggressive attitude has been stimulated towards those angiomas, which had to be regarded as inoperable only a few years ago. Among these many improvements and technical advances include microsurgical techniques, combined stereotactic and microsurgical procedures, artificial embolization of different kinds and the cryosurgical management.

Multiple variables such as the age of the patient, the type, localization, and size of the angioma, its clinical picture and the possible complications, such as hemorrhage have been analysed and are understood better. These factors influence the indication for, and choice of, the appropriate procedure to a great extent.

The 20th anniversary of my work in Giessen, after 20 years of neurosurgery at Giessen University, has given the occasion for the discussion of the present position with regard to assessment and treatment of inoperable angiomas in a workshop together with experts in this field. The workshop, which was held Jan. 11.–12. 1974, was possible thanks to the generosity of Mr. and Mrs. H. KRÖNER of Fresenius Company, Bad Homburg. This volume contains the papers and discussions of this meeting. The interest, which it aroused, led to rapid publication. The editors take great pleasure in expressing their thanks to the contributors for their participation and cooperation, and to Springer-Verlag for personal and technical aid in preparing and publishing the proceedings.

ipation and cooperation, and to Springer-Verlag for personal and technical aid in preparing and publishing the proceedings.
We hope that they may contribute to giving a more precise answer to the problem of how to deal with patients who suffer from the so-called inoperable cerebral angiomas.

Giessen, November 1974 HANS WERNER PIA

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The Indications and Contraindications for Treatment or Assessment

Introduction

H. W. PIA

It is believed that Fedor KRAUSE was one of the first neurosurgeons to operate upon cerebral angiomas when in 1908 in Berlin he tried to ligate the feeding vessels. However, the era of operative treatment did not start until the thirties during which period CUSHING and BAILEY reported 9 cases, DANDY 8 cases, and OLIVECRONA, TÖNNIS, and BERGSTRAND published their classical monograph in 1936. They established the principles of diagnosis and treatment: diagnosis based on the angiographic evaluation of the structure of the malformation, the number, localization, and function of the feeding arterial vessels; and treatment based on the complete extirpation of the malformation where possible. Out of 20 intracranial arterio-venous angiomas 3 were totally excised, the first by OLIVECRONA in 1932. In 2 cases the attempt had to be abandoned, 4 cases were only explored, 3 patients had the intracranial arterial feeding vessels ligated; in 4 patients ligature of the internal carotid artery was performed, and 3 patients were treated by X-ray therapy.

Total excision was limited to small angiomas, in whom the occlusion of the feeding vessels was possible before the draining veins were approached. Angiomas situated in the motor area, the Sylvian fissure, and the speech areas were considered inoperable. "The risk of rendering a patient with seizures and perhaps only slight disturbance of function hemiplegic and aphasic is not abolished by the technique of vessel ligation which is of doubtful value". The intracranial ligation of vessels was successful in only one case with a single feeding artery.

This pioneering work of OLIVECRONA and TÖNNIS is particularly impressive even on review of their results 20 years later. OLIVECRONA and LADENHEIM reported in 1957 a series of 125 angiomas, out of which 81 were totally excised. 7 patients died, 50 were cured, 15 improved, and 7 became worse. TÖNNIS reported a series of 134 angiomas, out of which 56 were totally excised. 5 patients died. 61% of the survivors had full working capacity, 22% were partially employed, and 11% were invalids. Other large series from KRAYENBÜHL (15a) with 72 cases, MCKISSOCK and HANKINSON (21) with 106 cases, NORLEN (23) with 57 cases, KRENCHEL (16) with 98 cases, and FRENCH, CHOU and STORY (12) with 82 cases confirmed such results.

OLIVECRONA summarized his experiences in 1957 with the following words: "As matters stand today, if a patient with a lesion, which meets the standards of operability undergoes extirpative surgery under hypotensive anesthesia, the likelihood of surgical mortality should be negligible. About 85% of these patients will be clinically improved with

complete excision of the angiomatous tissue and freedom from future hemorrhage. Few of the remaining 15% will suffer any detriment.

1. The lesion may be totally removed, but the preexisting symptoms will be somewhat aggravated. This is a small premium to pay for insurance against further vascular accidents.

2. A small residue of angiomatous tissue may remain, but the symptoms will be improved.

3. Residual tissue may persist with aggravation of the preoperative symptoms. This last group may require further surgical intervention at a later date".

In spite of the overwhelming evidence of the effectiveness of extirpation of operable angiomas even in such cases many questions remain unsolved. This concerns particularly the prognosis of such important symptoms as epilepsy and hemorrhage which of course has a bearing on the question of the indications for surgery. Epileptic seizures were abolished in approximately 50% of patients (MCKISSOCK, TÖNNIS, NORLEN). In a few cases extirpative surgery led to epilepsy. Elimination of the danger of hemorrhage by total extirpation must be considered in relation to its prognosis which in spite of a recurrent hemorrhage rate between 40 and 60% is often quite good.

