THE ROLE OF SMALL ANGIOMATOUS MALFORMATIONS IN THE PRODUCTION OF INTRACEREBRAL HEMATOMAS

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The etiologic background of intracerebral hemorrhage falls readily into certain well defined categories, which include hypertensive-arteriosclerotic apoplexy, aneurysms of the berry or medial-defect type, angiomatous malformations, trauma, blood dyscrasias, and neoplasms. But in any series of cerebral hemorrhages one invariably encounters a group of unknown etiology. To this group certain features are common, namely:

1) occurrence in a relatively young age group;
2) absence of any known predisposing factors, except an occasional history of trauma;
3) suddenness of onset, usually with headache;
4) delayed loss of consciousness;
5) location of the bleeding usually within the white matter;
6) absence of a demonstrable anatomic cause for the bleeding.

Several excellent reviews have presented detailed discussions of all of the etiologic factors postulated for these hemorrhages. The various concepts dealing with the pathogenesis of apoplexy have been considered, but the age-distribution and the absence of significant arteriosclerosis in this group argue convincingly against vascular degenerative disease being the common background for these lesions. The relationship to trauma is inconstant, remote, and often equivocal. Infectious and toxic agents have been excluded in carefully studied cases. Most reports ascribe no definite causative factor. It is of historical interest that Hawthorne in 1922 was impressed by their occurrence in apparently healthy children and adolescents.

The relationship of large vascular anomalies to cerebral hemorrhage is well known, an incidence of 50 per cent being reported by Olivecrona and Riives, and a frequency of 41 per cent being found in a review of Dandy limited to arteriovenous aneurysms. But the role of small angiomatous formations in the production of intracranial hemorrhage is less well known. Three examples of bleeding from small macroscopic lesions have been reported from the clinic of Olivecrona. Hawkins and Rewell described 2 cases, and Bagley another, of fatal hemorrhage related to cavernous
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angiomas that were demonstrable only at a microscopic level. Kidd and Cumings discussed a family presenting a history of 10 deaths in two generations from cerebrovascular accidents occurring in youth. Two of these subjects were autopsied, one showing a hemorrhage associated with an angioma revealed in sections, the other presenting old and recent hematomas without a demonstrable background.

A survey of the problem of intracranial hemorrhage currently being undertaken at Duke Hospital has indicated (1) that a considerable proportion of hemorrhages are without demonstrable cause and (2) that small angiomatous malformations may play a more significant role than usually recognized. Over a 20-year period 14 hematomas of unknown etiology and 9 instances of hemorrhage from angiomatous malformations have been found, compared with 55 cases of hypertensive-arteriosclerotic apoplexy and 29 hematomas resulting from ruptured berry aneurysms.

Six fatal hematomas related to vascular malformations were studied at autopsy. Four were found to have been produced by rupture of small lesions. The minute size of one of these, the difficulty in demonstrating the second, and the deep-seated situation of the others point to their possible role in the production of so-called spontaneous intracranial hemorrhage. The incidental finding of two other examples of cerebral vascular lesions susceptible of being obliterated by related hemorrhage again emphasizes this possibility. These cases are being reported in this aspect.

CASE REPORTS

Two examples of massive fatal intracranial hemorrhage arose from vascular malformations that were not evident grossly. In Case 1, a tiny arteriovenous angioma was demonstrated only after multiple sections of the wall of a subcortical hematoma had been studied. In Case 2, a pontine lesion was so nearly destroyed by the related hemorrhage that its classification as a cavernous angioma is difficult. With neither of these lesions was the afferent or efferent circulation demonstrable.

Case 1. J. B., a 12-year-old white male, had sudden onset of a headache during a contest with his playmates “to see who could blow a harmonica the loudest.” The child became progressively less responsive and vomited frequently.

On admission 12 hours later temperature was 37°C, pulse 96, respirations 22, and B. P. 118/66. The principal neurological findings included semicoma, left hyperreflexia, and a left Babinski sign. The CSF was clear and free of RBC; the pressure was not recorded. Bilateral pyramidal signs developed and death occurred on the 4th day of illness.

Autopsy findings were irrelevant except for the cerebral changes. A massive subcortical hematoma, 6×4×4 cm., was found in the left parietal lobe, confined to the white matter (Fig. 1A). No subarachnoid bleeding was present. No gross source of the bleeding was demonstrated, and study of the lesion with a dissecting microscope gave no further indication of the background of the hematoma. The parenchyma in the wall of the hemorrhage was unaltered except for congestion and multiple small hemorrhages.