Palliative surgical procedures proved to be unsatisfactory. Out of 44 cases treated by OLIVECRONA 14 died, 2 following surgery, and 12 probably from later intracranial hemorrhage. 30 patients survived, 14 in good condition, but 9 were partial and 7 were complete invalids.

Afferent ligation, as performed by DANDY, should be considered only in cases where one single artery feeds the malformation, but even here the temptation should be resisted as arteriography is not infallible, and contributing vessels may not be demonstrated.

The electrocoagulation of surface vessels (TRUPP and SACHS) gave unsatisfactory results.

Carotid ligation should be given up. Not only was there no effect on the angioma, but the procedure was dangerous.

After ligation the blood flow through the angioma was derived from collateral sources, thus increasing, as we would say now, the steal effect. Radiation, as employed by CUSHING, was without the slightest discernible benefit. OLIVECRONA took a strong stand against the revival of this ineffectual form of therapy for arterio-venous malformations.

The indications for operation followed the rules of tumor surgery. Angiomas located in the functionally less important areas were considered to be surgically approachable. Small angiomas in the motor cortex were also removed; but in larger lesions the postoperative deficit constituted a greater handicap than the preoperative occassional seizure. Angiomas in the Sylvian fissure were often removed.

Intracerebral hemorrhage per se did not contraindicate prompt surgery when the condition of the patient might otherwise be endangered by delay, for the removal of the clot often exposes a partially mobilized lesion.

In angiomas in the region of the basal ganglia and internal capsule, in large malformations of the dominant hemisphere, and in most angio-

mas of the cerebellum and cerebello-pontine angle, surgery was contra-indicated. The same was true in cases of epilepsy of long standing, as there was little to gain from operation.

"In the absence of an absolute rule to guide his decision, the surgeon must carefully plot each borderline case against the abscissa of danger and the ordinate of benefit".

The last decade has brought important advances. The clinical symptomatology and course, and last but not least the natural history of unoperated cases has been studied and understood better, especially in the cooperative study by PERRET and NISHIOKA (26) and by WALTER (1970). Further progress was made by the introduction of preoperative total angiography and peroperative angiography, superselective angiography (DJINDJIAN), and magnification angiography. These investigations improved the diagnosis of small malformations and our knowledge of the pathomorphology of angiomas.

The pathophysiological aspects were advanced among others by SCHIEFER and TÖNNIS (39) and FEINDEL (9, 10) who elaborated upon the general and local cerebral blood flow disturbances. Current investigations by BECK and BAUER on the influence of the cerebral shunt on the heart and circulation may contribute to the better understanding of local and general compensation and decompensation mechanisms.

Parallel to these developments operative technique itself has been improved and this has stimulated a more aggressive attitude towards angiomas in functionally important areas. KUNC (18) reported on 97 angiomas out of which 58 were operated on; 7 were located in the speech area, 19 in the sensori-motor cortex, 5 in the Sylvian fissure, and 27 close to the above regions. Radical removal was possible in 48 cases with 8 deaths. On the other hand SANO, AIBA and JIMBO (36) stressed the high morbidity. In their series of 108 patients, 81 were operated upon. There was only one postoperative death, but 81% of patients had definite though variable residual neurological deficits.

The indications for radical removal of medial and paramedial angiomas are far from well defined and considerable reluctance is apparent. An important contribution was made by RIECHERT and MUNDINGER (35) who introduced a combined stereotactic and conventional procedure. The advantage of their method is the precise localization of the angioma with the possibility of an approach which causes the least functional disability. RIECHERT reported in 1972 16 cases of deep lying malformations on which successful operations have been carried out.

The use of the operating microscope was an undubitable and important step forward in the treatment of angiomas as was shown by RIECHERT and many others at the microsurgical symposia in Zürich (1968), Vienna (1972), and Kyoto (1973).

BUSHE (3) in a series of 42 cases treated 6 angiomas located in the midline, 7 in the ventricular and paraventricular region, and 2 in the basal ganglia. Radical removal was performed in 10 cases, vessel ligation in 2, and no operation in 3 cases. 2 patients died. DRAKE reported recently on 6 angiomas in the midbrain and pons, all successfully removed. The review of WALTER and BISCHOF (45) comprising 72 cases with angiomas located in those regions revealed that there were only 16 patients in whom the angioma was completely extirpated and 3 of them died. 16 further patients had partial ligation and out of this number 6 died.

Angiomas of the cerebellum were reported less frequently in earlier series. Latterly VERBIEST (42), PIA (29), LAPRAS (1972) and YASARGIL (46) and others have shown that even huge angiomas of this area have become accessible due to improved technique especially the use of the microscope.

Huge angiomas of the external and internal carotid system are regarded as inoperable in most cases.

My own contributions to the emergency treatment of angiomas associated with intracerebral and intraventricular hematomas (PIA 1968, 1972) have shown that this can be a life-saving procedure with considerable benefit also on morbidity. The operation risk is minimal, a contraindication exceptional.

It is to LUESSENHOP that the credit is due for introducing artificial embolization in 1960 for the treatment of inoperable angiomas. He also showed the effectiveness of such treatment. Incomplete embolization of the angioma may facilitate or permit a total extirpation at a second stage. SANO in 1965, and in Germany TZONOS, BERGLEITER and PAMPUS, as well as SEEGER, have applied and modified the method of LUESSENHOP. A further step forward was made by DJINDJIAN in 1970 who combined superselective angiography with embolization in a one stage procedure and reported excellent results in arteriovenous malformations of the skull and soft tissues of the face as well as in spinal angiomas. A glomus tumor was successfully treated by embolization by HEKSTER, LUYENDIJK and MATRICALI (13).

The freezing technique was developed by WALDER (1970) in order to thrombose the pathological vessels of inoperable and deep seated angiomas. Cryosurgery may also be used as a complementary procedure after incomplete extirpation of a deep lying angioma.

X-ray therapy of arteriovenous angiomas is almost generally regarded as ineffective. However, in 1955 POTTER reported on 10 patients treated by X-ray therapy out of which one died and 9 survived, 4 of them for more than 20 years.

The similar experience of JOHNSON (14) suggests that X-ray therapy may exert a favourable effect at least upon some forms of arteriovenous malformations, so that we may have to reconsider the indications for X-ray therapy in selected inoperable cases.

The basic principles of management of cerebral arteriovenous malformations were laid down during the first phase of therapy between 1932 and the mid-sixties. Experience showed that complete excision of the angioma was the only method of cure, and therefore was the procedure of choice.

In the present second phase not only have new techniques been developed but also multiple variables such as the age of the patient, the type, location, and size of the angioma; its influence on the cerebral and general circulation, the clinical picture and possible complications, such as hemorrhage and deficits produced by surgery, were analysed and understood better. These factors influence the indication for and choice of the appropriate procedure to a great extent.

The subject of this workshop is the treatment of inoperable angiomas. I think that starting from this borderline position, we should try to define the principles of management of patients with such lesions. I hope also that we will try to define objective criteria for the choice

of treatment of different types of angiomas in general. The participation of so many distinguished experts from different specialties makes me confident that we will come closer to our aim of answering more precisely the question of how to deal with those patients who have so-called "inoperable angiomas".

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Chapter I
Morphological Aspects

The Morphology of Centrally-situated Angiomas

K. JELLINGER

Angiomas are localized collections of blood vessels, abnormal in structure and in number, and composed of normal or malformed arteries, veins, capillaries, or a mixture of these. Within the CNS, vascular malformations are usually congenital in origin and caused by faulty embryologic development. They represent persistence of a primitive pattern of vascular pathways with anomalous structures and altered hemodynamics. Though not true neoplasms, these vascular hamartomas may grow and inflict acute or progressive damage on the brain tissue.

Incidence

The rarity of these vascular lesions suggested by COURVILLE (3) is neither confirmed by clinical experience nor by personal figures from two autopsy series in a University Medical Center and a Regional Neuro-Psychiatric Center, where CNS angiomas were found in 0.35 and 0.87% of the cases respectively (Table 1).

Table 1. Incidence of cerebral vascular malformations (CVM)

COURVILLE (1945):	30000 autopsies (1100 cerebral hemorrhages)	- 29 CVM (= 0.1 %) 18 telangiectasias 5 venous angiomas 5 venous varices 1 AV angioma
Institute of Path. Univ. of Vienna (1969-1971)	5553 autopsies (166 cerebral hemorrhages)	- 20 CVM (= 0.35 %) 6 telangiectasias 1 cavernous angioma 1 venous angioma 12 AV angiomas
Neuro-Psychiatr. Hospital Linz (1968-1972)	1600 autopsies (43 cerebral hemorrhages)	- 14 CVM (= 0.87 %) 3 telangiectasias 2 cavernous angiomas 3 venous angiomas 6 AV angiomas

In neurosurgical material the incidence of cerebral angiomas varies from 0.5 - 7% and was 3.5% of the brain tumors confirmed by biopsy 1964-1972 at the Dept. of Neurosurgery, Univ. of Vienna, and 5% of the total of brain tumors in the files of the Neurological Institute Vienna. Vascular malformations are responsible for 10 - 40% of surgically treated intracranial hemorrhages (11) and for 1 - 5% of cerebral hematomas found at autopsy (5).

Classification

Several types of vascular malformations in the CNS have been described. They have been classified 1. by size: a) small ("cryptic") angiomas (less than 2 cm in diameter), b) medium-sized (2-4 cm), and c) large angiomas (over 4 cm); 2. according to the location: a) supra- and infratentorial; b) superficial: cortical, subcortical; c) paramedial: basal ganglia and ventricles; d) medial: corpus callosum and brainstem; e) solitary, multiple and diffuse; and 3. by morphological appearance, a simple scheme distinguishing: a) capillary telangiectasias, b) cavernous angiomas, c) arteriovenous malformations, d) venous angiomas including varix formation. For review of the various malformations and of the distinguishing anatomical features of each of these types (7, 11).

Table 2. Morphological type of angiomas by location

a) McCORMICK et al. (7)

Type	Cerebral (n=346)	Cerebellum (n = 164)	Brainstem	Total
AV Angioma	217	52	18	287
Cavernous	59	11	10	80
Venous	46	20	11	77
Telangiect.	22	6	32	60
Varix	2	2	2	6

b) Personal series: 117 autopsies and 83 biopsies( )

Type ($^{+}$mult. 8)	Cerebral Hemisph.	Choroid Plexus	Basal Ganglia	Cereb ellum	Brain Stem	Total
AV Angioma	33^{+}(73)	4 (2)	12^{+}(1)	4 (2)	12	56(78)
Cavernous	4^{+} (3)	-	1^{+}	-	2	7 (3)
Venous	10^{+}	2 (1)	-	3	4	19 (1)
Telangiect.	5^{+} (1)	-	1	5	15	26 (1)
Total	52 (77)	6 (3)	14 (1)	12 (2)	33	117(83)

Table 2 compares the morphological type and location of cerebral angiomas in the series reported by McCORMICK et al. (7) and in a personal material of 200 cases (117 autopsies and 83 biopsies).

Morphological Types

a) Telangiectasias are small, mostly solitary groups of abnormally dilated capillaries separated by normal neural tissue. They are usually found incidentally at necropsy. Unlike other small or "cryptic" angiomas, they are most uncommonly associated with spontaneous hemorrhage. The most common site is the pons, less often the cerebellum and cerebral hemispheres are involved.

b) Cavernomas are composed of closely clustered, sinusoidal thin-walled vessels with no intervening nervous tissue. They range in size from pinpoint lesions to foci of several cm in diameter. They may be multiple and have been found in all parts of the CNS including the brain stem where they may cause fatal hemorrhage. Neither of these types of lesion have extra development of the vessels of supply which is typical for the next two types.

c) Arteriovenous (AV) angiomas constitute the most frequent vascular malformations of the CNS with greatly dilated and thickened vessels. There are irregular vascular cavities formed by arteries and veins of all sizes closely clustered and replacing the intervening and adjacent nervous tissue. Their size ranges from small examples to extensive tortuous masses occupying large parts of the brain. They occur in all parts of the CNS, but the larger ones most often involve the area supplied by the middle cerebral artery (6). Previously regarded as rare in other locations, almost 25% of all AV malformations are found in the posterior fossa (Table 2). Secondary degeneration, with fibrosis, venous dilatation, calcification, thrombosis with organisation, and evidence of old or recent hemorrhages, is frequently found. A rare variety in infancy and childhood is a dilatation or varix of the great vein of Galen which is often associated with obstructive hydrocephalus and congenital cardiac defects (9).

d) Venous angiomas are composed entirely of veins with one or more large draining veins. As in AV malformations, but unlike cavernomas, nervous parenchyma is found (Fig. 10). They occur in all parts of the CNS but are considered to be less common in the brain than in the spinal cord.

e) In small "cryptic" vascular malformations ("microangiomas") which are undetected by clinical investigations the gross appearance is unrewarding. The disclosure of these small lesions which include all morphological types, though chiefly AV angiomas, is dependent on careful microscopic examination. They have been found in all parts of the CNS.

Major Sites and Associated Lesions

With regard to treatment the morphologiacal classification of cerebral angiomas is of minor importance. The ideal treatment of a vascular malformation in the brain is radical excision, provided that the lesion is surgically accessible, and the risk of neurological deficit following surgery is minimal. Hence, the choice of treatment chiefly depends on the location and extent of the lesion and its vascular supply. Table 3 summarizes the major sites of cerebral angiomas, mainly AV mal-